PV001 / #262

POSTER SESSION 01: IMMUNE SYSTEM AND AUTOIMMUNITY
03-06-2025 4:50 PM - 5:50 PM

METABOLIC STRESS ELEVATES TIGIT IN TREGS

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Background and Aims: A decreased number or dysfunction in regulatory T-cells is a key factor in many autoimmune diseases. Current approaches are centered on manipulating the cell machinery to promote tolerance and treat autoimmune conditions. This study aims to identify any changes in T cells between autoimmunity and health using a metabolomic approach.

Methods: In this study, we used T regulatory and T effector cells from healthy blood donors and type 1 diabetic patients. The cells were cultured for 11 days under GMP conditions. For cells isolated from healthy patients, we utilized a cell culture medium with different glucose concentrations (0, 100, 1000mg/dl of glucose) to mimic hypo-, hyper-, and normoglycemic conditions. On day 11 cells were harvested, washed, and suspended in PBS. Next, cells were analyzed using spectrophotometry to determine the activities of hexokinase, glucose-6-phosphate dehydrogenase, isocitrate dehydrogenase, lactate dehydrogenase, aconitase, and fatty acid synthase. Cell purity and phenotype were assessed using flow cytometry.

Results: T-regulatory cells cultured in conditions with a limited glucose concentration are characterized by an elevated expression of immune checkpoint receptor-TIGIT (Kruskall Walis test <0.0001). Additionally, the expression rates increase with the lowering concentration of glucose. There were no differences in enzymatic activity.

Conclusions: T regulatory cells may compensate for low glucose availability by overexpressing TIGIT, highlighting the key role of this receptor in Treg adaptation to metabolic changes and enhancing their immunosuppressive function. This insight could inform new treatments for autoimmune diseases, such as type 1 diabetes, by targeting TIGIT to boost Treg activity.

Keywords: autoimmune diseases, type 1 diabetes, metabolomics

PV002 / #299

POSTER SESSION 01: IMMUNE SYSTEM AND AUTOIMMUNITY
03-06-2025 4:50 PM - 5:50 PM

METABOLIC REWIRING SHAPES DENDRITIC CELL DISEASE PROMOTING FUNCTION IN AUTOIMMUNITY

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Background and Aims: Dendritic cells (DCs) possess a double-edged sword ability, to prime both immunogenic and tolerogenic responses, as deregulation of their proinflammatory potential can lead to excessive immune responses, a cardinal feature of autoimmune manifestations. Yet, the cellular and molecular mechanisms dictating DC imbalance in autoimmunity remain ill-defined. Driven by increasing evidence suggesting an essential link between DC metabolic alterations and function, we investigated whether autoimmune environments instruct such DC reprogramming and its potential implication in disease induction and progression.

Methods: Using experimental autoimmune encephalomyelitis (EAE), a multiple sclerosis mouse model and diverse single cell approaches,

Results: we identified an unprecedented shift in DC metabolic function. Specifically, bulk and single-cell multi-omic analyses revealed that DCs from EAE mice, compared to their counterparts from naïve mice, exhibited increased glycolytic behavior and a remarkable enrichment in pentose phosphate pathway (PPP). Moreover, the bioenergetic balance of DCs from EAE mice, on a single cell resolution, exhibited increased susceptibility to metabolic blockade of key glycolysis and PPP reactions. Importantly, our data demonstrate that abrogating the heightened activity of PPP has a functional impact on EAE DCs, as pharmacological inhibition of transketolase (TKT), a core PPP enzyme, reduced their maturation status and profoundly restrained their immunogenicity *in vitro*.

Conclusions: Collectively, our findings suggest that DCs undergo a robust metabolic reprogramming in autoimmune environments and reveal an until now unappreciated central role of PPP in this process. Thus, they define PPP as a new DC checkpoint in

autoimmune diseases and render its selective inhibition in DCs as an attractive therapeutic target.

Keywords: autoimmune diseases, dendritic cells, Immunometabolism

PV003 / #361

POSTER SESSION 01: IMMUNE SYSTEM AND AUTOIMMUNITY
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COMPONENTS OF DIFFERENT POLARITIES OBTAINED FROM BIDENS PILOSA L MODULATE THE ACTIVITY OF MACROPHAGES AND DENDRITIC HUMAN CELLS TOWARDS AN ANTI-INFLAMMATORY PROFILE

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Background and Aims: *Bidens pilosa L*. is a plant traditionally used by various communities in Colombia and around the world for its anti-inflammatory, antinociceptive and antioxidant properties. The immunomodulation of antigen presenting cells (APCs) such as macrophages and DCs with botanical extracts is currently an area of interest in the development of new treatments, specifically for autoimmune diseases. This study focused on evaluating the immunomodulatory effects of *Bidens* on PBMCs, macrophage polarisation (M0, M1 and M2) and human DC maturation.

Methods: Different polar and non-polar extracts and fractions were prepared from the aerial part of *B. pilosa* and used to determine their ability to modulate macrophage polarisation (M1/M2), DC maturation and cytokine production using multiparameter flow cytometry. The chemical components present in the extracts or fractions responsible for the modulation of the cellular response were identified using mass gas chromatography.

Results: The petroleum ether extract, the ethyl acetate and the hydroalcoholic fraction obtained from *B. pilosa* showed low cytotoxicity and modulated the PHA-stimulated proliferation of PBMCs. Furthermore, the *B. pilosa* petroleum ether extract induced M2 polarization or a hybrid M1/M2 phenotype in MØs and a semi-mature status in DCs, regardless of exposure to a maturation stimulus.

Conclusions: The immunomodulatory activity of the non-polar (petroleum ether) extract of *B. pilosa* on human PBMC proliferation, M2 polarization of MØs, and semi-mature status in DCs could be attributed to the low–medium polarity components in the extract, such as phytosterol terpenes and fatty acid esters.

Keywords: Autoimmunity, Immunomodulation, Bidens pilosa, Macrophages and Dendritic cells

PV004 / #135

POSTER SESSION 01: IMMUNE SYSTEM AND AUTOIMMUNITY
03-06-2025 4:50 PM - 5:50 PM

THE CASE FOR THE EXISTENCE OF MICROBIAL SENTINEL CELLS: ROUTES TO RHEUMATISM, DIABETES AND OTHER CHRONIC DISEASES

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Background and Aims: Background: While David Strachan gave us the "hygiene hypothesis", still the oft-quoted explanation for the rise in autoimmune disease, he neglected to mention the early 19th century description of hay fever by John Bostock, nor the observation of only 28 cases throughout Britain during his day [1]. We now know that hay fever is only one aspect of the so-called "atopic march", often starting with babyhood eczema [2]. 1. Bostock, J. Of the catarrhus aestivus or summer catarrh. *Med. Chir. Trans*. **1828**, *14*, 437-446. 2. Hill, D.A.; Spergel, J.M. The atopic march: critical evidence and clinical relevance. *Ann. Allergy Asthma Immunol*. **2018**, *120*, 131-137. **Aims**: To introduce the concept of microbial sentinel cells: hypothetical unicellular eukaryotes transferring immune-related information from one generation to the next. We suggest that it is the absence of these entities in the "modern western" microbiome that confers non-communicable disease upon subsequent generations.

Methods: This work relies only on literature review.

Results: We have recently published a "dual inheritance" mechanism, in which parental genes are complemented by a maternal microbiome delivered during the birth process. We speculate that key microeukaryote microbial sentinel cells are removed from the microbiome following interaction with the toxic metal ions liberated during industrialisation [3]. 3. Smith, D. et al. On the inheritance of microbiome-deficiency: paediatric functional gastrointestinal disorders, the immune system and the gut-brain axis. *Gastrointest. Disord*. **2023**, *5*, 209-232.

Conclusions: Our analysis points the way to both amelioration and prevention of future disease, if key microeukaryotes can be recovered from unaffected populations.

Keywords: Autism;, hay fever, hygiene hypothesis

PV005 / #155

POSTER SESSION 01: IMMUNE SYSTEM AND AUTOIMMUNITY
03-06-2025 4:50 PM - 5:50 PM

EVALUATION OF NUTRITIONAL STATUS WITH CLINICAL/LABORATORY TEST AND ANTHROPOMETRIC MEASUREMENTS IN SINGLE CENTER COHORT OF PATIENTS SUFFERING OF SYSTEMIC SCLEROSIS: LONGITUDINAL OBSERVATIONAL STUDY _

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Background and Aims: Systemic sclerosis (SSc) is a rare connective tissue disease characterized by multi-system involvement. The organs most frequently involved are skin and gastrointestinal (GI) tract. These are also those that have the greatest impact on the QoL of subjects, while pulmonary and cardiac involvements are those that have the greatest impact on life expectancy. The symptoms of GI involvement are different depending on the affected tract, dyspeptic symptoms and alternation of bowel habits are common. However, inflammation and fibrosis can affect not only the muscle layers, but also the intestinal mucosa responsible for nutrient absorption. This study aims to evaluate the nutritional status of a monocentric cohort of patients affected by SSc, compared with a group of healthy subjects.

Methods: The evaluation of nutritional status will be expressed through four different domains: anthropometry, nutritional anamnesis, use of specific questionnaires and analysis of blood chemistry parameters.

Results: We observed a higher risk of malnutrition in patients affected by SSc. This data emerged with particular statistical significance from the analysis of the MUST results and was partially confirmed also by the analysis of the PGSGA. Blood tests highlighted micronutrient deficiencies in patients affected by SSc. Instead there aren't statistically significant differences in the analysis of anthropometric measurements or nutritional history between the two groups.

Conclusions: The study confirmed the increased risk of malnutrition in patients with SSc, the risk does not appear to be secondary to a reduced intake of macro or micronutrients but rather to a picture of malabsorption.

Keywords: systemic sclerosis, Nutritional status, Malnutrition

PV006 / #165

POSTER SESSION 01: IMMUNE SYSTEM AND AUTOIMMUNITY
03-06-2025 4:50 PM - 5:50 PM

WOMAN WITH CONCURRENT VIOLACEOUS ULCERATED NASAL SKIN LESION AND RAYNAUD PHENOMENO: WHERE TH1 AND TH2 RESPONSE COEXIST.

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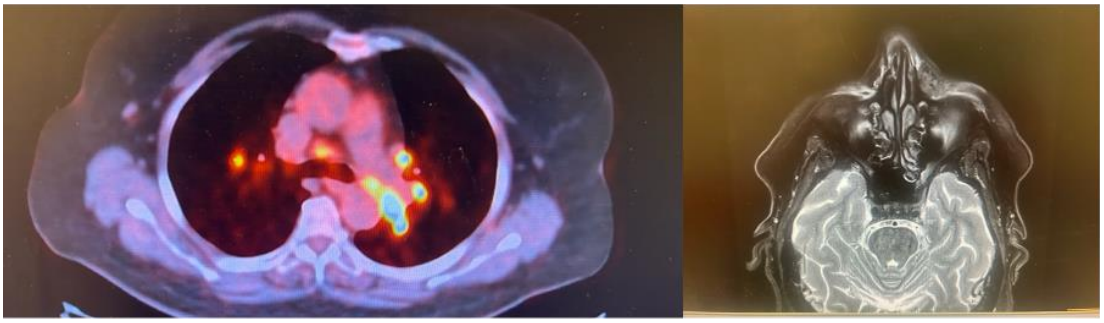
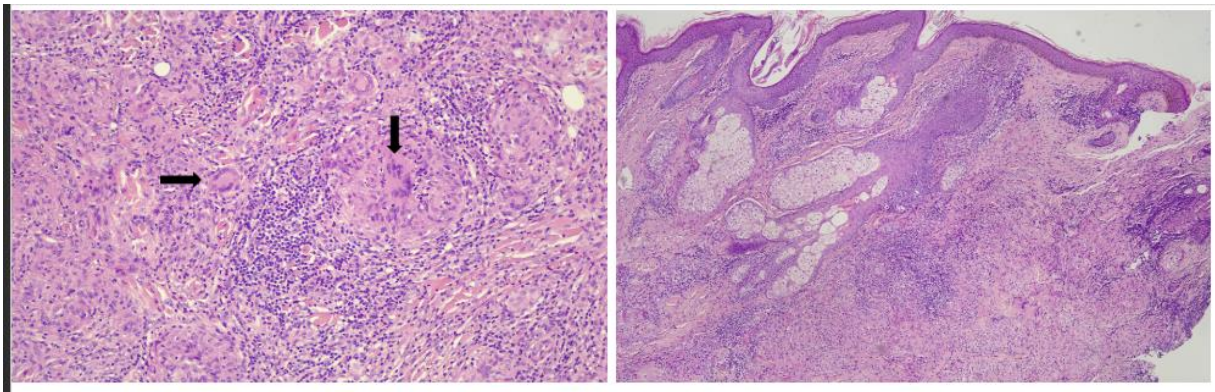
Background and Aims: Sarcoidosis is aTH1 response driven disease while Systemic Sclerosis a TH2. Lupus pernio(LP) is a distinct form of cutaneous sarcoidosis characterized by non-caseating granulomas.Raynaud phenomenon is the clinical hallmark of systemic sclerosis. We present the case of a woman with newly diagnosed lupus pernio and systemic sclerosis.

Methods:



Picture 2: Left image:PET-CT showed hypermetabolic mediastinal and hilar lymph nodes, lung nodules (<0.7mm) and 2 liver foci, Right Image MRI revealed soft tissue mass with contrast enhancement evading nasal bone causing a bony splinter to protrude

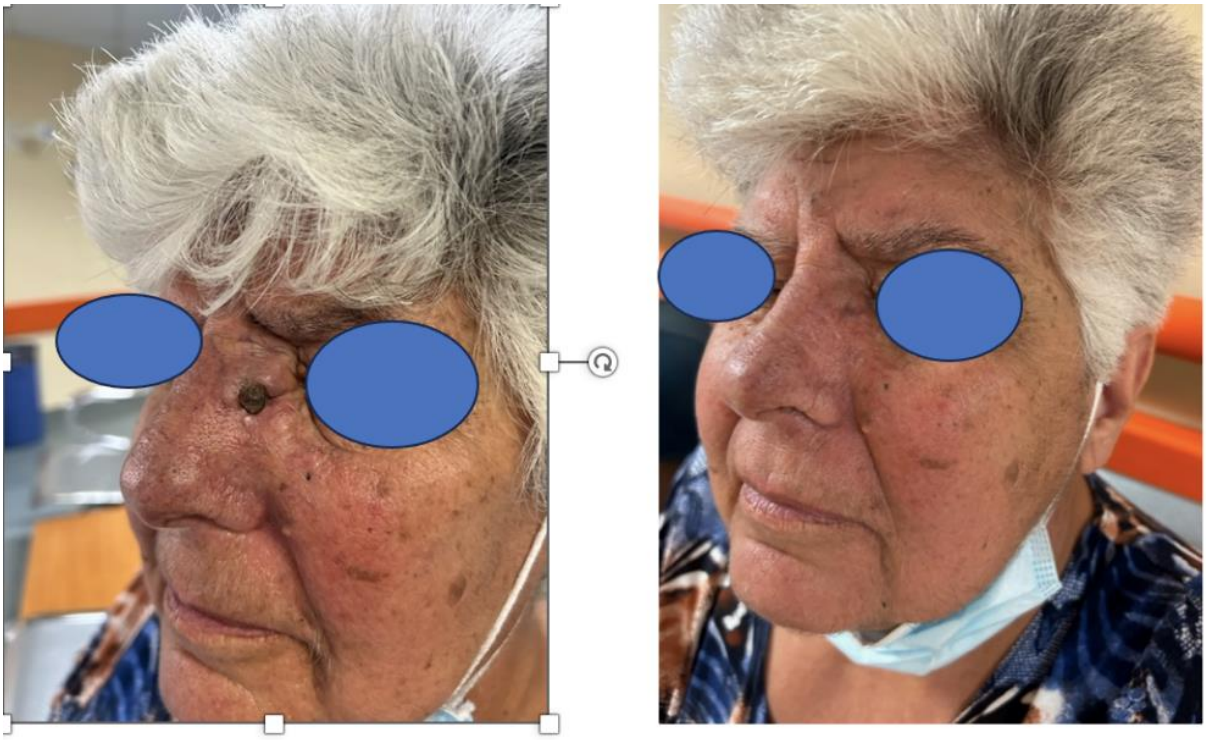


Picture1:Multiple noncaseating granulomas(black arrows with epithelioid histiocytes and multinucleated giant cells in the dermis compatible with sarcoidosis evading the underlying cartilage

A 74y.o. woman presented with violaceous cutaneous lesion on the left side of the nose over the last 6 months. The lesion was painless, growing, saddling the nose bridge and limiting vision. No signs of infection noticed. She reported Raynaud's phenomenon upon

exposure to cold, had puffy fingers and facial teleangiectasias. Capillaroscopy revealed dilatated capillaries and microbleeds. MRI revealed soft tissue mass evading nasal bone(Picture2). FNA and skin biopsy revealed multiple noncaseating granulomas penetrating the underlying cartilage(Picture1). Acid-fast stain was negative for mycobacteria and Mantoux test(-). Serum calcium level was normal. Immunology screen revealed ANA=1/1280 by IFL with ACA >200(NV<20 AU/ml), p-/c-ANCA and MPO,PR3(-).PET-CT showed hypermetabolic mediastinal and hilar lymph nodes(Picture2). In BALF CDA/CD8 ratio was increased=8.12(<4).

Results:



Picture 3:patient before treatment and 6 months after initiation of treatment

Treatement included local injections of triamcinolone, systemic corticosteroids pos and methotrexate with rapid response. The patient was monitored closely for scleroderma renal crisis which was not observed.

Conclusions: LP requires systemic treatment while steroids should be administered cautiously to SSc patients to avoid renal crisis. Coexistence of these two rare pathological entities cannot be pathophysiologically explained as they are caused by different autoimmunity pathways and must be considered a random phenomenon.Nevertheless,

clinicians should be aware of this possibility to fully explain symptoms and thus, guide treatment accordingly.

PV007 / #410

POSTER SESSION 01: IMMUNE SYSTEM AND AUTOIMMUNITY
03-06-2025 4:50 PM - 5:50 PM

SYSTEMIC LUPUS ERYTHEMATOSUS IN A YOUNG WOMAN: WHEN TO THINK OF AN INBORN ERROR OF IMMUNITY

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Background and Aims: Systemic lupus erythematosus (SLE) is known to mimic plenty of other diseases. The diagnostic of skin lesions in patients with SLE might be challenging in daily clinical practice. Pathogenic STAT1 gain of function mutations are associated with cutaneous lesions (mucocutaneous candidiasis) in the majority of the cases. Also, in some of these patients, serology suggestive of SLE may be present leading to a false diagnosis of SLE.

Methods: We analyzed the patient's medical history and files up to present.

Results: A 24-year-old patient was addressed to our department for generalized erythematous-squamous skin lesions. She had a diagnosis of SLE from childhood and was treated with systemic glucocorticoids, azathioprine and hydroxychloroquine with little benefit. Clinically, a generalized erythematous-squamous rash was observed, associated with scarred skin lesions and lateral cervical lymphadenopathy. Her laboratory tests showed hypochromic microcytic anemia and a slight inflammatory syndrome. She was positive for anti-dsDNA antibodies, without hypocomplementemia and proteinuria. The patient had a history of multiple skin biopsies indicating fungal skin infection, without findings suggestive of SLE-related skin involvement. Thus, we decided to perform genetic testing, conducting a primary immunodeficiency panel, which indicated a pathogenic mutation in the STAT1 gene – gain of function variant (c.820C>T p.Arg274Trp). Additionally, we decided to perform a thoraco-abdominal CT scan, which revealed a large aortic aneurysm (5.1/5.3 cm).

Conclusions: This case emphasizes the importance of including primary immunodeficiencies in the differential diagnosis of autoimmune diseases with atypical presentations and refractory to specific treatment. The large aortic aneurysm may significantly influence the patient's prognosis.

Keywords: lupus, mimicker, immunity

PV008 / #496

POSTER SESSION 01: IMMUNE SYSTEM AND AUTOIMMUNITY
03-06-2025 4:50 PM - 5:50 PM

CONTRIBUTION OF INNATE SIGNALS TO B-CELL DYSFUNCTION IN DIFFUSE CUTANEOUS SYSTEMIC SCLEROSIS

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Background and Aims: B cells play a crucial role in the pathogenesis of systemic sclerosis (SSc) and B-cell activation has been described as an early event of the disease. Abnormal B-cell activation and function can be influenced by innate immune molecules, such as Toll-like receptors (TLRs), complement receptors (CRs) and type I interferons (IFN-Is). Previously, we observed reduced expression of the TLR homologue CD180 molecule in B cells of diffuse cutaneous SSc (dcSSc) patients compared to healthy controls (HCs). Furthermore, we found that CD180 ligation had different effects on the activation and cytokine production of B cells in dcSSc and HCs. In this study, we aimed to investigate the effect of CD180 ligation on IFN-I signaling and CRs expression.

Methods: Peripheral blood mononuclear cells of dcSSc patients and age-matched HCs were isolated using Ficoll gradient centrifugation and to investigate the effect of activation via CD180 the cells were stimulated with anti-human CD180 antibody. Expression of IFN-I receptor (IFNAR) and CRs (CD21, CD35, CD11b, CD11c), and the changes in the phosphorylation of STAT1 were detected by flow cytometry.

Results: We found upregulated STAT1 phosphorylation and IFNAR expression in B cells of dcSSc patients compared to HC, which was further increased by CD180 ligation in dcSSc B cells. We observed a different expression pattern of CRs in dcSSc and HC B cells and the difference was further enhanced by stimulation via CD180.

Conclusions: These results suggest that B-cell activation via CD180 may contribute to B-cell dysfunction through IFN-I signaling and CRs.

Keywords: systemic sclerosis, B cells, CD180

PV009 / #200

POSTER SESSION 01: IMMUNE SYSTEM AND AUTOIMMUNITY
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OVERCOMING CHALLENGES: SECUKINUMAB SUCCESSFULLY MANAGES SPONDYLOARTHRITIS AFTER RECOVERY FROM MYCOBACTERIUM ABSCESSUS INFECTION – CASE REPORT

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Background and Aims: Introduction: Secukinumab, a fully human monoclonal antibody that directly inhibits IL-17A, has consistently demonstrated significant and sustained improvements in the sign/symptoms of moderate-to-severe spondyloarthritis (SpA). Non-tuberculous mycobacteria (NTM) infections are increasingly recognized as a global health concern, with species such as Mycobacterium abscessus causing serious opportunistic infections in humans.

Methods: This case describes the clinical course of a patient with SpA who successfully recovered from M. abscessus infection and was subsequently treated with secukinumab.

Results: Case report: A 52-year-old woman known to have SpA and bronchiectasis had admissions for management of Mycobacterium abscessus in 2022. She had received a combination of antituberculotic drugs and fully recovered. However, prolonged immobility during hospitalization led to a pelvic osteoporotic fracture, for which teriparatide was initiated. Following discharge, her SpA symptoms worsened, prompting an escalation in NSAID therapy, although the disease remained highly active. Simultaneously, she underwent physical rehabilitation and was followed by pulmonologists through high-resolution CT scans and sputum analysis. Given the persistent disease activity and following multidisciplinary evaluation, secukinumab

150 mg was initiated in May 2023. Three months post-initiation, the patient developed neutropenia, which was monitored by a haematologist. Despite this, secukinumab treatment led to a marked and sustained reduction in SpA activity.

Conclusions: Discussion: To our knowledge, this is the first reported case of secukinumab being introduced in a patient with SpA after recovery from M. abscessus infection. This case emphasizes the importance of further exploring whether anti-IL-17 therapeutics can be safely given to individuals convalescing from NTM infection, with benefits outweigh the risks.

Keywords: Mycobacterium abscessus, Secukinumab, Spondyloarthritis

PV010 / #220

POSTER SESSION 01: IMMUNE SYSTEM AND AUTOIMMUNITY
03-06-2025 4:50 PM - 5:50 PM

CASE REPORT: ANTI-FIBRILLARIN AUTOANTIBODIES INDUCED BY VIRAL MOLECULAR MIMICRY IN A PEDIATRIC PATIENT

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Background and Aims: Molecular mimicry of host antigens by virus is one of the leading mechanisms which, in susceptible individuals, may induce autoimmunity. Anti-fibrillarin autoantibodies (AFA) are detected in 7-48% of systemic sclerosis. The fibrillarin NH2-hexapeptide sequence is shared with an Epstein-Barr-virus (EBV)-encoded nuclear antigen. Here, we report the first case, to our knowledge, of a transient AFA positivity induced by EBV mimicry in a pediatric patient.

Methods: A 14-year-old girl was admitted to the Emergency Department with a history of vomiting, sore throat and fever started 6 days before.

Results: Laboratory tests revealed leucocytosis with a high lymphocytes count, an increased level of CRP, transaminases and total/direct bilirubin with normal liver function. On pediatric evaluation viral infections, celiac disease and anti-nuclear antibody (ANA) screening was required. High EBV-VCA-IgM and a slight increase of EBV-VCA-IgG were detected, helping establish a diagnosis of ongoing EBV infection. A positivity of ANA testing showing a clumpy nucleolar indirect immunofluorescence pattern, was observed on HEp-2000 substrate. Sample was tested for ELiA-CTD screen and Immunoblotting Scleroderma Profile and a positivity of anti-fibrillarin antibodies was determined. After a follow-up of six months, together with infection resolution, the negativity of ANA was determined, confirming the transient nature of the autoimmune phenomenon.

Conclusions: Our findings confirm how molecular mimicry may play an important role in the viral-induced autoimmunity. Given the significant homology between fibrillarin and EBV protein, caution in interpreting AFA positivity is suggested, especially in pediatric patients without clinical evidences of an autoimmune condition and a simultaneous screening for EBV infections is recommended.

Keywords: AUTOANTIBODIES, FIBRILLARIN, INFECTION DISEASES AND AUTOIMMUNITY

PV011 / #310

POSTER SESSION 01: IMMUNE SYSTEM AND AUTOIMMUNITY
03-06-2025 4:50 PM - 5:50 PM

TEMPERATURE, PH DEPENDENCY AND ACTIVITY OF MICROBIAL TRANSGLUTAMINASE AND ITS GLIADIN CROSS-LINKED NEO-COMPLEXES

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Background and Aims: Microbial transglutaminase (mTG) is a survival factor for bacteria and is heavily used as a food additive in the processed food industry. Being an enzyme, its temperature and pH range of activity are sensitive. Therefore, we studied the mTG temperature and pH operating ranges by exploring its capacity to cross-link gliadin peptides.

Methods: After optimizing the conditions to cross-link gliadin peptides by mTG (Zedira, Germany), temperature and pH dose-response curves were explored. Gliadin peptides, mTG, and cross-linked products were analyzed on SDS gels.

Results: mTG showed activity at 60°C by cross-link gliadin peptides. Also, various processed food products are not boiled during production processes. On the other hand, the mTG-gliadin docked complexes turn more immunogenic when heated to 90°C. Most probably, more epitopes are exposed to the immune system during denaturation. Concerning the pH impact on mTG activity, the enzyme is active at pH 4.0 and above.

Conclusions: Generally, during processed food preparation, the mTG cross-linked complexes are created before heating or boiling. The resulting covalent isopeptide bonds are incredibly resistant to the luminal proteases. During meal intake, gastric acidity is neutralized, and the pH can reach 4.5. Many children and adults consume acid-suppressive medications, infants and the elderly have a higher gastric pH, and alkaline reflux is not rare. Temperature and pH do not jeopardize the mTG induced cross-linking of gliadin peptides during food preparation. The stomach pH allows those cross-linked complexes to pass and reach the gut lumen.

Keywords: microbial Transglutaminase, Celiac Disease, Gluten Related Disorders

PV012 / #565

POSTER SESSION 01: IMMUNE SYSTEM AND AUTOIMMUNITY
03-06-2025 4:50 PM - 5:50 PM

EVALUATING GUT MICROBIOME IN AUTOIMMUNE DISEASES

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Background and Aims: The link between gut microbiome and autoimmune diseases has been a study subject in the last decade. Dysbiosis of gut microbiota has been shown to be related to alterations of the immune system and can contribute to the development of autoimmune diseases through several proposed mechanisms (translocation of pathobionts and their proinflammatory products as lipopolysaccharides (LPS), molecular mimicry, and disordered metabolome). Different studies have characterized microbiome alterations in patients with SLE, inflammatory bowel diseases, type 1 diabetes mellitus patients and in the pathogenesis of autoimmune liver diseases. This study aims to analyze the casuistry of our laboratory over the past five years emphasizing the clinical significance and potential applications of interventions based on gut microbiota in autoimmune diseases.

Methods: Gut microbiome was analyzed among patients with newly diagnosed AAV, systemic lupus erythematosus, rheumatoid arthritis, Sjogren's syndrome, type 1 diabetes mellitus, liver autoimmune diseases and inflammatory bowel disease. Fecal samples were collected using standardized methods (GutHealth® kit) and bacterial 16S rRNA genes analyzed by Next Generation Sequencing (NGS).

Results: The authors present 5 years revised casuistic from January 2020 to December 2024 as a reference clinical laboratory center in autoimmune diseases diagnosis focusing on gut microbiome profile among patients with several autoimmune diseases.

Conclusions: Data regarding the key role of gut microbiome in the pathogenesis of chronic autoimmune diseases is undeniable. Further investigations are needed to identify specific biomarkers that can accurately distinguish between healthy and compromised microbiota states in autoimmune diseases. These insights may reveal diagnostic markers and therapeutic targets in autoimmunity patients.

Keywords: Autoimmunity diagnosis, gut microbiota, microbiome

PV013 / #179

POSTER SESSION 02: SPONDYLOARTHRITIS DISORDERS
03-06-2025 4:50 PM - 5:50 PM

PREVALENCE AND CLINICAL CHARACTERISTICS OF HLA-B27 IN JAPANESE PATIENTS WITH PSORIATIC ARTHRITIS

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Background and Aims: The prevalence of HLA-B27 in PsA patients has been reported to be as high as 40-50% in some regions. However, in Japan, where the HLA-B27 carrier rate is relatively low (0.2-0.6%), there are few reports on the prevalence of HLA-B27 in PsA patients. This study aims to investigate the prevalence of HLA-B27 in PsA patients in Japan and examine the clinical characteristics of HLA-B27-positive cases.

Methods: We conducted a retrospective study of 342 PsA patients who attended Nippon Life Hospital between 2019 and 2020. HLA-B27 testing was performed, and the patients were divided into HLA-B27 positive and negative groups for comparative analysis. Clinical parameters such as pain, joint involvement, and inflammatory markers were analyzed between the two groups. Case studies of HLA-B27-positive patients were also presented, highlighting their clinical features.

Results: Out of the 342 PsA patients, only 3 were HLA-B27 positive, giving a prevalence rate of 0.88%. All HLA-B27-positive patients were female and presented with the axial type of PsA. In comparison to the HLA-B27-negative group, these patients showed distinctive clinical features, such as a higher frequency of axial involvement and a notable incidence of uveitis. Notably, the CRP was elevated in the HLA-B27-positive group.

Conclusions: The prevalence of HLA-B27 in Japanese PsA patients is lower than reported in other regions, at 0.88%. HLA-B27-positive cases were associated with female gender, spinal involvement, and an elevated risk of uveitis. These findings highlight the need for special attention to ocular complications in HLA-B27-positive PsA patients and suggest potential ethnic differences in the expression of HLA-B27-related PsA

Keywords: HLA-B27, Psoriatic Arthritis (PsA), Prevalence

PV014 / #185

POSTER SESSION 02: SPONDYLOARTHRITIS DISORDERS
03-06-2025 4:50 PM - 5:50 PM

IN SPONDYLOARTHRITIS TNF-A INHIBITORS NOT ONLY REDUCE DISEASE ACTIVITY BUT ALSO PREVENT FUNCTIONAL LIMITATION OF THE SPINE UNLIKE NSAIDS AND SYNTHETIC DMARDS.

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Background and Aims: Seronegative spondyloarthritis (SpA) is a group of inflammatory diseases characterized by axial and peripheral arthritis, enthesitis, and extra-articular manifestations. First-line treatment comprises NSAIDs, GCs, and DMARDs for peripheral SpA. Second-line therapies include TNF-alpha inhibitors, IL-17 inhibitors, and JAK inhibitors.

Methods: Data from 127 patients (92 males, 72.4%; 35 females, 27.6%; mean age 34.8±9.9 years; mean disease duration 6.06±4.93 years) were analyzed. Between January and June 2021, patients were treated with NSAIDs and synthetic DMARDs. BASDAI, BASFI indices, CRP levels, X-ray, and MRI were used to assess treatment efficacy. Non-responders were switched to TNF-alpha therapy after six months and reevaluated after two months. Paired-sample t-tests assessed differences in BASDAI, BASFI, and CRP values.

Results: Axial disease was present in 78/61.4% and peripheral in 49/38.6% patients. HLA-B27 positivity was found in 87/68.5%, with 4/3.1% uveitis cases, 75/59.1% with r-SpA, and 52/40.9% with nr-SpA. Treatment included GCs 35.4%, NSAIDs 91.3%,sulfasalazine 55.9%, methotrexate 9.4%, and leflunomide 2.4%.BASDAI, BASFI, and CRP were 5.12±1.2, 4.87±1.17, and 13.44±9.92, with no significant change after 6 months. MRI findings were unchanged in 31/24.4%, improved in 60/47.2%, and worsened in 36/28.3%.TNF-alpha therapy in 22 patients showed rapid improvement after 2 months:BASDAI 7.77±0.68 to 3.45±0.85, BASFI 7.55±0.85 to 3.5±0.85, and CRP 43.41±19.08 to 3.82±1.0 ($p<0.01$ for all).

Conclusions: In SpA NSAID and sDMARD therapy reduces disease activity, but doesn't prevent functional limitation of the spine. Patients who had negative response to conventional therapy rapidly improved after the prescription of TNF-alpha inhibitors. Moreover, TNF- alpha inhibitors may considerably improve quality of the life in SpA.

Keyword: TNF-alfa-inhibitors, spondyloarthritis, treatment

PV015 / #196

POSTER SESSION 02: SPONDYLOARTHRITIS DISORDERS
03-06-2025 4:50 PM - 5:50 PM

THE EFFICACY NSAIDS AND CONVENTIONAL SYNTHETIC DMARDS IN AXIAL AND PERIPHERAL SPONDYLOARTHRITIS

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Background and Aims: Seronegative spondyloarthritis (SpA) is a chronic inflammatory disorder primarily affecting the sacroiliac joints and axial skeleton, with potential peripheral arthritis and extra-articular involvement. NSAIDs are first-line treatment, while DMARDs and glucocorticosteroids may be added for peripheral SpA. This study evaluates the impact of NSAIDs and DMARDs on disease activity and radiographic progression in SpA.

Methods: Data from 105 patients (73male,32 female; mean age 35.12 ± 9.8 years; mean disease duration 5.53 ± 4.8 years)were analyzed from June to December 2021.Patients received NSAIDs and conventional synthetic DMARDs(csDMARDs).Efficacy was evaluated using BASDAI, BASFI, CRP, X-ray,and MRI assessments.Paired-sample T-tests compared BASDAI, BASFI,and CRP values.Results are presented as odds ratios (OR) with 95% confidence intervals (CI), with significance defined as $p<0.05$.

Results: Axial disease was diagnosed in 61 patients 58.1% and peripheral disease in 43/41%. HLA-B27 was positive in 68/64.8%,with uveitis in 2/1.9%, r-SpA in 54(51.4%), and nr-SpA in 51/48.6%.reatment included glucocorticoids (40),NSAIDs (etoricoxib,94), sulfasalazine (64),methotrexate (11), and leflunomide (3).At 6 months, BASDAI decreased from 4.86 to 4.62 ($p<0.05$) and CRP from 13.84 to 4.98 ($p<0.05$),while BASFI remained unchanged. MRI showed no change in 31 patients 29.5%,improvement in 38/36.2%,and worsening in 36/34.3%. Seven patients 14% of nr-SpA converted to r-SpA. The protective role of csDMARDs against radiographic progression was not significant(OR 0.28,95% CI 0.05-1.44,$p>0.05$).

Conclusions: In patients with axial and peripheral SpA NSAIDs and csDMARDs are able to reduce disease activity, i.e., pain, stiffness and CRP levels, but do not prevent functional limitation of the spine. Conventional therapy of SpA doesn't prevent conversion of nr-SpA to r-SpA.

Keyword: spondyloarthritis, NSAID

PV016 / #207

POSTER SESSION 02: SPONDYLOARTHRITIS DISORDERS
03-06-2025 4:50 PM - 5:50 PM

UNRAVELING THE HIDDEN BURDEN: SEXUAL DYSFUNCTION, MENTAL HEALTH, AND QUALITY OF LIFE IN WOMEN WITH NON-RADIOGRAPHIC AXIAL SPONDYLOARTHRITIS – A CROSS-SECTIONAL STUDY

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Background and Aims: Axial spondyloarthritis (ax-SpA) significantly impacts patients' well-being, yet female sexual function, a process influenced by both physiological and psychological factors, is often neglected in women with this condition. This study aimed to assess the prevalence of sexual dysfunction (SD), reduced quality of life (QoL), anxiety, and depression in female patients with non-radiographic ax-SpA (nr-axSpA).

Methods: We conducted a cross-sectional study involving 60 sexually active women diagnosed with nr-axSpA and an age-matched control group of 60 healthy women. Data were collected from electronic medical records and through three validated questionnaires: Female Sexual Function Index (FSFI), Hospital Anxiety and Depression Scale (HADS), and Short Form-36 (SF-36).

Results: SD was significantly more prevalent in nr-axSpA patients compared to controls (65% vs. 40%; p=0.006). The total FSFI score was lower in the patient group than in controls [19.71 ± 11.32 vs. 24.75 ± 8.36; p=0.006], with significant differences in the desire, arousal, lubrication and pain domains. Anxiety (HADS-A: 8.52±3.62 vs. 5.88±3.83, p<0.001) and depression (HADS-D: 6.27±3.38 vs. 3.28±2.77, p<0.001) scores were also higher among nr-axSpA group. Nr-axSpA individuals had significantly lower physical (171.0±72.9 vs. 301.5±81.2; p<0.001) and mental (204.9±83.9 vs. 277.1±74.5; p<0.001) QoL scores compared to their healthy counterparts.

Conclusions: Women affected by nr-axSpA exhibit greater prevalence of SD, elevated levels of anxiety and depression as well as lower QoL. Therefore, comprehensive healthcare addressing both physical and mental wellness is essential for these individuals.

Keywords: quality of life, Female sexual dysfunction, Non-radiographic axial spondyloarthritis

PV017 / #249

POSTER SESSION 02: SPONDYLOARTHRITIS DISORDERS
03-06-2025 4:50 PM - 5:50 PM

TH2 CYTOKINE PROFILE IN ATOPIC SPONDYLOARTHRITIS PATIENTS: CLINICAL CORRELATION FOR THERAPEUTIC APPROACHES?

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Background and Aims: The coexistence of Th2 and Th1-driven diseases is increasingly recognized, posing therapeutic challenges, particularly for eligible patients for biologic treatments. This study explores the Th2 cytokine profile in atopic patients with spondyloarthritis (SpA) to assess its clinical relevance and potential implications for therapeutic strategies.

Methods: The Th2 cytokine profile was analyzed in 39 atopic SpA patients, divided into biologically treated (BT) and non-biologically treated (NBT) groups. Atopic conditions included allergic rhinitis (AR), atopic dermatitis (AD), and allergic asthma (AA). Serum levels of interleukins (IL)-4, IL-5, and IL-13 were measured using a Luminex multiplex immunoassay.

Results: The cohort had a mean SpA duration of 13.64 ± 7.25 years, with a mean age of 49.25 ± 12.83 years. Atopy onset occurred after the third decade in 31 cases (79.49%). In the BT group (22 patients), 16 cases (41%) presented with AR, 5 (12.82%) AA, and 8 (20.51%) AD. These patients had been on biologics for an average of 7.9 ± 3.7 years (77.27% on TNF inhibitors). The NBT group (17 patients) had 8 cases (20.51%) of AR, 8 (20.51%) of AA, and 2 (5.13%) of AD. IL-4 and IL-5 serum levels were comparable between both groups. The BT group exhibited significantly higher IL-13 levels (1.6-fold, $p = 0.0006$), though no correlation was observed with SpA activity

Conclusions: The rising levels of serum IL-13 in biologically treated SpA patients with coexisting atopy suggest that incorporating small molecule inhibitors tailored to the patient's atopic profile could offer a viable treatment strategy.

PV018 / #301

POSTER SESSION 02: SPONDYLOARTHRITIS DISORDERS
03-06-2025 4:50 PM - 5:50 PM

BIOLOGICAL THERAPY IN PATIENT WTH COEXISTENCE OF MULTIPLE SCLEROSIS AND ANKYLOSING SPONDYLITIS: A CASE REPORT

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Background and Aims: Multiple sclerosis (MS) and ankylosing spondylitis (AS) rarely coexist. Both entities require biological drugs at some point of treatment, and given the fact that immunopathology of both is partly contradictory choosing a right biological drug for both without causing interactions may be challenging. Tumor necrosis alpha (TNF-α) inhibitors are a class of biologic drugs approved for treatment of AS. Therefore, these drugs should be avoided in treating AS in patients with definite MS.

Methods: We present a 40-year-old man diagnosed with multiple sclerosis in 2012. He was initially treated with pulse therapy of metylprednisolone without significant improvement of symptoms. In 2017 he started taking teriflunomide, but 2 weeks after he started having frequent diarrheas. In 2018 he started taking ocrelizumab. In 2022 he was admitted in our clinic because of back pain, artrhalgia, night pain and heel pain. HLA B 27 was positive and MRI showed acute left sacroiliitis with edema and erosions.

Results: We decided to start biological therapy, so we consulted neurologist and decided to requested approval for IL-17 inhibitor secukinumab which he started taking in November 2023. Secukinumab had excellent effect, BASDAI decreased from 5.3 before starting secukinumab therapy to 2.95 in 6 months of therapy.

Conclusions: The co-existence of MS and AS is rare, it might be due to differences in immunopathogy and immunogenetics. The prevalence of HLA B27 in MS patients is 10.2% which is close to prevalence in healthy population. On the other hand, there are possible common immunopathological pathways in MS and AS linked to Th17 cells.

Keywords: ankylosing spondylitis, Multiple Sclerosis, biological therapy

PV019 / #352

POSTER SESSION 02: SPONDYLOARTHRITIS DISORDERS
03-06-2025 4:50 PM - 5:50 PM

GENETIC INTERACTIONS BETWEEN COMMON HLA B* AND HLA B*27 SUBTYPES IN COLOMBIAN SPONDYLOARTHRITIS AND HEALTH INDIVIDUALS.

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Background and Aims: HLA antigen distribution varies significantly by ethnicity. In Colombia, the main groups are mestizos, Native Americans, and Afro-Colombians. Given the importance of gene interactions, we propose to evaluate the B alleles that frequently accompany HLA B27-positive Spondyloarthitis(SpA) patients by subtype.

Methods: 121 SpA and 742 healthy controls (HC) were included. Luminex and *Illumina NGS* was used to analyze exons. Shannon index (H') was calculated to assess the diversity of subtypes alleles of HLA-B.

Results: 31.4% of patients were HLA-B27 positive, with HLA-B27:05:02 as the most common subtype (78.9%). Genetic diversity was greater in HLA-B27:05:02 (H' = 3.016) than in HLA-B27:02:01 (H' = 1.099). In HC, 29 were HLA-B27 positive (3.9%), including 13 (44.8%) with HLA-B27:05:01 and 37.9% with B27:03:01. A difference was found in the frequency of HLA-B27:05:02 between SpA patients and healthy controls ($P < 0.005$), indicating this subtype is more common in patients. Sequence alignment analysis showed HLA-B27:05:02 and HLA-B*27:02:01 are similar in most regions but have a section of different amino acids. For the interaction between two HLA antigens, the most common B alleles in HLA-B27+ group, show the predominant combinations were HLA-B27:05:02+/HLA-B35:43:01+ and HLA-B27:05:02+/HLA-B40:02:01+ in SpA. In contrast, HLA-B27:05:01 and HLA-B03:01 didn't show any predominant B alleles in healthy subjects

Conclusions: The restriction of HLA-B* subtypes in SpA patients suggests that HLA-B27:05:02 is the ancestral allele, while HLA-B27:05:01 predominates in healthy controls.

Notably, the combinations of HLA-B27 subtypes differ between patients and controls. Additionally, *a stronger association between HLA-B27:*05:02 and HLA-B*35:43:01+ identified indicating a strong genetic interaction. **Acknowledgements ASOREUMA and COLCIENCIAS**

Keywords: HLA antigen, Spondyloarthitis, genetic interaction

PV020 / #548

POSTER SESSION 02: SPONDYLOARTHRITIS DISORDERS
03-06-2025 4:50 PM - 5:50 PM

SAFETY OF THE NOVEL IL-17A INHIBITOR NETAKIMAB ACCORDING TO THE REAL CLINICAL PRACTICE

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Background and Aims: Safety data can vary greatly between randomized clinical trials and a real clinical practice. To analyze the 104-weeks Netakimab (NTK) safety records in r-axSpA in the real clinical practice.

Methods: Study BCD-085-NIS-02/LIBRA was non-interventional and observational. All the adverse events were registered during 104 weeks of NTK treatment.

Results: Patients with definite r-axSpA (n=137) were involved at the study in 23 centers (mean age 42.3(±11.9) y.o.; 68% were male; symptoms duration 13.9±9.2 years; 34.3% had previously received biologics). At least one AE/SAE related to therapy (due to opinion of the investigator), was reported in 8 (6,0%) cases (Table 1). Table 1. Adverse effects observed for NTK in the study.

AE/AR	NTK discontinuation	AE/AR Outcome	Seriousness criteria	Relation of the AE to the NTK therapy
COVID-19	No	Recovered without sequelae	No	Doubtful
Latent tuberculosis infection	No	Improvement	No	Possibly related

AE/AR	NTK discontinuation	AE/AR Outcome	Seriousness criteria	Relation of the AE to the NTK therapy
Alopecia	No	Improvement	No	Possibly related
Terminal ileitis, minimal signs of inflammation	No	Improvement	No	Possibly related
Nasopharyngitis	No	Recovered without sequelae	No	Possibly related
Colitisexacerbation	Yes	Recovered without sequelae	Yes	Doubtful
Acute sinusitis	No	Unknown	No	Doubtful

During the 104 weeks study period, only 1 (0.7%) case fulfilled the seriousness criteria (colitis) was checked. No deaths were reported. Most cases of infections reported for NTK were mild or moderate and did not require antibiotics and /or NTK treatment discontinuation.

Conclusions: Treatment of r-axSpA with NTK was well tolerated – most of registered AEs were expected and previously described for IL-17 inhibitors.

PV021 / #552

POSTER SESSION 02: SPONDYLOARTHRITIS DISORDERS
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SURVIVAL RATE IL-17A INHIBITOR NETAKIMAB IN RADIOGRAPHIC AXIAL SPONDYLOARTHRITIS (R-AXSPA) – REAL WORLD DATA FROM LIBRA STUDY

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Background and Aims: The survival of biologics can vary greatly between trials and clinical practice. Netakimab (NTK) is a novel IL-17A inhibitor for r-axSpA with unknown survival rate. To evaluate the retention rate of NTK in r-axSpA in clinical practice.

Methods: NTK retention rate was analyzed in the study BCD-085-NIS-02/LIBRA in 23 centers. Survival rate was calculated with Kaplan–Meier analysis.

Results: Mean age of 137 r-axSpA patients was 42.3(±11.9) years old; 68% were male, symptoms duration was 13.9±9.2 years; 34.3% had history of biologics. At week 104 85.5% of r-axSpA patients continued NTK therapy (95% CI 79.7-91.8). Patients received biologics previously and bionaïve patients were comparable in NTK retention rate, p=0.16. Survival rate was similar in NTK as the 1st line of biologics or as the second line 75.2% (95% CI 54.2-100.0) and 80.4% (95% CI 67.4-95.8) respectively (p = 0.33) (Fig.1); at week 104 52.9% r-axSpA patients maintain LDA, and 21.3% – remission. The main reasons of NTK discontinuations were loss of follow-up (5.5%) and loss of response (4.4%). Fig.1. Kaplan-Meier curves of NTK survival rate in r-axSpA at week 104

Conclusions: The survival rate in NTK treatment in r-axSpA is satisfactory – most of the patients maintain low disease activity or remission at week 104 without AE or loss of response.

PV022 / #530

POSTER SESSION 02: SPONDYLOARTHRITIS DISORDERS
03-06-2025 4:50 PM - 5:50 PM

THE EFFECTS OF PILATES EXERCISE TRAINING COMBINED WITH WALKING ON CARDIORESPIRATORY FITNESS, FUNCTIONAL CAPACITY AND DISEASE AC-TIVITY IN PATIENTS WITH NON-RADIOLOGICALLY CONFIRMED AXIAL SPONDYLITIS

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Background and Aims: Background and Aims The objective of the study was to examine the effects of pilates exercise training combined with walking on cardiorespiratory fitness, functional capacity and disease activity in patients with non-radiologically confirmed axial spondylitis (nr-axSpA).

Methods: Methods Thirty patients with nr-axSpA [27 women (90%), with a mean age of 46.07±10.48 years old and C-reactive protein (CRP) 2.26±2.14 mg/l] were randomly divided into two groups: A (n_1==15 patients) and B (n_2=15 patients). Group A followed a 6-month home-based pilates exercise training program, while group B remained untrained until the end of the study. CPET, TUG, 5xSTS, SR, BSR and BSL, BASDAI and ASDAS, were applied to all patients, both at the begin and at the end of the study.

Results: Results After 6 months, group A showed higher values in exercise time by 37.41% (p=0.001), VO_2peak by 25.41% (p=0.01, VO_2/HRmax by 14.83% (p=0.04) and SR by 18.70% (p=0.007), while lower values were observed in TUG by 24.32% (p=0.001), 5xSTS by 12.13% (p=0.001), BASDAI by 20.00% (p=0.04) and ASDAS score by 23.41% (p=0.03), compared to group B. Linear regression analysis showed a positive correlation in group A between BASDAI and 5xSTS (r=0.584, p=0.02), BASDAI and TUG (r=0.538, p=0.03), and ASDAS and 5xSTS (r=0.538, p=0.03), while a negative correlation was found between BASDAI and VO_2peak (r=-0.782, p<0.001), ASDAS and SR (r=-0.548, p=0.03) and ASDAS and VO_2peak (r=-0.659, p=0.008).

Conclusions: Conclusions To sum up, cardiorespiratory fitness, functional capacity and disease activity improved after a long-term pilates exercise training program in patients with nr-axSpA..

Keywords: disease activity, spondylarthritis, exercise

PV023 / #170

POSTER SESSION 03: SYSTEMIC SCLEROSIS – CLINICAL ASPECTS AND TREATMENT & PATHOGENESIS
03-06-2025 4:50 PM - 5:50 PM

PREVALENCE, CLINICAL FEATURES, AND MANAGEMENT OF SKIN CALCIFICATIONS IN SCLERODERMA

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Background and Aims: Background and aims: Systemic sclerosis (SSc), commonly known as scleroderma, is a chronic autoimmune condition marked by fibrosis of the skin and internal organs. Calcinosis cutis, or skin calcifications, is a common complication resulting from abnormal calcium deposition in the dermis and subcutaneous tissue. These calcifications can cause significant discomfort, including pain, ulcerations, and infections, adversely affecting patients' quality of life.This study evaluates the prevalence, clinical presentation, and management strategies for skin calcifications in patients with scleroderma.

Methods: Methods: A retrospective analysis was performed on 68 scleroderma patients diagnosed with skin calcifications based on clinical evaluation and imaging. Data on patient demographics, disease duration, extent of skin involvement, and treatments were collected. The effectiveness of therapeutic options, including calcium channel blockers, surgical removal, and extracorporeal shockwave therapy (ESWT), was assessed.

Results: Results: Skin calcifications were present in 35% of patients, primarily those with long-standing disease. Commonly affected areas included the fingers, elbows, and knees. Patients with diffuse cutaneous SSc had a higher likelihood of developing calcifications than those with limited cutaneous SSc. Treatment outcomes varied, with calcium channel blockers showing limited effectiveness, while surgical excision and ESWT provided symptom relief in some cases.

Conclusions: Conclusion: Skin calcifications are a common and disabling feature in scleroderma. Early recognition and individualized treatment are critical for reducing complications. Additional research is required to identify more effective therapies.

Keywords: calcifications, systemic sclerosis, scleroderma

PV024 / #203

POSTER SESSION 03: SYSTEMIC SCLEROSIS – CLINICAL ASPECTS AND TREATMENT & PATHOGENESIS
03-06-2025 4:50 PM - 5:50 PM

MANIFESTATIONS THAT MOST STRONGLY AFFECTS THE QUALITY OF LIFE IN PATIENTS WITH SYSTEMIC SCLEROSIS

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Background and Aims: To evaluate the quality of life in patients with systemic sclerosis(SSc) by using a Health Assessment Questionnaire(HAQ) and parameters that most strongly affects it.

Methods: 151 patients with SSc were included in this study. The mean age was 48±13years, female-83% patients. 81% of patient had interstitial lung disease.

Results: The mean baseline HAQ value was 1.35±0.8 points, indicating that the group had moderate disability on average. 53 patients (35%) had minimal disability, 59(39%) had moderate disability, and 39(26%) had severe disability. Patients were divided into groups depending on HAQ values: group 1 - severe impairments of life activity and group 2 - moderate and minimal impairments of life activity (Table 1). Group 1 had significantly more patients with arthritis, arthralgia, joint contractures, muscle weakness, pain associated with digital ulcer, gastrointestinal dysfunction (dysphagia, early satiety, diarrhea). Also, these patients had lower LVEF, higher PASP, lower FVC and DLCO. There was a direct moderate correlations between HAQ and arthralgia (r=0.432; p=0.01), contractures (r=0.391; p=0.00004), arthritis (r=0.263; p=0.001), pain associated with digital ulcers (r=0.410; p=0.001), dysphagia (r=0.323; p=0.0005), CRP (r=0.502; p=0.0005) and

prednisolone dose (r=0.432; p=0.0007).

Table 1. Characteristics of main parameters of systemic sclerosis depending on the presence of functional disability according to the HAQ questionnaire.

Parameters	Group 1 (n = 39)	Group 2 (n = 112)	p
Female/male, n (%)	36 (92) / 3 (8)	89 (79) /23 (21)	0.01
Disease onset, n (%): - limited - diffuse - overlap	 9 (23) 21 (54) 9 (23)	 47 (42) 59 (53) 6 (5)	 0.001 NS 0.04
Age, years, M±σ	50±13.8	47.1±13.3	0,02
Disease duration, years, M±σ	6.4±4.7	6.4±6.1	NS
Digital ulcers, n (%)	6 (15)	15 (13)	NS
Pain associated with digital ulcers, n (%)	14 (36)	31 (28)	0.03
Arthritis, n (%)	14 (36)	16 (14)	0.00001
Arthralgia, n (%)	24 (62)	43 (38)	0.00001
Contractures, n (%)	28 (72)	69 (62)	0.0001
Muscle weakness, n (%)	8 (21)	14 (13)	0.01
Dysphagia, n (%)	31 (79)	75 (70)	0.01
Early satiety, n (%)	16 (41)	28 (25)	0.0002
Diarrhea, n (%)	9 (23)	16 (14)	0.004
Modified Rodnan skin score, Me [25th; 75th quartile]	9 [3; 22]	9 [3; 15]	0.00002
Left ventricular ejection fraction, %, M±σ	62.5±7.6	64.2±6.5	0.0006
PASP*, mm Hg, M±σ	40.6±18.7	35.1±13.2	0.0001
FVC**, %, M±σ	72.8±17.9	80.2±22.8	0.01
DLCO***, %, M±σ	44.2±14.4	48.7±21.4	0.02
CRP, mg/l, Me [25th; 75th quartile]	8 [3.1; 18.8]	4.9 [1.5; 10.5]	0.00001
Disease activity index, M±σ	3.4±1.7	3.1±1.8	0.01
Prednisolone, mg/day, M±σ	13.1±6.6	11.1±4	0.00001
Immunosuppressants, n (%)	16 (43)	56 (50)	0.03
Cyclophosphamide, n (%)	12 (32)	25 (22)	NS
Mycophenolate mofetil, n (%)	4 (11)	28 (25)	0.0004

*PASP - pulmonary artery systolic pressure, **FVC - forced vital capacity % predicted, ***DLCO - diffusion capacity for carbon monoxide % predicted.

Conclusions: The quality of life of patients with SSc associated with the severity of skin fibrosis, the presence of arthritis and joint contractures, digital ulcers, dyspnea and the dose of prednisolone. The worst values of quality of life were associated with inflammatory activity of the disease, musculoskeletal involvement, gastrointestinal dysfunction, and a decrease in pulmonary functional tests.

Keywords: systemic sclerosis, quality of life, HAQ

PV025 / #212

POSTER SESSION 03: SYSTEMIC SCLEROSIS – CLINICAL ASPECTS AND TREATMENT & PATHOGENESIS
03-06-2025 4:50 PM - 5:50 PM

CLINICAL SIGNIFICANCE OF TNF-ALPHA IN SYSTEMIC SCLERODERMA

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Background and Aims: Objectives: To study the association of serum TNF-alpha levels with the main clinical manifestations and activity of SSc.

Methods: 66 patients with SSc-ILD verified by MSCT were enrolled into the study (disease duration 7.2±5.6 years, diffused/limited SSc 1.7/1, average age 50.3 ± 12.6 years, females 77%). All patients received low or moderate dose glucocorticoids and immunosuppressants, 45 (68%) received rituximab (RTM) at cumulative dose 2.4±1.5 grams. The level of TNF-alpha was determined by the ELISA method. Subsequently correlation analysis was made to clarify the association of the TNF-alpha level with the main clinical manifestations, activity of SSc (EScSG, points), laboratory parameters (ESR, CRP, ANA-HEP-2, a-Scl-70, B cell count).

Results: There was a significant correlation between TNF-alpha levels and the progression of vascular disorders (r=0.43), including digital scars (r=0.27), ulcers (r=0.28) and necrosis (r=0.34), the presence of cardiopathy (r=0.31), proteinuria (r=0.37), and SSc activity (r=0.37). There was no correlation of TNF-alpha levels with indicators of inflammatory and immunological activity and indicators of functional pulmonary tests. However, an inverse correlation of TNF-alpha levels with the development of pulmonary fibrosis was revealed according to MSCT data (r=-0.53).

Conclusions: The serum levels of TNF-α may promote the progression of the endothelial dysfunction. Randomized controlled trials with TNF-α inhibitors in patients with SSc are needed to confirm the potential role of these agents in the treatment of patients with peripheral vascular disorders and without interstitial lung disease.

Keyword: systemic sclerosis, TNF-alfa

PV026 / #213

POSTER SESSION 03: SYSTEMIC SCLEROSIS – CLINICAL ASPECTS AND TREATMENT & PATHOGENESIS
03-06-2025 4:50 PM - 5:50 PM

THE ASSOCIATIONS OF THE MCP-1 LEVEL WITH THE CLINICAL MANIFESTATIONS AND ACTIVITY OF SYSTEMIC SCLEROSIS DURING IMMUNOSUPPRESSIVE AND ANTI-B-CELL THERAPY

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Background and Aims: Objectives: To study the associations of the MCP-1 level with the clinical manifestations and activity of systemic sclerosis during active therapy.

Methods: 66 patients with SSc-ILD verified by MSCT were enrolled into the study (disease duration 7.2±5.6 years, diffused/limited SSc 1.7/1, average age 50.3 ± 12.6 years, females 77%). All patients received low or moderate dose glucocorticoids and immunosuppressants, 45 (68%) received RTM at cumulative dose 2.4±1.5 grams. The level of MCP-1 was determined by the ELISA method. Subsequently correlation analysis was made to clarify the association of the MCP-1 level with the main clinical manifestations, activity of SSc (EScSG, points), laboratory parameters (ESR, CRP, ANA-HEP-2, a-Scl-70, B cell count), glucocorticoid and immunosuppressant therapy and cumulative rituximab dose.

Results: The level of MCP-1 was significantly correlated with the development of arthritis(r=0.29), cardiopathy(r=0.37), diastolic dysfunction of the left ventricle(r=0.26), decreased glomerular filtration rate (r=0.31), modified skin score (r=0.31), SSc activity (r=0.29), levels of CRP (r=0.38), ESR (r=0.36). An inverse correlation was found between the level of MCP-1 and the total dose of rituximab (r= -0.37), but not with the dose of glucocorticoids and the use of immunosuppressants. There was no correlation between the level of MSR-1 and the indicators of functional pulmonary tests and MSCT data.

Conclusions: The data obtained indicate the role of MCP-1 in maintaining the inflammatory activity of SSc,the progression of cutaneous fibrosis and,probably, endothelial dysfunction.A decrease in the level of MCP-1 during rituximab therapy may be a

potential predictor of the effectiveness of this drug, however,this issue requires further study.

PV027 / #214

POSTER SESSION 03: SYSTEMIC SCLEROSIS – CLINICAL ASPECTS AND TREATMENT & PATHOGENESIS
03-06-2025 4:50 PM - 5:50 PM

THE USE OF KL-6 TO PREDICT THE PROGRESSION OF INTERSTITIAL LUNG DISEASE IN SYSTEMIC SCLEROSIS

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Background and Aims: Objectives: To study of the value KL-6 serum level in assessing the progression of SSc-ILD during active therapy.

Methods: 66 patients with SSc and interstitial lung disease(SSc-ILD) verified by MSCT were enrolled into the study (disease duration 7.2±5.6 years,diffused/limited SSc 1.7/1,average age 50.3 ± 12.6 years,females 77%).The duration of follow-up was 19±14.3 months.All patients received low or moderate dose glucocorticoids and immunosuppressants,45 (68%) received rituximab(RTM) at cumulative dose 2.4±1.5 grams.The level of KL-6 was determined by the ELISA method. Subsequently correlation analysis was made to clarify the association of the KL-6 level with the FVC, DCLO,various X-ray symptoms of ILD (ground-glass changes, reticular changes, traction bronchiectasis, and honeycombs), activity of SSc (EScSG, points), laboratory parameters (ESR, CRP, ANA-HEP-2, a-Scl-70, B cell count), glucocorticoid and immunosuppressant therapy and cumulative rituximab dose.

Results: An inverse correlation was found between the level of KL-6 at the time of inclusion in the study and Δ FVC(R=-0.310),but not with the FVC and DLCO.The level of KL-6 was significantly correlated with the B cell count at the beginning of the study(R=0,306).There was no correlation between the level of KL-6 and the X-ray symptoms of ILD, activity of SSc,indicators of laboratory and inflammatory activity,therapy.

Conclusions: The data obtained indicate the possibility of using the KL-6 level to predict the progression of ILD, but not to confirm the presence of a pulmonary lesion.The association of the KL-6 level with the B cells count allows discussing the study of this biomarker to determine the indications for the appointment of anti-B cell therapy.

PV028 / #216

POSTER SESSION 03: SYSTEMIC SCLEROSIS – CLINICAL ASPECTS AND TREATMENT & PATHOGENESIS
03-06-2025 4:50 PM - 5:50 PM

SAFETY OF COMBINATION TREATMENT WITH RITUXIMAB AND MYCOPHENOLATE MOFETIL COMPARED TO RITUXIMAB MONOTHERAPY IN PATIENTS WITH SYSTEMIC SCLEROSIS - A REAL LIFE, RETROSPECTIVE, MONOCENTRIC STUDY

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Background and Aims: Our study aims to compare the safety of combination treatment with mycophenolate and rituximab (MMF-RTX) versus rituximab monotherapy (RTX) in a single-center cohort of patients affected by systemic sclerosis (SSc).

Methods: Adult patients with SSc (ACR/EULAR 2013 criteria) who were treated at our center from 2012 to 2023 with either RTX or MMF-RTX were enrolled. Patients with a follow-up of 12 up to 36 months after the start of therapy were included. During biannual outpatient visits, the following adverse events were recorded: infusion reactions, infections, severe infections (requiring hospitalization), hypogammaglobulinemia (IgG < 4 g/L), or leukopenia (WBC < 4000/mm3).

Results: 15 patients in the RTX group and 17 patients in the MMF-RTX group were included. Patients were followed up until the occurrence of an adverse event or until the end of the observation period, and were similar in terms of sex, disease duration, cutaneous form, and follow-up duration. Cardiac involvement was more frequent in the combination therapy group (p=0.003). During follow-up, two therapy-unrelated deaths occurred in the MMF-RTX group. At the end of follow-up, a total of 15 adverse events were recorded, of which 6 in the RTX group and 9 in the MMF-RTX group (p=0.464). Of the 15 adverse events, 13 were infections (p=1.000), 4 severe (p=1.000); the remaining 2 were hypogammaglobulinemia and an infusion reaction (cutaneous rash), both in the MMF-RTX group. Event-free survival times were similar between the two groups (p=0.420).

Conclusions: Combination therapy with MMF and RTX appears to be well-tolerated and safe in patients with SSc.

Keywords: Rituximab, micofenolato, eventi avversi

PV029 / #261

POSTER SESSION 03: SYSTEMIC SCLEROSIS – CLINICAL ASPECTS AND TREATMENT & PATHOGENESIS
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TH1, TH2 AND TH17 CYTOKINE PROFILE IN PATIENTS WITH MULTIPLE SCLEROSIS FOLLOWING TREATMENT WITH RAPAMYCIN

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Background and Aims: The management of multiple sclerosis (MS) primarily involves immune-modulating medications, with cytokines playing a crucial role in the disease's pathogenesis. Understanding the effects of specific treatments on cytokine levels is vital. This study aims to evaluate the impact of rapamycin on the concentrations of Th1, Th2, and Th17 serum cytokines in patients with MS.

Methods: Six patients with relapsing-remitting MS were enrolled as a case group, alongside six healthy individuals as a control group. Patients received 2 mg of rapamycin daily for 6 months, while the control group received no treatment. Enzyme-linked immunosorbent assay (Simultaneous Multi-Analyte ELISA) was utilized to determine serum concentrations of IL-2, IL-4, IL-5, IL-6, IL-10, IL-12, IL-13, IL-17, IFN-γ, TNF-α, G-CSF, and TGF-β before and after therapy with rapamycin.

Results: The mean absorbance of 10 out of the 12 studied cytokines showed reduction after the therapy with rapamycin including IL-2, IL-4, IL-5, IL-6, IL-10, IL-12, IL-13, IL-17, IFN-γ, and TNF-α. The only statistically significant reduction was observed in the absorbance of IFN-γ ($p=0.028$). Two cytokines illustrated an increase in the patient's sera after the therapy, including G-CSF and TGF-β, but only the increase in TGF-β was statistically significant ($p=0.046$). None of the controlled group's studied cytokines varied significantly after 6 months.

Conclusions: Based on the findings of this study, rapamycin has some immunosuppressive effects, such as decreasing IFN-γ, which can improve the quality of life of patients with multiple sclerosis. Also, the increased level of TGF-β may benefit the disease, which needs further clinical studies.

Keywords: Cytokine Profile, Multiple Sclerosis, Rapamycin

PV030 / #304

POSTER SESSION 03: SYSTEMIC SCLEROSIS – CLINICAL ASPECTS AND TREATMENT & PATHOGENESIS
03-06-2025 4:50 PM - 5:50 PM

COMORBIDITIES IN SYSTEMIC SCLEROSIS-ASSOCIATED INTERSTITIAL LUNG DISEASE. EVALUATION OF THE CHARLSON COMORBIDITY INDEX

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Background and Aims: to estimate the frequency of comorbid conditions in patients with systemic sclerosis associated with interstitial lung disease (SSc-ILD).

Methods: One hundred in-patients with SSc-ILD were included in the study. There were 87 women, mean age – 49.5±12 yrs and disease duration – 8.7±7.2 yrs. The comorbid conditions have been accounted for and Charlson comorbidity index (CCI) was calculated.

Results: The most common comorbidities were hypertension in 25%, coronary heart disease in 25% (myocardial infarction in 6%), osteoarthritis in 27%, osteoporosis in 22% and thyroid disease in 12%. Diabetes mellitus was diagnosed in only 2%, body mass index was 25.2±6.1 (decrease <25 in 17% and obesity in 8%). Neoplasms before the onset of SSc were diagnosed in 2%. Eight patients were smokers. Disability was established in 64% of patients. An increase in CCI > 1 was noted in 96% of patients, >3 in 35%. The average CCI (2.04±1.15) was significantly higher in men, menopausal women, people ≥45 years old, in patients with disease duration > 5 years (p=0.01). Forced vital capacity and diffusing capacity of the lungs for carbon monoxide were significantly lower in patients with CCI≥3 (p=0.001).

Conclusions: Patients with SSc-ILD are often diagnosed with comorbid diseases. CCI depends on gender, age, and duration of the disease. Patients with CCI≥3 had worse pulmonary function measures, suggesting a potential worsening of the SSc-ILD prognosis

associated with increased comorbidity. Therefore, the severity of comorbidity should be taken into account due to planning therapy for SSc-ILD patients.

Keywords: systemic sclerosis, comorbidity

PV031 / #340

POSTER SESSION 03: SYSTEMIC SCLEROSIS – CLINICAL ASPECTS AND TREATMENT & PATHOGENESIS
03-06-2025 4:50 PM - 5:50 PM

LEFT ATRIAL VOLUMETRIC-MECHANICAL COUPLING INDEX AS A MARKER OF SUBLINICAL HEART INVOLVEMENT IN SYSTEMIC SCLEROSIS

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Background and Aims: Background: Primary cardiac involvement in systemic sclerosis (SSc) is common, but clinically silent. Left atrial volumetric-mechanical coupling index (LACI), a new echocardiographic parameter, has been shown to predict cardiovascular outcomes in heart failure patients and the general population, but it has not been studied in SSc. **Objective:** To identify new echocardiographic parameters, particularly related to left atrial volume and function, as markers of increased NTproBNP in SSc patients without overt heart involvement.

Methods: Methods: Consecutive SSc patients without heart involvement underwent 2D and 3D transthoracic echocardiography. LACI was calculated by both 2D and 3D echocardiography Logistic regression, Pearson correlation coefficients, and area under the receiver-operating characteristic curve (AUC) are used to analyze the association and predictive power of LACI for elevated NTproBNP.

Results: Results: A total of 36 SSc patients (55±10 years old; 34 (94%) women; mean disease duration 14±11 years) were included. Seventeen (47%) patients had increased NTproBNP levels (i.e. >125 pg/mL). Among all tested variables, 3D LACI had the best correlation with NTproBNP values ($r = 0.63$, $p<0.001$). A 3D LACI of >2 associated with NTproBNP of >125 pg/mL (odds ratio 7.33 [95%CI 1.38- 38.8]; $p=0.01$) and predicted these increased values with a sensitivity of 76.9% and a specificity of 81.2% (AUC 0.78).

Conclusions: Conclusion: Left atrial volumetric-mechanical coupling index assessed by 3D echocardiography is significantly associated with and predictive of elevated NTproBNP

levels in SSc patients without overt heart disease and may be a potentially useful non-invasive marker for screening for early subclinical heart involvement in SSc patients.

Keywords: systemic sclerosis, heart involvement, Left atrial volumetric-mechanical coupling index

PV032 / #382

POSTER SESSION 03: SYSTEMIC SCLEROSIS – CLINICAL ASPECTS AND TREATMENT & PATHOGENESIS
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MALNUTRITION IS CONNECTED TO GASTROINTESTINAL SYMPTOMS AND DISEASE ACTIVITY IN SYSTEMIC SCLEROSIS

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Background and Aims: Multiple factors may cause changes in nutritional status in systemic sclerosis (SS). Our aim was to investigate the connections between SS activity, malnutrition, and intestinal involvement.

Methods: We conducted a prospective observational study that included adult patients diagnosed with SS according to the ACR/EULAR 2013 classification criteria. To assess digestive symptoms, we used a translated and validated form of the UCLA GIT 2.0 questionnaire. Nutritional status was evaluated using MNA, MUST, and GLIM tools. Disease activity was assessed with EUSTAR-AI (European Scleroderma Trials and Research Group Activity Index).

Results: The final study group consisted of 87 patients, including 46 with a diffuse form and 41 with a limited form of SS. Most were women (85.1%), and the mean disease duration was 9.39 (6.52). Malnourished patients, according to MNA, had higher scores for UCLA (p=0.001), UCLA-reflux (p=0.009), UCLA distension (p=0.033), and UCLA emotional well-being (p=0.000). According to the MUST criteria, patients with a high risk of malnutrition showed clinically significant digestive involvement (UCLA – p= 0.036; UCLA-emotional well-being p=0.002). Active disease (EUSTAR-AI) correlated with MNA assessment (p = 0.027) and MNA malnutrition indicator score (p = 0.023). According to GLIM, malnourished patients had higher EUSTAR-AI scores, but the results did not reach statistical significance (p

=0.1).

Clinical characteristic	Mean ± SD / Number (%)	
Age	56.37 ± 11.57	
mRSS	12.79 ± 8.27	
Inter-incisor distance (cm)	4.31 ± 1.38	
Unintentional weight loss	34 (39.1%)	
BMI (kg/m^2)	< 18.50	7 (8%)
	18.50 – 24.99	46 (52.9%)
	25 – 29.99	25 (28.7%)
	>30	9 (10.3%)
EUSTAR-AI	2.20 ± 1.66	
MNA class	Normal nutritional status	43 (49.4%)
	At risk	34 (39.1%)
	Malnourished	10 (11.5%)
GLIM	Not malnourished	60 (69%)
	Malnourished Stage 1	14 (14.9%)
	Malnourished Stage 2	13 (16.1%)
MUST class	Low risk	60 (69%)
	Medium risk	11 (12.6%)
	High risk	16 (18.4%)
UCLA	0.45 ± 0.41	

Conclusions: In our study group, MNA, MUST, GLIM, and EUSTAR-AI criteria were associated with certain aspects of gastrointestinal involvement in SS.

Keywords: systemic sclerosis, Malnutrition, gastrointestinal

PV033 / #488

POSTER SESSION 03: SYSTEMIC SCLEROSIS – CLINICAL ASPECTS AND TREATMENT & PATHOGENESIS
03-06-2025 4:50 PM - 5:50 PM

SERUM ADAM12 LEVELS INVERSELY CORRELATE WITH LUNG FIBROSIS AND DIGITAL ULCERS IN PATIENTS WITH SYSTEMIC SCLEROSIS

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Background and Aims: The development of dermal and lung fibrosis is an important pathophysiological mechanism in the progression of systemic sclerosis (SSc). Epithelial cell-promoting fibroblast activation is a key element of this underlying process.A recent study showed an increased expression of ADAM12 in fibroblasts of patients with SSc. The aim of the study was to assess the levels of ADAM12 in sera of patients with SSc and to investigate its clinical significance.

Methods: Serum ADAM12 levels were tested by a commercial quantitative sandwich ELISA (R & D Systems) in 50 patients with SSc (25 limited, 25 diffused, 40 females, all ANA positive). Detection of disease-specific or disease-related autoantibodies were tested by a line immunoassay (Euroimmun).

Results: The mean levels ± SD (ng/ml) of ADAM12 in our cohort was 0.138 ± 0.33 ng/ml, range: 0.000-19.19 ng/ml). There was an inverse correlation with the presence of pulmonary fibrosis (0.029 ± 0.081 ng/ml vs 0.162 ± 0.36 ng/ml, p=0.038) and the presence of digital ulcers (0.041 ± 0.083 ng/ml vs 0.202 ± 0.412 ng/ml, p=0.029). There was no correlation between the levels of ADAM12 with sex, the type of the disease, the presence of pulmonary hypertension, GI involvement, arthritis, telangiectasias, serositis, calcinosis, cancer, or the presence of SSc-related autoantibodies (anti-Scl70, anti-centromere, anti-RNA polymerase III and anti-Ro52).

Conclusions: Our study suggests that ADAM12 could be a potential biomarker indicating the absence of pulmonary fibrosis and digital ulcers in patients with SSc, it remains to be seen whether is an epiphenomenon or a marker of the disease.

Keyword: ADAM12, fibrosis, Fibroblasts, Scleroderma, pathophysiology

PV034 / #221

POSTER SESSION 04: PEDIATRIC RHEUMATOLOGY
03-06-2025 4:50 PM - 5:50 PM

CLINICAL FEATURES OF NON-SYSTEMIC JUVENILE IDIOPATHIC ARTHRITIS IN PATIENTS WITH CONGENITAL HEART DISEASE

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Background and Aims: The relationship between congenital heart disease (CHD) and juvenile idiopathic arthritis (JIA) has not been well known. This study aimed to analyze the clinical features of patients with CHD who subsequently developed non-systemic JIA.

Methods: In this single-center, retrospective case series of six patients of JIA with CHD, we analyze clinical features, laboratory data, and treatments collected from the medical record.

Results: Six patients (3 males and 3 females) with JIA were complicated with CHD. Four patients were rheumatoid factor (RF)-positive polyarthritis, one patient was RF-negative polyarthritis, and the rest was oligoarthritis. Four patients required surgical repair of CHD within one year of birth. The median age of onset of JIA was 7 years. Five patients have mild mental retardation (MR), and growth disorder at the time of JIA diagnosis, and two of them were Down syndrome. Interstitial lung disease (ILD) was identified in 3 patients without obvious respiratory symptoms. All of them were RF positive, with elevated KL-6 and ground glass opacity on chest CT scan at the diagnosis of JIA. The arthritis in patients with ILD was particularly refractory, with markedly elevated anti-cyclic citrullinated peptide antibody (mean 2,215U/ml) and matrix metalloproteinase-3 levels (mean 268mg/ml), and all required biologic agents. One of them had multiple joint deformities and contractures. On the other hand, the severity of JIA without complication of ILD was not high.

Conclusions: In JIA with CHD, arthritis in patients with ILD is particularly refractory, and requires prompt and appropriate treatment, with attention to delayed diagnosis due to MR.

Keywords: Juvenile Idiopathic Arthritis , interstitial lung disease, congenital heart disease

PV035 / #293

POSTER SESSION 04: PEDIATRIC RHEUMATOLOGY
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DRY EYE SYNDROME IN PEDIATRIC-ONSET STILL'S DISEASE: A CLINICAL CASE

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Background and Aims: Systemic juvenile idiopathic arthritis (sJIA) is a rare and severe form of juvenile arthritis characterized by systemic inflammation and joint involvement. This case report presents an 8-year-old girl with sJIA who developed severe dry eye syndrome (DES), likely due to underlying autoimmune mechanisms. The aim is to emphasize the importance of early detection and comprehensive management of ocular complications in sJIA.

Methods: The patient presented with high-grade fever lasting over two weeks (peaking at 39°C), inflammatory arthralgia affecting the right knee, talocrural joint, and bilateral radiocarpal joints, along with evanescent rashes. A thorough diagnostic workup was performed, including inflammatory marker profiles, Schirmer's test, and autoimmune antibody screening.

Results: Laboratory results confirmed heightened systemic inflammation, with elevated ferritin (162.6 ng/ml), fibrinogen (5.19 g/l), C-reactive protein (CRP) (109.1 mg/l), and leukocytosis with neutrophilia (66.2%). Protein S100 levels were also elevated (0.37 mcg/l; normal <0.15 mcg/l). Anti-Ro/SSA antibodies were negative, but the presence of anti-NOR90 IgG antibodies suggested an autoimmune component contributing to both systemic and ocular symptoms. Schirmer's test showed severe DES with a result of 1 mm/5 minutes without anesthesia. Following treatment with Tocilizumab, systemic inflammation decreased, and the patient experienced significant improvement in fever, joint pain, and ocular dryness.

Conclusions: This case underscores the need for routine ophthalmologic evaluations in sJIA patients, as ocular involvement like DES can be an under-recognized yet significant complication. The detection of anti-NOR90 IgG antibodies points to an autoimmune

mechanism, and Tocilizumab effectively managed both systemic and ocular symptoms, highlighting its role in addressing autoimmune-driven complications in sJIA.

Keywords: sJIA, dry eye syndrome, anti-NOR90 IgG

PV036 / #331

POSTER SESSION 04: PEDIATRIC RHEUMATOLOGY
03-06-2025 4:50 PM - 5:50 PM

AUTOIMMUNE ENDOCRINOPATHIES IN PEDIATRIC PATIENTS WITH COMMON VARIABLE IMMUNODEFICIENCY RECEIVING IMMUNOGLOBULIN REPLACEMENT THERAPY

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Background and Aims: Common variable immunodeficiency (CVID) is the most frequent symptomatic disorder of antibody biosynthesis, characterized by a marked heterogeneity of genetic underpinnings, immune system dysfunctions, and clinical manifestations associated with infectious manifestations but also with a spectrum of immune dysregulation, autoimmune, allergic, lymphoproliferative, and malignant disorders. In this study, we aimed to evaluate the role of autoimmune complications among children with CVID.

Methods: Retrospective review of the study cohort comprising 39 children, in whom the diagnosis of CVID had been established and the regular immunoglobulin replacement therapy had been implemented. In all the children studied clinical evaluation of autoimmune disorders, autoantibodies, and peripheral blood B and T cell subsets were analyzed.

Results: Anti-thyroid peroxidase antibodies showed a striking prevalence among CVID children, identified in 18 (46.15%) of them, followed by anti-pancreatic islet anti-glutamic acid decarboxylase antibodies in 13 (33.33%) children. Clinically, the most frequent immune dysregulation group was autoimmune disorders, present in 18 (46.15%) of the children studied with a high rate of autoimmune thyroiditis diagnosed in as many as 10 (25.64%) of the CVID children. The most prominent abnormalities in the B and T cell compartment contributing to complex immune deficiency and immune dysregulation, autoimmunity phenotypes, were significant reductions in the switched memory B cell, naive T helper cell, and regulatory T cell subsets.

Conclusions: We document a previously unreported high rate of immune dysregulation and autoimmune disorders in pediatric CVID as a clinical and diagnostic challenge with the variability of defects in the humoral and cellular immune responses.

Keywords: Autoimmunity, Common Variable Immunodeficiency, anti-TPO antibodies

PV037 / #337

POSTER SESSION 04: PEDIATRIC RHEUMATOLOGY
03-06-2025 4:50 PM - 5:50 PM

OCULAR MICROVASCULAR CHANGES IN JUVENILE IDIOPATHIC ARTHRITIS

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Background and Aims: Juvenile idiopathic arthritis (JIA) is a chronic autoimmune disorder in children, often accompanied by ocular complications such as uveitis. Optical coherence tomography angiography (OCTA) is a non-invasive imaging technique that provides detailed visualization of retinal microvascular changes in various ocular conditions. However, its application in detecting retinal microvascular alterations in JIA patients remains underexplored. This study examines the connection between disease activity and retinal microvascular alterations, as assessed by OCTA, in children with JIA.

Methods: In this prospective study, 55 pediatric patients diagnosed with JIA underwent comprehensive rheumatologic and ophthalmologic evaluations, including OCTA imaging. Disease activity was measured using the JADAS-10. Participants were categorized into two groups: mild disease activity (n = 27) and moderate/severe activity (n = 28). OCTA metrics—vascular density of the superficial (SCP) and deep (DCP) capillary plexuses, FAZ, and CMT—were analyzed using an Optopol device (Poland).

Results: The cohort had a mean age of 10.65 ± 4.37 years, with 67.3% female participants. Patients with moderate/severe disease activity exhibited significantly reduced vascular density in the SCP (14.4 ± 4.7% vs. 17.6 ± 3.7%) and DCP (28.3 ± 3.5% vs. 31.85 ± 2.67%) compared to those with mild activity. Additionally, the FAZ area was enlarged (0.38 ± 0.1 mm^2 vs. 0.22 ± 0.04 mm^2), and CMT was increased (286.2 ± 160.2 µm vs. 239.4 ± 29.7 µm) in the moderate/severe group.

Conclusions: OCTA reveals significant retinal microvascular changes in children with JIA, especially in those with higher disease activity, highlighting its potential as a tool for monitoring ocular involvement in JIA.

Keywords: juvenile idiopathic arthritis, OCT-A, disease activity

PV038 / #341

POSTER SESSION 04: PEDIATRIC RHEUMATOLOGY
03-06-2025 4:50 PM - 5:50 PM

S100 PROTEIN AND DISEASE ACTIVITY IN JUVENILE IDIOPATHIC ARTHRITIS

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Background and Aims: Juvenile Idiopathic Arthritis (JIA) is the most common chronic rheumatic disease in children, with diverse clinical presentations and varying disease severity. Identifying reliable biomarkers for assessing disease activity is crucial for optimizing treatment decisions. S100 proteins, particularly S100A8/A9 and S100A12, are involved in JIA inflammation and show promise as disease activity biomarkers. This study aims to evaluate the relationship between serum S100 proteins, interleukin-8 (IL-8), and JIA disease activity, as measured by the JADAS-10.

Methods: A cohort of 31 pediatric JIA patients, diagnosed according to ILAR criteria, was prospectively studied. Serum levels of S100 proteins and IL-8 were measured using ELISA at baseline and follow-up visits. Disease activity was assessed via JADAS10 scores, active joint count, ESR, and CRP levels. Patients were stratified into mild, moderate, and severe disease categories based on JADAS10 scores.

Results: Elevated S100 protein levels (>0.15 mcg/l) were observed in 35.5% of the cohort, with a mean S100 concentration of 0.13 ± 0.08 mcg/l. All patients with elevated S100 levels were ANA-positive, suggesting a link to more severe disease phenotypes. IL-8 levels were within the reference range (<62 pg/ml) in 96.8% of patients, with a mean concentration of 20.27 ± 44.4 pg/ml.

Conclusions: Serum S100 proteins are strongly associated with disease activity in JIA and may serve as valuable biomarkers for monitoring severity, particularly in ANA-positive patients. While IL-8 levels were generally normal, further studies are needed to explore its role in JIA. Incorporating S100 protein measurements into routine practice could improve disease stratification and management.

Keywords: juvenile idiopathic arthritis, S100 proteins, IL-8

PV039 / #342

POSTER SESSION 04: PEDIATRIC RHEUMATOLOGY
03-06-2025 4:50 PM - 5:50 PM

COEXISTENCE OF AUTOIMMUNE THYROIDITIS AND JUVENILE STILL DISEASE

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Background and Aims: Autoimmune thyroiditis, which can manifest in chronic arthritis, involves immune-mediated damage to the thyroid, potentially leading to hypothyroidism or hyperthyroidism. However, data on thyroid involvement in Still's disease is limited, presenting a gap in understanding the interaction between autoimmune and autoinflammatory processes in this condition. This study investigates hormonal and autoimmune thyroid changes in autoinflammatory versus autoimmune arthritis.

Methods: Ninety juvenile idiopathic arthritis (JIA) patients were included, with 45 in the prepubertal group and 45 in the pubertal group. Clinical and laboratory data on thyroid involvement were analyzed. Comparative analysis focused on different subtypes of arthritis onset, including systemic juvenile idiopathic arthritis (sJIA), oligoarticular JIA (oJIA), and polyarticular JIA (pJIA). Statistical methods were employed to assess differences based on age, sex, and disease subtype. The study was approved by the national doctoral school ethics committee.

Results: The analysis revealed no significant difference in the incidence of clinical manifestations based on age distribution (χ^2 = 10.37; DF=5; p=0.06), but a significant difference was observed by sex (χ^2 = 15.008; DF=5; p=0.01). No gender differences were found in thyroid autoantibodies. Notably, children with sJIA showed no thyroid autoantibody positivity, unlike those with oJIA and pJIA. Anti-Tg antibodies showed a significant association with disease duration and treatment response, particularly in relation to age at JIA onset.

Conclusions: Thyroid involvement in pediatric rheumatic conditions should not be overlooked. Our findings suggest that sJIA is an autoinflammatory condition, while other JIA subtypes are autoimmune. Further research is needed to identify children at risk for overlap or polyautoimmune syndromes.

Keywords: Autoimmune thyroiditis, juvenile Still disease, polyautoimmune syndrome

PV040 / #344

POSTER SESSION 04: PEDIATRIC RHEUMATOLOGY
03-06-2025 4:50 PM - 5:50 PM

THE IMPACT OF NUTRITIONAL STATUS ON VITAMIN D LEVELS IN PEDIATRIC RHEUMATOLOGICAL CONDITIONS

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Background and Aims: Chronic rheumatological conditions can significantly affect nutritional status and vitamin D levels in children, which are essential for bone health and disease protection. Vitamin D deficiency, prevalent among youth, negatively impacts bone density and muscle function. This study aims to analyze the relationship between nutritional status and vitamin D levels in children with rheumatological conditions.

Methods: The study was a retrospective and descriptive analysis of children admitted to the Rheumatology Department during 3 months. Data included anthropometric measurements and laboratory analyses evaluating biochemical parameters, including vitamin D levels.

Results: The study included 67 patients, 61.19% of whom were girls. Age distribution showed that 14.93% were under 5 years old, with most aged 6-13 years. Nutritionally, 46.27% were classified as normal weight, while 20.89% were overweight or obese. The most common rheumatological conditions were idiopathic juvenile arthritis (55.22%) and reactive arthropathy (17.91%). Approximately 49% had vitamin D deficiencies or insufficiencies, with a higher prevalence in girls (66.67%). Average BMI was 20.84 kg/m^2, and triponderal index (TPI) was 16.18 kg/m^3. Vitamin D levels showed an inverse correlation with BMI ($r=-0.2$, $r^2=0.04$, $p<0.001$) and a direct correlation with TPI ($r=0.2$, $r^2=0.04$, $p<0.001$), though neither was statistically significant.

Conclusions: The factors influencing vitamin D levels in children with chronic rheumatological conditions are complex. Regular monitoring of vitamin D levels is essential, particularly for girls, who may be more susceptible to deficiencies.

Keywords: Nutritional status, Children, Rheumatological conditions

PV041 / #357

POSTER SESSION 04: PEDIATRIC RHEUMATOLOGY
03-06-2025 4:50 PM - 5:50 PM

NAILFOLD CAPILLAROSCOPY IN JUVENILE ONSET AUTOIMMUNE DISORDERS WITH PUFFY HANDS BUT WITHOUT RAYNAUD´S PHENOMENON

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Background and Aims: Noninvasive screening and disease monitoring are an unmet need in pediatric autoimmune disease. Nailfold video capillaroscopy (NFC) is a validated technique for microvascular surveillance in rheumatologic diseases and should be considered as an early screening tool for the detection of microangiopathy in young patients with unclear autoimmune disorders. We aimed to identify variations in NFC in pediatric patients with puffy hands without Raynaud´s phenomenon and the association with disease activity.

Methods: We present five young patients (3 boys and 2 girls), with puffy hands onset between 5-13 years. NFC was performed using video capillaroscopy equipped with a 200x magnification contact lens (Video 3.0) at 8 digits in a quiet, temperature-controlled room (22–24 °C) according to expert recommendations.

Results: They suffered persistent and progressive polyarthralgia, myalgia, swelling and stiffness of the bilateral fingers, hands, ankles. The Raynaud's phenomenon was absent at that moment. Inflammatory markers were only mildly elevated. Serologic analysis included an elevated anti-nuclear antibody with negative result for other extractable nuclear antigen and other relevant antibodies. The nailfold capillaroscopy performed at diagnosis was abnormal and confirmed an 'active' scleroderma pattern. These aspects guided and influenced the treatment with good resolution of many symptoms after 6 months of therapy. Significant amelioration of capillaroscopic changes in young patients as response to the treatment has been reported.

Conclusions: We think that nailfold video capillaroscopy should be included as a screen for early assessment of all young patients with puffy hands and other nonspecific

symptoms suggestive for autoimmune disease, and as part of routine follow-up for these subjects.

PV042 / #415

POSTER SESSION 04: PEDIATRIC RHEUMATOLOGY
03-06-2025 4:50 PM - 5:50 PM

COMPOUND HETEROZYGOUS UNC13D MUTATION IN A PATIENT WITH JUVENILE IDIOPATHIC ARTHRITIS

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Background and Aims: Juvenile idiopathic arthritis (JIA) is an autoimmune/autoinflammatory joint disease that frequently occurs along with other inflammatory or haematological diseases. Despite the identification of numerous gene variants through genome-wide association studies, their role in the pathogenesis of JIA and of associated diseases remains unclear. Potentially causal JIA genes participate in pathways such as antigen presentation, cytokine signalling or their regulatory mechanisms.

Methods: We present a 28-yr adult female patient diagnosed with polyarticular JIA at the age of ten, who was found to be compound heterozygous for UNC13D variants of unknown significance (VUS).

Results: She presented with symmetric arthritis. Radiographs of the hands and feet showed juxtaarticular osteoporosis and a left tibio-tarsal joint erosion. She had mild inflammation (ESR 24 mm/h, CRP 0.8 mg/dl, normal<0.5) and rheumatoid factor was positive (32IU, normal <8), while anti-nuclear and anti-CCP antibodies were negative. She is currently well on baricitinib. Her brother carrying the same VUS had a prolonged post-Covid reactive arthritis. During a cautious follow-up, none of them had signs of macrophage activation syndrome (MAS).

Conclusions: UNC13D (Munc13-4) has been described as one of the most important genes in lymphohistiocytosis. Nevertheless, JIA has been reported in a patient with systemic JIA, without MAS, who was compound heterozygous for UNC13D and had defective NK cytotoxic function. As cytotoxic deficiency cannot be ruled out in our patient, JAK inhibitors may reduce the risk of MAS. JIA phenotypes may be influenced by different

gene variants. Genetic testing could pave the way for improved patient stratification and shared therapeutic targets.

Keywords: juvenile idiopathic arthritis, UNC13D, lymphohistiocytosis

PV043 / #431

POSTER SESSION 04: PEDIATRIC RHEUMATOLOGY
03-06-2025 4:50 PM - 5:50 PM

OCULAR SARCOIDOSIS IN PEDIATRIC PATIENT: A CASE REPORT

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Background and Aims: Sarcoidosis is a systemic granulomatous disease that rarely presents in the pediatric population.[1, 2] The eye remains as a common site for presenting symptoms in up to 40% of the patients diagnosed with sarcoidosis. [3] The diagnosis is based on three major criteria: a compatible clinical presentation, finding non-necrotizing granulomatous inflammation in one or more tissue samples, and the exclusion of alternative causes of granulomatous disease.

Methods: Case Report.

Results: A nine-year-old male initially presented with a history of painless, bilateral visual acuity decline. Fundoscopic examination revealed findings consistent with posterior uveitis and bilateral vitritis, with a predominance in the left eye. Although a biopsy was not performed, a HRCT scan revealed pulmonary lesions, consistent with a diagnosis of systemic sarcoidosis involving the eyes and the lungs. Other diseases were ruled out (PPD negative, ANCA negative, Lyme is not relevant in Paraguay). The patient was initially treated with prednisone at a dosage of 30 mg, methotrexate (25mg subcutaneously weekly) for 7 months, resulting in clinical improvement, which further allowed to confirm a diagnosis of sarcoidosis. A transition to adalimumab was planned; however, due to financial constraints, the initiation of adalimumab was delayed to . Despite this, the patient continued to show improvement during the interim period, with lungs clear of nodules by CT scan.

Conclusions: This case demonstrates the importance of ruling out sarcoidosis in young patients presenting with posterior uveitis. The prompt treatment with adalimumab is ideal,

however, prednisone and methotrexate demonstrated to be good enough given the economic situation.

Keywords: Sarcoidosis, pediatric, ocular

PV044 / #436

POSTER SESSION 04: PEDIATRIC RHEUMATOLOGY
03-06-2025 4:50 PM - 5:50 PM

KLOTHO PROTEIN IN PATIENTS WITH JIA

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Background and Aims: Klotho (KL) is a protein that seems to play a role in various childhood disorders. We have studied bone metabolism in children with juvenile idiopathic arthritis (JIA), including the Klotho.

Methods: The study included 72 patients with JIA aged 4-18 years. The control group consisted of 29 healthy children matched for age. In both groups, parameters of bone metabolism in blood serum were examined, including Klotho protein. Klotho detection was performed using a commercially available enzyme-linked immunosorbent assay (ELISA) kit. Bioethical committee no: KE-0254/112/2017 (funding: DS 410 Medical University of Lublin).

Results: In the JIA group, the mean KL concentrations were: 1716.7 ± 975.7 pg/ml, while in the control group: 2337.9 ± 966.7 pg/ml. Comparison of KL values showed significantly lower values in the JIA group compared to the control ($p = 0.003$). The KL value was compared in the JIA group depending on the course of the disease: oligoarthritis, polyarthritis and enthesitis. No significant differences were found between these groups. The differences in KL values between the JIA group treated with glucocorticosteroids and the untreated were also not statistically significant. No significant correlations of KL with disease activity parameters or bone metabolism parameters, including vitamin D were found in the JIA group.

Conclusions: It has been determined that KL deficiency may cause mineral metabolism disorders, impaired growth, calcification of blood vessels and soft tissues. Studies also indicate chondroprotective effects of KL. Therefore, reduced concentration in JIA may indirectly indicate bone metabolism disorders and other tissues in children with JIA.

Keyword: JIA, Klotho

PV045 / #130

POSTER SESSION 05: AUTOIMMUNE DISEASE MANAGEMENT
03-07-2025 2:10 PM - 3:10 PM

IMUNOMODULATORY EFFECTS OF YU-PING-FENG FORMULA ON PRIMARY SJO_GREN'S SYNDROME: INTERROGATING THE T CELL RESPONSE

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Background and Aims: Ethnopharmacological treatments have shown beneficial effects in the clinical practice of autoimmune disorders. However, the underlying mechanism of immunomodulatory effects remains challenging, given the complicate composition of herbal medicines. Here, we developed an immunological approach to interrogate the T helper cell response.

Methods: Through data mining, we hypothesized that Chinese medicine formula Yu-Ping-Feng might be a promising candidate for treating primary Sjögren syndrome, a common autoimmune disease manifested by exocrine gland dysfunction. Mice with active Sjögren syndrome were treated with Yu-Ping-Feng formula for a specified duration to evaluate its impact on disease progression. Salivary function, serum autoantibody levels, and immune cell populations were assessed post-treatment. Coculture assays and adoptive transfer models were employed to investigate the direct effects of Yu-Ping-Feng on different immune cells. We also recruited 20 patients with primary Sjögren syndrome and conducted a pilot study of 8-weeks therapy of Yu-Ping-Feng formula.

Results: Yu-Ping-Feng therapy ameliorated the experimental Sjögren syndrome pathology in mice, showing improved salivary function and decreased serum levels of autoantibodies. Both effector T and B cells were significantly suppressed. Further analyses using coculture assays and adoptive transfer models demonstrated direct inhibition by Yu-Ping-Feng on the expansion and differentiation of effector/memory T-cells relevant to Sjögren syndrome. Yu-Ping-Feng treatment also effectively improved fatigue symptoms and exocrine gland functions, as well as reduced serum IgG/IgA levels, while effector T- and B-cell subsets were significantly decreased in patients.

Conclusions: Our findings suggest a novel approach to assess the immunomodulatory effects of Yu-Ping-Feng formula, which may be favorable for patients with autoimmune disorders.

Keywords: adoptive transfer, Chinese medicine formula, Sjögren syndrome

PV046 / #131

POSTER SESSION 05: AUTOIMMUNE DISEASE MANAGEMENT
03-07-2025 2:10 PM - 3:10 PM

GINSENG-EPIMEDII FORMULA AMELIORATED EXPERIMENTAL SJÖGREN'S SYNDROME VIA REDUCING IL-6 PRODUCTION

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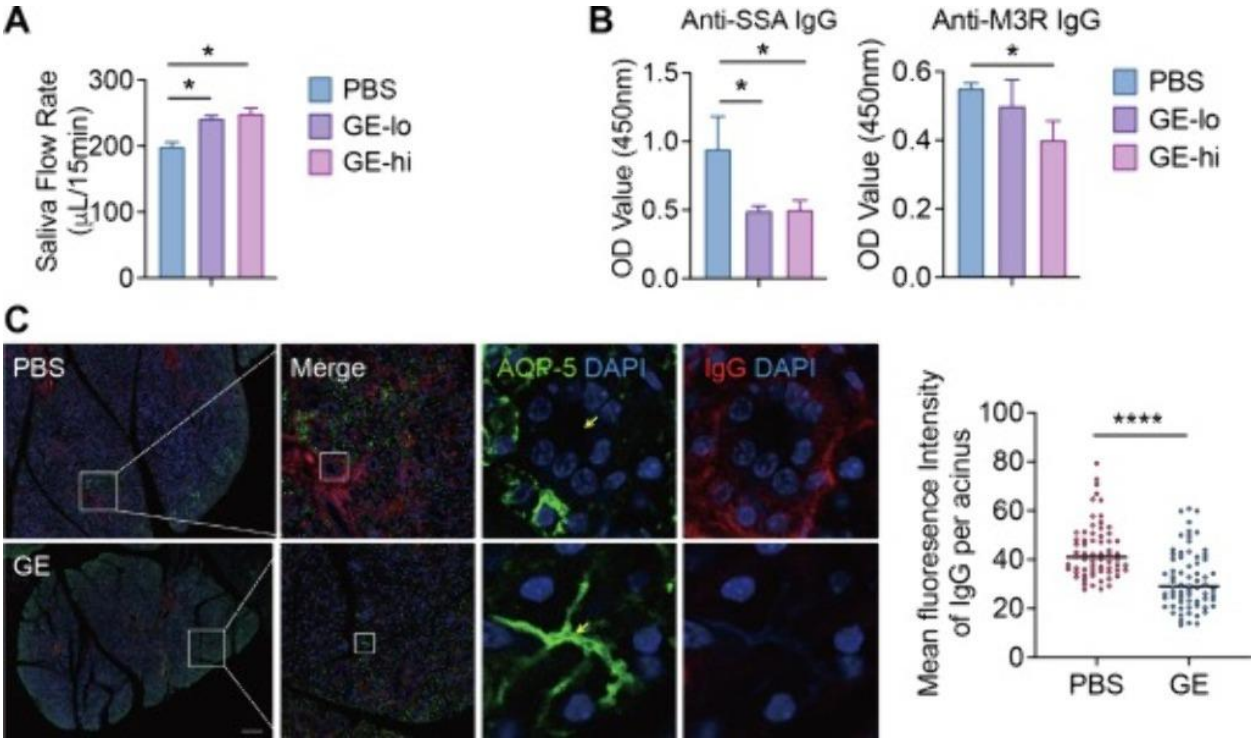
Background and Aims: Primary Sjögren's syndrome (pSS) is an autoimmune disease marked by exocrine gland dysfunction and systemic immune dysregulation. Current therapies, such as B cell depletion, offer limited efficacy in restoring glandular function. This study examines the therapeutic effects of the Ginseng-Epimedii (GE) formula, a traditional herbal remedy, on experimental Sjögren's syndrome (ESS), with a focus on its modulation of interleukin-6 (IL-6) and effector T cell responses.

Methods: ESS was induced in C57BL/6N mice via immunization with salivary gland proteins. Mice received oral GE treatment for 14 days. Salivary gland function was assessed through pilocarpine-stimulated saliva collection, and serum autoantibodies were quantified by ELISA. Flow cytometry was used to measure IL-6 production by antigen-presenting cells and the differentiation of Th17 and T follicular helper (Tfh) cells. Immunofluorescence was performed to assess IgG deposition and AQP5 expression in the salivary gland. A network pharmacology analysis was conducted to explore GE's active components and their target pathways in pSS.

Results: GE treatment significantly improved salivary function and reduced anti-SSA autoantibodies. IL-6 production by B cells and dendritic cells was markedly suppressed, along with reduced Th17 and Tfh cell differentiation. These results indicate that GE modulates the immune microenvironment, limiting effector T cell activation and autoantibody
production.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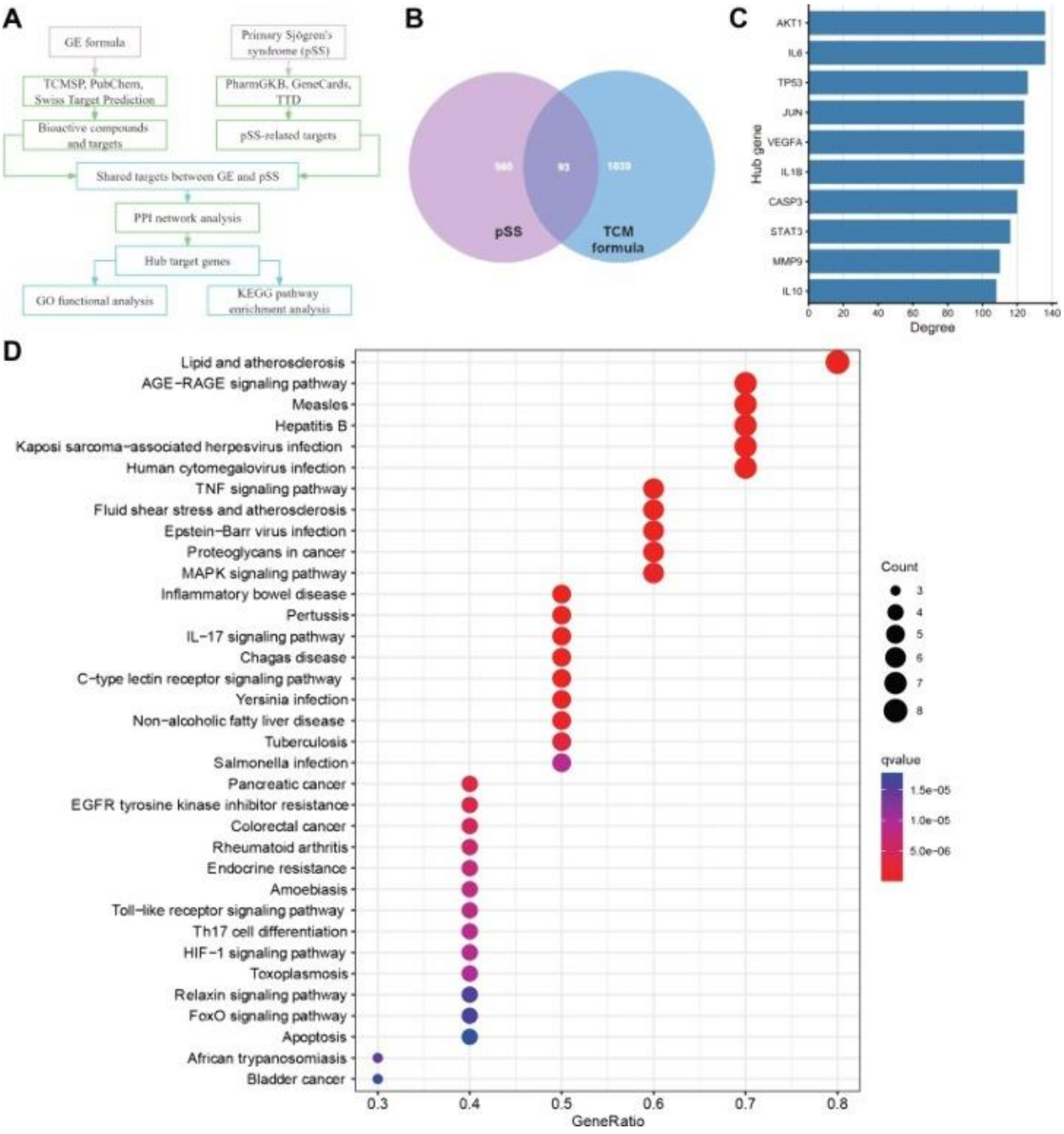
A
GE formula
Primary Sjögren's syndrome (pSS)
TCMSP, PubChem, Swiss Target Prediction
PharmGKB, GeneCards, TTD
Bioactive compounds and targets
pSS-related targets
Shared targets between GE and pSS
PPI network analysis
Hub target genes
GO functional analysis
KEGG pathway enrichment analysis
B
pSS
TCM formula
C
Hub gene
AKT1
IL6
TP53
JUN
VEGFA
IL1B
CASP3
STAT3
MMP9
IL10
Degree
D
Lipid and atherosclerosis
AGE−RAGE signaling pathway
Measles
Hepatitis B
Kaposi sarcoma−associated herpesvirus infection
Human cytomegalovirus infection
TNF signaling pathway
Fluid shear stress and atherosclerosis
Epstein−Barr virus infection
Proteoglycans in cancer
MAPK signaling pathway
Inflammatory bowel disease
Pertussis
IL−17 signaling pathway
Chagas disease
C−type lectin receptor signaling pathway
Yersinia infection
Non−alcoholic fatty liver disease
Tuberculosis
Salmonella infection
Pancreatic cancer
EGFR tyrosine kinase inhibitor resistance
Colorectal cancer
Rheumatoid arthritis
Endocrine resistance
Amoebiasis
Toll−like receptor signaling pathway
Th17 cell differentiation
HIF−1 signaling pathway
Toxoplasmosis
Relaxin signaling pathway
FoxO signaling pathway
Apoptosis
African trypanosomiasis
Bladder cancer
GeneRatio
Count
qvalue
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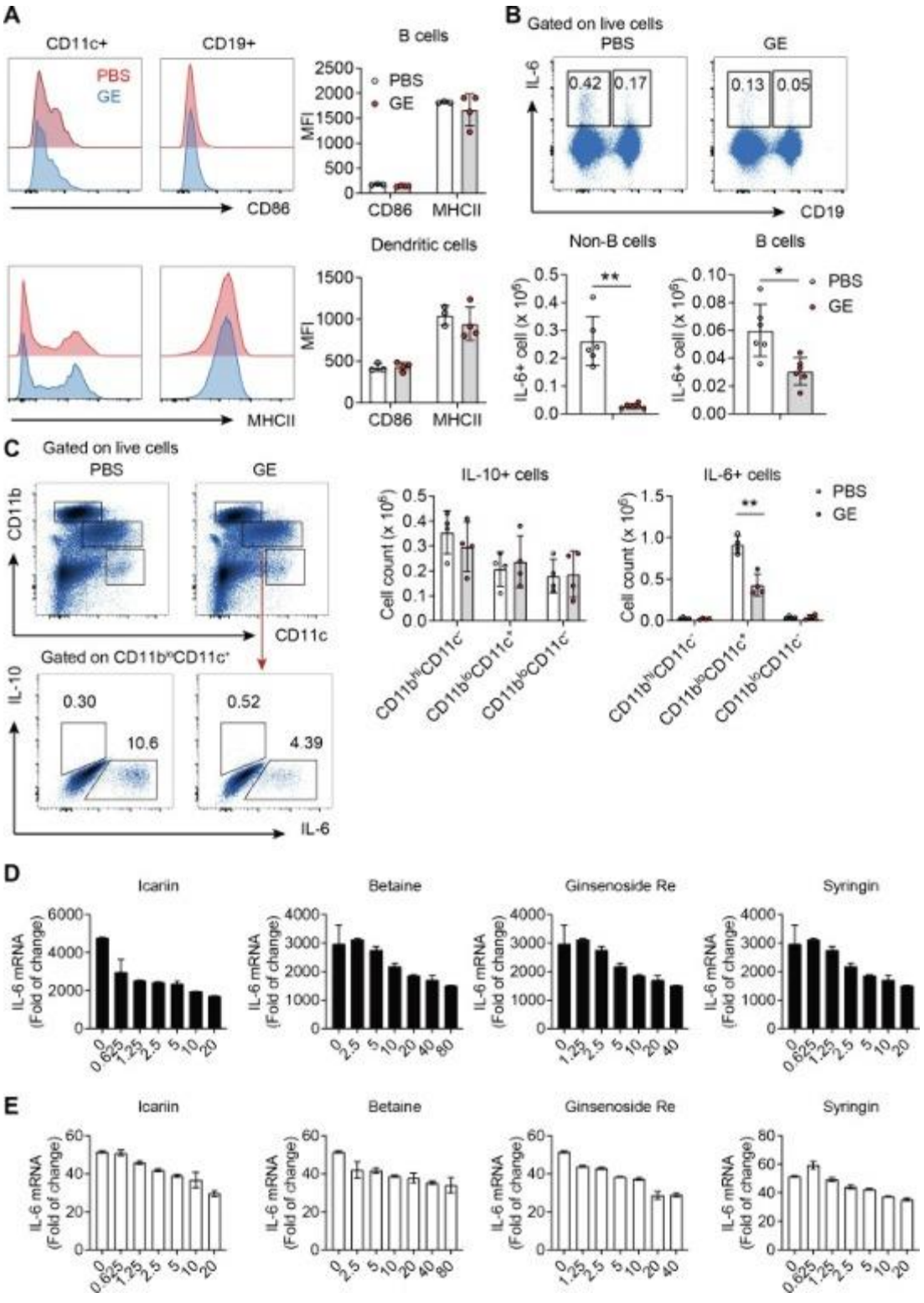
1.0e−05
5.0e−06



Conclusions: GE formula shows significant immunomodulatory effects in ESS by reducing IL-6 production and restraining effector T cell responses, highlighting its potential as a novel therapeutic candidate for treating pSS and related autoimmune disorders.

Keywords: Autoimmunity, Sjogren's syndrome, Chinese medicine

PV047 / #149

POSTER SESSION 05: AUTOIMMUNE DISEASE MANAGEMENT
03-07-2025 2:10 PM - 3:10 PM

NAVIGATING THE DIAGNOSTIC MAZE AND TREATMENT DIFFICULTIES IN REFRACTORY MULTICENTRIC RETICULOHISTIOCYTOSIS

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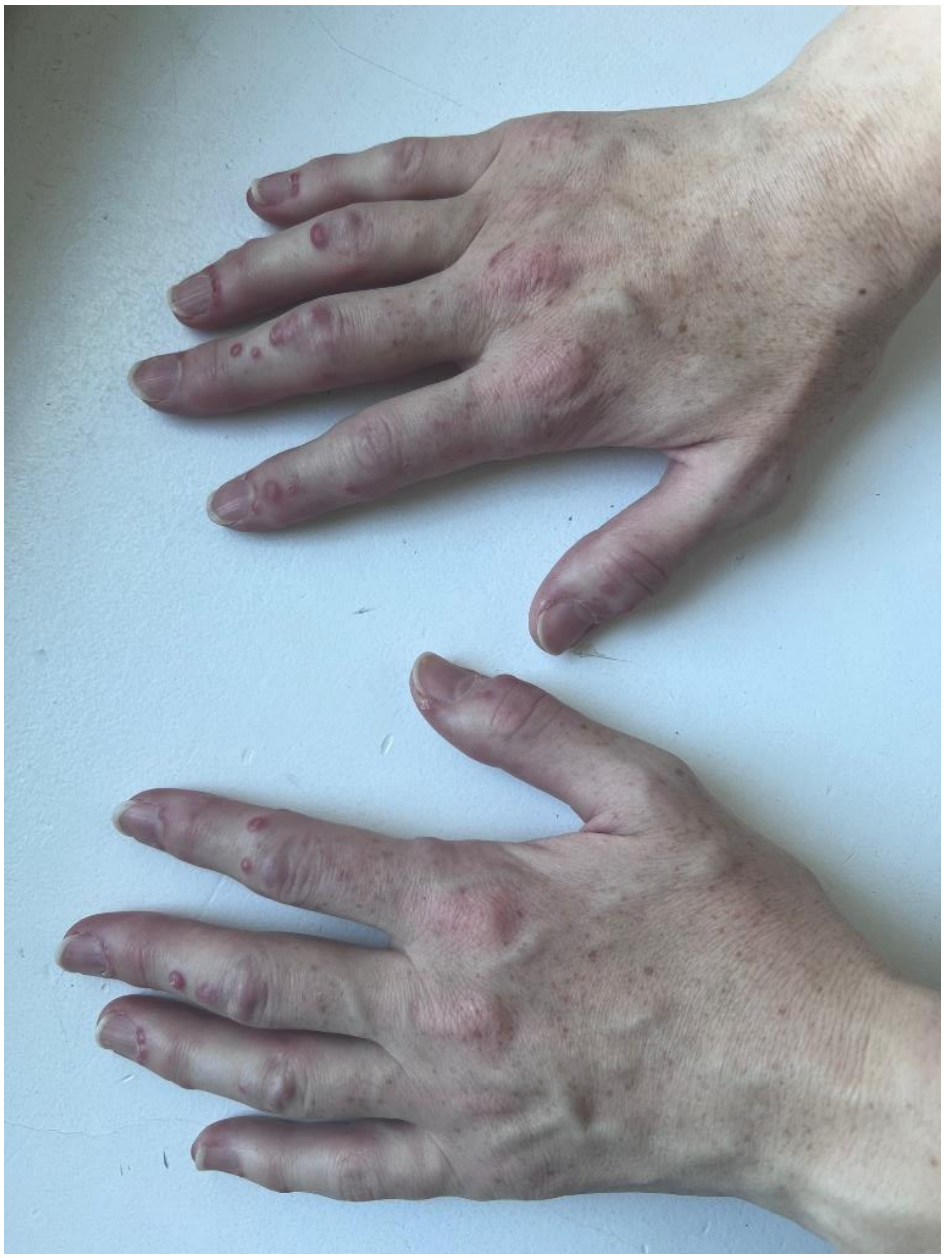
Background and Aims: Multicentric reticulohistiocytosis (MRH) is a rare systemic granulomatous disorder of unknown etiology, characterized by papulonodular skin lesions and destructive arthritis.With fewer than 300 cases reported in the literature, its true prevalence remains unclear. We present a particularly challenging case of a patient with refractory MRH, highlighting the complexities in diagnosis and management.

Methods: A case description of MRH .

Results: A 35-year-old female presenting with arthritis affecting the knees and small joints of her hands, accompanied by papulonodular rashes on the fingers **(Image 1),** was admitted to Vilnius University Hospital, Santaros Klinikos, Department of Rheumatology in September 2023. Examination revealed positive anti-Ro-52 antibodies, atypical findings in videocapillaroscopy and sonoscopic evaluation showed active synovitis. Synovial fluid analysis suggested lymphocitic inflammation. A thorough investigation ruled out systemic connective tissue diseases, paraneoplastic syndrome and potential infectious causes. Although the skin biopsy was suggestive of sarcoidosis, further testing excluded this condition, leading to a diagnosis of MRH. Initial therapies with NSAIDs, corticosteroids, hydroxychloroquine, methotrexate were inadequate as follow-up sonoscopic evaluations revealed the development of joint erosions. Consequently a combination therapy of adalimumab and methotrexate was initiated but after three months, there was no

noticeable improvement. **Image 1. Typical to MRH skin rash.**



Conclusions: This case highlights the challenges in managing MRH. Due to the rarity of this disease, the treatment is largely empirical. Literature suggests that combining TNF-alpha inhibitors with methotrexate yields the best results in controlling arthritis. Other therapeutic options include bisphophonates, other synthetic disease-modifying drugs, and biologic agents such as tocilizumab, anakinra and JAK inhibitors.

Keywords: arthritis, multicentric reticulohistiocytosis, TNF-alpha inhibitors

PV048 / #225

POSTER SESSION 05: AUTOIMMUNE DISEASE MANAGEMENT
03-07-2025 2:10 PM - 3:10 PM

EVALUATION OF GENES INVOLVED WITH METABOLISM IN PATIENTS WITH SJOGREN"S DISEASE

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Background and Aims: The aim of the current study was to evaluate mutations in genes affecting metabolism in a population of patients with Sjogren's disease.

Methods: Whole exome sequencing and sequencing of mitochondrial DNA was performed on 194 patients meeting clinical criteria for Sjogren's disease with autoantibodies associated with Sjogren's disease (SSA, SSB, anti-SP1, anti-CA6, anti-PSP)

Results: Of 194 patients (Age 21-81; mean age 54.2 +/- 13.5 years) evaluated, significant mutations were identified in 192. Many patients had mutations in several genes. The most common mutations were mitochondrial respiratory chain genes (complex 1 – 76, complex 3 – 44, complex 4 – 19, complex 5 – 49). Other mutations affecting the mitochondrial were MT-DLOOP mutations (10), mutations in M-tRNA genes (17), genes affecting carnitine (CPT2 – 4 ; SLC22A5 – 2), genes causing mitochondrial depletion (MCME1 – 1, RRMP8 – 1, POLG – 3, TK – 1), gene causing CoQ10 deficiency (PDSS2 – 1) and succinate dehydrogenase deficiency (1). Glycogen storage diseases were identified in 7 patients (Type II – 1, Type IIIa – 1, Type V – 1, Type IX – 1 and Type XI – 3). Treatment of these disorders with appropriate diet and medications resulted in significant improvement in fatigue, exercise intolerance, gastrointestinal dysmotility, joint pain and recurrent infections. Interestingly, 53 patients had various food hypersensitivities – appropriate elimination diets contributed to improved energy and gastrointestinal function.

Conclusions: Metabolic disorders are common in patients with Sjogren's disease. Treatment of the underlying metabolic disorders benefits patients symptomatically.

PV049 / #226

POSTER SESSION 05: AUTOIMMUNE DISEASE MANAGEMENT
03-07-2025 2:10 PM - 3:10 PM

INHIBITION OF GLYCOLYSIS PREVENTS EARLY MANIFESTATIONS OF SJOGREN'S DISEASE IN AN ANIMAL MODEL

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Background and Aims: Metabolic abnormalities are associated with multiple autoimmune diseases. We described metabolic abnormalities in the IL14a transgenic mouse model (TG) for Sjogren's disease (SD) (Clin. Exp. Rheum. 36: 301, 2018). The aim of the current study was to determine affects of blocking glycolysis on manifestations of SD in TG mice at different stages of disease.

Methods: Groups of 6 TG mice were treated with either deoxy-glucose (DG), metformin (M) or rapamycin (RP) from either 6-7 months of age or 10-14 months of age. The presence of eye disease was determined by observation, the presence of salivary gland disease determined by measurement of salivary gland secretions after pilocarpine stimulation and the level of immunoglobulins in serum by ELISA .

Results: Mice treated with DG starting from 6 -7 months of age showed normalization of salivary gland secretions, eye disease and hypergammaglobulinemia. Mice treated with RP had modest improvement in salivary gland secretions, no change in eye disease, reduced IgM, but no changes in IgG. In contrast, M treatment did not improve any of these clinical manifestations. DG completely blocks glycolysis while RP reduces the reliance on glycolysis by blocking mTOR. In total contrast, TG mice treated with DG, M or RP from 10-14 months of age exhibited no significant changes in disease manifestations.

Conclusions: Early manifestations of SD in TG mice rely on glycolysis. Later manifestations of SD in TG mice do not rely on either glycolysis or mitochondrial respiratory chain function. Metabolism differs at different stages of SD.

PV050 / #227

POSTER SESSION 05: AUTOIMMUNE DISEASE MANAGEMENT
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AUTOANTIBODIES IN A COHORT OF PATIENTS WITH CLINICALLY DEFINED SJOGREN'S DISEASE

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Background and Aims: Many patients with Sjogren's disease lack SSA. Antibodies to salivary gland protein 1 (SP1), carbonic anhydrase 6 (CA6) and parotid secretory protein (PSP) are seen in patients with Sjogren's Disease (Clin. Immunol. 182: 24, 2017; Cornea 4: 405, 2018; Clin. Exp. Rheum. 40: 2387, 2022). These studies evaluated antibodies to SP1, CA6 and /or PSP in a cohort of patients with clinically defined Sjogren's syndrome.

Methods: Charts were reviewed of 194 patients with xerostomia, xerophthalmia, positive Schirmer's tests, fatigue, arthralgias and often gastrointestinal dysmotility ((Age 21-81; mean age 54.2 +/- 13.5 years; 11% Male) who had autoantibody testing done as part of routine care.

Results: All patients had autoantibodies to SSA, SSB, SP1, CA6 and / or PSP. Only 7 patients had antibodies to SSA and 1 to SSB. Ninety patients had antibodies to SP1, 102 to CA6 and 49 to PSP. Ninety-seven patients had autoantibodies to only one autoantigens: 35 to SP1(19 IgM), 49 to CA6 (33 IgG) and 13 to PSP. Food hypersensitivities were identified in 53 patients, 40 of whom had antibodies to CA6.

Conclusions: Many patients with clinically defined Sjogren's disease lack antibodies to SSA, but have antibodies to SP1, CA6 and / or PSP. Many patients with Sjogren's disease and food hypersensitivities have antibodies to CA6. Further study is needed to evaluate evolution of autoantibodies in patients with Sjogren's disease over time as well as to determine whether a particular autoantibody correlates with or predicts a particular clinical manifestation.

Keyword: Sjogren's, autoantibodies

PV051 / #259

POSTER SESSION 05: AUTOIMMUNE DISEASE MANAGEMENT
03-07-2025 2:10 PM - 3:10 PM

FROM DRY EYES TO NEUROLOGICAL SURPRISES: A UNIQUE SJÖGREN'S SYNDROME CASE

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Background and Aims: Primary Sjögren's syndrome (PSS) is a chronic autoimmune disorder marked by xerostomia and xerophthalmia, alongside various neurological manifestations. While involvement of peripheral nervous system (PNS) is common, involvement of the central nervous system (CNS) is rare. This report highlights an unusual case of autoimmune CNS vasculitis and myeloradiculitis as the initial manifestation of PSS.

Methods: We present a case study of a PSS associated with myeloradiculitis and CNS vasculitis.

Results: A 74-year-old female was admitted to the Neurology department with a four-month history of progressive, asymmetric lower limb weakness and hypoesthesia in the perineal area and legs. Neurological examination revealed lower limb asymetric paraparesis with bowel and bladder dysfunction (ASIA C). The patient was thoroughly assesed for infections and paraneoplastic syndrome. Spine MRI indicated thoracic myelopathy at Th3-Th4, leptomeningeal enhancement in the conus medullaris, and root enhancement in the cauda equina. Brain MRI revealed ischemic changes characteristic of small vessel vasculitis. After consulting with a rheumatologist, PSS was suspected, as the patient had lymphopenia and, elevated inflammatory markers, and a history of dry eyes and mouth, positive Anti-Ro/SSA and borderline for Anti-Ro-52. The biopsy of small salivary glands confirmed the diagnosis of PSS based on the 2016 ACR-EULAR classification criteria. The patient was treated with IV methylprednisolon, plasmapheresis and cyclophosphamide. Although her condition stabilized, neurologic deficits persist.

Conclusions: Early diagnosis and proactive treatment of PSS are essential for effective management and positive long-term outcomes, though treatment for CNS involvement remains poorly defined.

Keywords: Primary Sjögren syndrome (PSS), autoimmune myeloradiculitis, CNS vasculitis

PV052 / #276

POSTER SESSION 05: AUTOIMMUNE DISEASE MANAGEMENT
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IGG4 RELATED DISEASE: UNCOMMON FEATURES OF A RARE DISEASE

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Background and Aims: We present a case series of three patients with IgG4-related disease (IgG4RD) exhibiting atypical presentations that do not meet established classification criteria, underlining the need of broader ones.

Methods: A 30 years-old man presented with unexplained hyperemesis, ascites, and patchy gastrointestinal thickening on CT scans. Ascitic fluid analysis revealed elevated IgG4 levels. Biopsies from colonoscopy and gastroscopy showed no neoplasm but indicated mild eosinophil and plasma cell infiltration. While eosinophil counts were insufficient for hypereosinophilia, a significant number of plasma cells were IgG4-positive. He was diagnosed with IgG4-related disease (IgG4-RD) and successfully treated with intravenous steroids and rituximab, leading to symptom resolution.

Results: A 35 years-old woman experienced asthenia, loss of appetite, diffuse itching, jaundice, and acholic stools. Blood tests revealed hemolytic anemia, and a CT scan identified a peri-hilar bile duct mass infiltrating the hepatic artery and portal vein. She underwent gastroscopy, echo-endoscopy, and ERCP, which confirmed the mass and allowed for stent placement. Biopsy showed an IgG4-positive plasma cell infiltrate with fibrosis and eosinophils. She was successfully treated with rituximab and prednisone.

Conclusions: The third patient is a 64-year-old woman who underwent a sialadenectomy for a suspicious mass-forming lesion in the submandibular gland. No neoplastic cells were found in the nearby lymph nodes or the mass itself; however, the pathology report revealed a rich IgG4-positive plasma cell infiltrate. No other organ involvement was clinically evident, but a PET-CT scan showed hypermetabolism in the right colon. A diagnosis of localized IgG4-related disease (IgG4-RD) was made, and she was successfully treated with steroids.

Keywords: Classification criteria, Rare feature

PV053 / #283

POSTER SESSION 05: AUTOIMMUNE DISEASE MANAGEMENT
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DEVELOPMENT OF PHYTOBIOACTIVE COMPOUNDS BASED NANO DRUG DELIVERY SYSTEM FOR IMMUNOMODULATORY EFFECT: FORMULATION OPTIMIZATION, IN VITRO AND IN SILICO STUDIES

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Background and Aims: Most synthetic immunomodulatory drugs are prohibitively costly, possess numerous drawbacks, and are associated with various adverse effects. So that, introduction of immunomodulatory drugs derived from natural sources is regarded as a potential and appealing alternative. Therefore, this study aims to develop a phytobioactive compound based nanoformulation with enhanced immunomodulatory activity via molecular docking and *in vitro* assay of selected phytobioactive compounds.

Methods: Molecular docking simulations were performed on the ligand binding regions of TNF-α, IL-6, and IL-1β, utilizing selected phytomedicines. Formulation were prepared by thin film hydration method using cholesterol, bile salts and surfactant. The optimization was performed using Design Expert (DoE) where vesicle size (nm), polydispersity index, and zeta potential (mV), were chosen as dependent variables whereas cholesterol, bile salts and surfactants were as independent variables. *In vitro* antioxidant assay were performed by DPPH method.

Results: *In silico* investigation showed a substantial affinity of selected phytobioactive compounds with TNF- α, IL-6 and IL-1β. DoE optimization identified specific combinations (17 runs). The quadratic model was identified as the most appropriate. The substantial model F-value and P-values below 0.05 indicate that the model is significant. The Predicted R^2 aligns reasonably with the Adjusted R^2, with a discrepancy of less than 0.2. Developed combination formulation showed high antioxidant potential and synergistic effect.

Conclusions: *In silico* studies of phytobioactive compounds showed suitable immunomodulatory effect, this compounds was further developed into nanoformulation

for better bioavailability that provide a foundation for future preclinical and clinical studies aimed at developing effective immunomodulatory therapies.

Keyword: Phytomedicine, immunomodulation, nanoformulation, in vitro assay, molecular docking

PV054 / #507

POSTER SESSION 05: AUTOIMMUNE DISEASE MANAGEMENT
03-07-2025 2:10 PM - 3:10 PM

ALTERATIONS IN THE NATURAL AUTOANTIBODY NETWORK REFLECT THE IMMUNOLOGICAL DIVERSITY OF SYSTEMIC AUTOIMMUNE RHEUMATIC DISEASES

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Background and Aims: Immunological dysregulation disrupts self-tolerance, driving the development of systemic autoimmune rheumatic diseases (SARDs), often associated with the disease-specific pathological autoantibodies (pAAbs). A distinct subset of the autoantibody repertoire, natural autoantibodies (nAAbs), plays a crucial role in maintaining immune homeostasis and tolerance. However, their involvement in SARD pathogenesis remains unclear. We aimed to investigate the associations between nAAbs and pAAbs in different SARDs.

Methods: We grouped 172 serum samples based on pAAb positivity characteristics for rheumatoid arthritis (RA), systemic lupus erythematosus (SLE), Sjögren's syndrome (SjS), and antiphospholipid syndrome (APS). Using in-house ELISAs, we quantified anti-citrate synthase (anti-CS) and anti-heat-shock protein 60/70 (anti-HSP60/70) IgM/G nAAbs. We compared the nAAb profiles of the pAAb-positive serogroups to those of 70 healthy controls (HCs).

Results: Our analysis revealed elevated IgM nAAb levels in the RA serogroup, reduced anti-CS IgM titers in SjS and APS, and a high variability in the SLE serogroup. Regarding IgG nAAbs, we found elevated levels in the SLE group but no significant difference in other serogroups.

Conclusions: Given the heterogeneity of SARDs, distinct immune mechanisms may cause these differences in nAAb levels between the predominantly T-cell-driven RA, where the pAAbs target neo-epitopes, and the immune-complex mediated SLE, characterized by impaired clearance of apoptotic materials. These findings underscore the distinct regulatory roles of nAAbs in SARDs, highlighting the need for further analysis in a clinical

context. Our results could lead to novel diagnostic approaches and treatment strategies for SARDs, including IgM-enriched intravenous immunoglobulin (IVIG) therapy. **Acknowledgment**: TKP2021-EGA10

PV055 / #521

POSTER SESSION 05: AUTOIMMUNE DISEASE MANAGEMENT
03-07-2025 2:10 PM - 3:10 PM

NATURAL AUTOANTIBODIES IN PREGNANT WOMEN WITH AUTOIMMUNE CHRONIC THYROIDITIS

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Background and Aims: The function of natural autoantibodies (nAAbs) and their network has been extensively studied; however, their function in pregnant patients with autoimmune diseases has not been thoroughly investigated. The altered natural autoantibody network in patients with systemic autoimmune diseases has been described, but Hashimoto's thyroiditis (HT), as the predominant organ-specific autoimmune disease of reproductive age in women, has been less investigated.

Methods: The aim of our study was to evaluate IgM and IgG nAAbs against mitochondrial citrate synthase (CS) and heat shock proteins (Hsp60 and Hsp70) in women diagnosed with HT who were pregnant (HTP). Serum samples collected from HTP and healthy pregnant (HP) women in the first and third trimesters were tested by ELISA.

Results: Our results indicate the stability of nAAbs to CS and Hsps throughout pregnancy in both healthy women and those with HT. However, during both trimesters, HTP patients showed increased levels of IgM isotype nAAbs against Hsp60 and Hsp70 compared to HP women, suggesting a regulatory role of IgM nAAbs during pregnancy in patients with HT.

Conclusions: However, the levels of IgG isotype nAAbs against Hsps were lower only in the third trimester in HTP patients, resulting in a higher IgM/IgG ratio, indicating their importance in alterations of the nAAb network during pregnancy in patients with HT.

Keyword: natural autoantibodies, autoimmune diseases, Hashimoto’s thyroiditis, pregnancy, Hsp70

PV056 / #132

POSTER SESSION 06: RHEUMATOID ARTHRITIS
03-07-2025 2:10 PM - 3:10 PM

IS DIAGONAL EARLOBE CREASE ASSOCIATED WITH CARDIOVASCULAR COMORBIDITY IN PATIENTS WITH RHEUMATOID ARTHRITIS?

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Background and Aims: Diagonal earlobe crease (DELC) is a skin clinical sign which can appear on the earlobe between the tragus and the earlobe's free edge. DELC is sometimes associated with the risk of developing vascular diseases of the heart, brain and peripheral arteries. The association of DELC and cardiovascular diseases (CVD) in patients with rheumatoid arthritis (RA) has not been fully investigated. Aim of study was to examine the association of presence and type of DELC with CVD comorbidity in patients with RA.

Methods: Retrospective and cross-sectional case-control study involved 122 patients with RA divided into two groups by whether they had CVD disease or not. Two groups were compared by different clinical and disease parameters.

Results: Patients with RA have positive association of DELC with vascular diseases (OR 1.56, p = 0.51). The association is more expressed with bilateral DELC (OR 1.72, p = 0.42) and with higer grade DELC (OR 2.31, p = 0.25).

Conclusions: In our patients with RA, more pronounced DELC may be associated with the presence of CVS disease.

Keyword: diagonal earlobe crease, cardiovascular diseases, rheumatoid arthritis

PV057 / #140

POSTER SESSION 06: RHEUMATOID ARTHRITIS
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COMPARISON OF EFFICACY BETWEEN OZORALIZUMAB AND GOLIMUMAB IN PATIENTS WITH RHEUMATOID ARTHRITIS

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Background and Aims: Ozoralizumab is a dual-specific antibody composed of an anti-human TNFα variable domain of heavy chain antibody (VHH) and an anti-human serum albumin (HSA) VHH, approved in September 2022 for rheumatoid arthritis (RA) patients inadequately treated with existing therapies in Japan. This study aims to investigate the advantages of ozoralizumab over the conventional TNF inhibitor golimumab.

Methods: We conducted a retrospective analysis of 23 patients treated with ozoralizumab and 20 with golimumab from April 2022 to July 2024, extracted from electronic medical records. Patient demographics, treatment history, and concurrent medication usage were compared.

Results: Both medications were primarily initiated in older patients with a disease duration of over 10 years. Prednisolone usage was similar between the two groups. A higher proportion of patients in the ozoralizumab group had prior biological treatments, especially those with difficult-to-treat cases (26% had received three or more previous treatments). Most discontinuations of ozoralizumab occurred in patients not receiving methotrexate (MTX) (6/7 cases). In contrast, about one-third of golimumab patients were maintained on a regimen of 100 mg every four weeks. The incidence of MTX non-use was greater in the ozoralizumab cohort (70% vs. 60%).

Conclusions: Ozoralizumab appears to be more frequently initiated in difficult-to-treat RA cases compared to golimumab, with comparable therapeutic outcomes. Both treatments were effective even without MTX; however, ozoralizumab may demonstrate superior efficacy, as indicated by its use in a higher proportion of complex cases and the dosing patterns observed in golimumab-treated patients. Further studies are warranted to confirm these findings.

Keywords: Ozoralizumab, Golimumab, Difficult-to-Treat Rheumatoid Arthritis, D2TRA

PV058 / #187

POSTER SESSION 06: RHEUMATOID ARTHRITIS
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COMPARISON OF EFFICACY AND SAFETY OF LOW-DOSE ROSUVASTATIN/EZETIMIBE IN PATIENTS WITH RHEUMATOID ARTHRITIS AND OSTEOARTHRITIS: : A MULTICENTER CLINICAL STUDY

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Background and Aims: To compare the effectiveness and safety of a low-dose rosuvastatin/ezetimibe formulation (rosuvastatin 5 mg/ezetimibe 10 mg) for dyslipidemia in patients with rheumatoid arthritis (RA) compared to those with osteoarthritis (OA).

Methods: This multicenter, open-label, clinical trial enrolled with RA and hand/knee OA. Patients over 19 years who met primary hypercholesterolemia or mixed dyslipidemia, as well as the Korean insurance criteria for rosuvastatin/ezetimibe, were enrolled in the study. The primary endpoint was LDL-C reduction ≥50% from baseline at 12 weeks. Critical secondary endpoints were LDL-C <70 mg/dL and significant adverse events.

Results: A total of 160 RA and 120 OA patients were recruited. Consequently, 145 RA and 110 OA patients completed the follow-up and were included in the final analysis. Compared to the OA group, the RA group had older age, high glucocorticoid usage, and higher acute phase reactants. The primary endpoint of LDL-C reduction ≥50% from baseline was achieved in 98 (67.6%) patients in the RA group and 67 (60.9%) patients in the OA group (p=0.216). No significant difference was found in the safety endpoints. In the multivariate regression analysis, baseline LDL-cholesterol was the only independent factor influencing an LDL reduction ≥50% (OR 1.039, p<0.001). Factors related to RA, such as Disease Activity Score-28 for RA with ESR, C-reactive protein, and steroid use, were not associated with the response to Rosuvastatin/Ezetimibe.

Conclusions: The efficacy and safety of low-dose rosuvastatin/ezetimibe in reducing LDL-C are similar in the RA and OA groups. Baseline LDL-C is the only independent factor influencing the achievement of ≥50% LDL-C reduction.

Keywords: Dyslipidemia, Rheumatoid Arthritis, Osteoarthritis

PV059 / #245

POSTER SESSION 06: RHEUMATOID ARTHRITIS
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THE DEVELOPMENT OF RHEUMATOID ARTHRITIS IN HIGH RISK INDIVIDUALS. CAN IT BE PREVENTED WITH THE USE OF STATINS?

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Background and Aims: Statins have anti-inflammatory and immunomodulatory effects. It has been suggested that statins may have a role in the primary prevention of rheumatoid arthritis (RA).

Methods: We examined the association between statin use and the risk of RA development in two large population cohorts. Furthermore, to investigate a causal relationship between statins and arthritis, we administered statins in the mouse collagen type II-induced arthritis model.

Results: In the first case control study performed using the Netherlands Information Network of General Practice database (n = 800.710 patients), we found an increased risk of incident RA in statin users (adjusted OR, 1.71; CI 1.16-2.53; p = 0.007) whereas the risk to develop RA was not increased in angiotensin converting enzyme inhibitor or angiotensin II receptor blocker users. In the second study performed using the UK Clinical Practice Research Datalink (n = 1.107.988 patients), we also found an increased risk of developing RA in recent statin users (HR adjusted 1.39, CI 1.01 - 1.90), whereas the risk to develop SLE was not increased. Oral statin administration accelerated arthritis onset and resulted in higher arthritic scores in the collagen type II-induced arthritis animal model.

Conclusions: There is no role for statins in the prevention of RA in high risk individuals. In contrast, an increased risk of developing RA was observed in our studies especially within the first six months of statin use. Patients with a high or intermediate 10-year risk of cardiovascular disease statin use should not be discouraged since in patients not prone to develop RA statins are probably safe.

Keywords: Rheumatoid Arthritis, statins, case-control study

PV060 / #247

POSTER SESSION 06: RHEUMATOID ARTHRITIS
03-07-2025 2:10 PM - 3:10 PM

NOURISHING CHANGE: ASSESSING DIETARY HABITS AND CHOICES IN RHEUMATOID ARTHRITIS PATIENTS

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Background and Aims: Diet and nutrition play a crucial role in understanding how environmental factors impact the progression and symptom severity of rheumatoid arthritis (RA). While multiple studies have examined diets that are beneficial for RA patients, few have comprehensively assessed patients' dietary habits, knowledge, and motivations.

Methods: This survey-based study sought to evaluate the dietary choices of RA patients, their willingness to change eating habits, and their baseline understanding of recommended diets. A total of 117 RA patients were recruited from clinic visits, with a demographic breakdown of 82% female, 61% White, 25% Hispanic, and 79% over age 45.

Results: Disease severity was categorized by monthly flares (0-1, 2-4, 5+), yet regardless of flare frequency, the standard American diet remained predominant (51%). Only half reported eating three meals daily, and 39% avoided certain foods due to symptom triggers, with sugary foods being the most common. Motivations for future dietary changes included improving overall health or RA symptoms (71%), weight loss, reducing medication side effects, or addressing non-autoimmune symptoms. A significant barrier to change was a lack of information (42%), highlighting an area for improvement in patient education. Notably, 61% of patients reported never discussing diet with their rheumatologist, while 58% expressed interest in doing so.

Conclusions: These findings underscore a critical opportunity for rheumatologists to engage in meaningful conversations about diet, offering guidance and support to help RA patients adopt healthier eating habits.

Keywords: Rheumatoid Arthritis, diet, nutrition

PV061 / #469

POSTER SESSION 06: RHEUMATOID ARTHRITIS
03-07-2025 2:10 PM - 3:10 PM

LONG-TERM PERSISTENCE OF METHOTREXATE AS A FIRST-LINE THERAPY IN PATIENTS WITH RHEUMATOID ARTHRITIS, PSORIATIC ARTHRITIS AND UNDIFFERENTIATED ARTHRITIS. SINGLE-CENTER RETROSPECTIVE STUDY.

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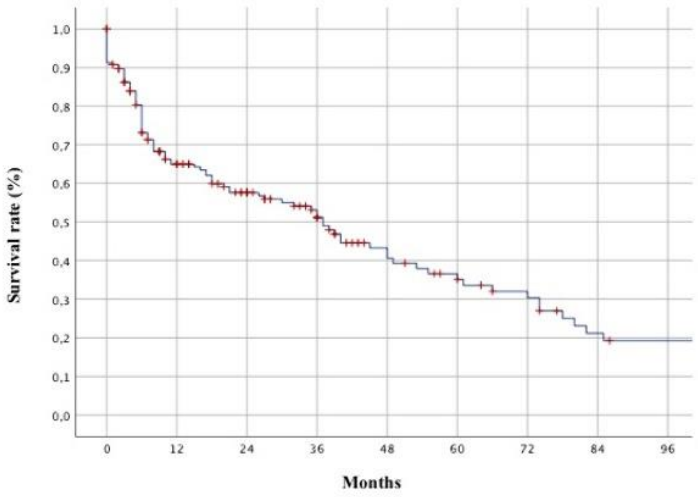
Background and Aims: In the era of biotechnological drugs, methotrexate (MTX) still represents the first-line treatment in chronic inflammatory arthritis such as rheumatoid arthritis (RA) and psoriatic arthritis (PsA). The aim of our study was to evaluate the persistence of MTX as a first-line treatment in a group of patients with chronic inflammatory arthritis (RA, PsA and undifferentiated arthritis-UA).

Methods: Retrospective analysis of our database. Data from patients with chronic inflammatory arthritis who referred to our Unit between January 2014 and January 2022 were analysed. In our outpatient clinic, clinical data (gender, age, smoking habit, body mass index - BMI, C-reactive protein-CRP, comorbidities and treatment are routinely collected. Kaplan-Meier curves (KM) were used to determine the persistence of MTX during follow-up.

Results:

Figure 1. Overall MTX survival rate at 96 months



658 subjects diagnosed with chronic inflammatory arthritis were evaluated. Patients with chronic inflammatory arthritis who started MTX as first-line therapy, with clinical data available, were 242. Of these, 130 (53.7%) with RA, 82 (33%) with PsA and 30 (16.3%) with UA. Overall, the survival rate of MTX at 24 months of follow-up was approximately 60%, while at 48 months and 96 months, it was 40% and 20%, respectively (figure 1). A statistically significant difference was found between RA and PsA compared to UA (Chi-square test=14.84; p=0.001). No statistically significant differences were found for the persistence of MTX in females compared to males, obese versus non-obese patients, older versus younger patients.

Conclusions: Our study confirmed the efficacy and overall safety of MTX in RA and PsA with good persistence even over the long term.

Keywords: Rheumatoid Arthritis, psoriatic arthritis, methotrexate

PV062 / #471

POSTER SESSION 06: RHEUMATOID ARTHRITIS
03-07-2025 2:10 PM - 3:10 PM

CHARACTERISTICS OF PATIENTS WITH RHEUMATOID ARTHRITIS REQUIRING SWITCHING OF BDMARDS AND JAK INHIBITORS IN REAL-WORLD CLINICAL PRACTICE

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Background and Aims: The purpose of this study is to evaluate clinical features and pharmacotherapy in RA patients who needed bDMARDs/JAKi switching.

Methods: 103 RA-patients were enrolled. All patients required switching bDMARDs/JAKi due to non-response, AEs. The majority were women 85.4%, mean age 46.9±13.7years, disease duration 11.0[6.0-16.5]. The patients were median SJC 6.0[4.0-9.0], PJC 11.0[7.0-15.5], PtPGA 70,0[60.0-80.0], PhPGA 70.0[60.0-70.0], CRP 14.6[4.05-33.15], ESR 36[14-64]. There was high disease activity (DAS28-ESR 5.87±1.11, DAS28-CRP 5.42±0.90, CDAI 32[23.5-37.5], SDAI 32.5[25.6-42.0]). Patients used to csDMARDs 77.7%, more often methotrexate 32.0%, leflunomide 30.1%, less frequently sulfasalazine, hydroxychloroquine 11.7% for each. The majority were taking glucocorticoids 62.1%, NSAIDs 80.6%. Most frequently prescribed for switch were rituximab 44.7%, iIL-6 30.1%, less frequently TNFi 14.6%, JAKi 9.6% of patients. Group 1 consist of patients who required the first switch(n=50), group 2 second switch(n=39), group 3 - ≥3 switches of bDMARDs/JAKi.

Results: PJC, PtPGA were significantly higher in groups 2 and 3. These groups showed significant difference CDAI(P_{1-2}=0.01,P_{1-3}=0.013), SDAI(P_{1-2}=0.015,P_{1-3}=0.011). The AEs frequency was significantly higher in group 3(P1-3=0.027, P2-3=0.016). The 41(39.8%) patients had previously used TNFi, 23(22.3%)JAKi, 17(16.5%) iIL-6, 11(10.7%)rituximab, 11(10.7%)abatacept. The 50% of group 1 had previously used TNFi, there were no such patients in group 3(p=0.003), in which iIL-6 were more frequently used 50%vs14.0% and 7.7%, respectively(P1-3=0.006, P2-3=0.002).

Conclusions: The group of patients with repeated switching was characterized by higher PJC, PtPGA and disease activity indices, than group with first switch. The most common bDMARDs for the first switch were TNFi, but for group 3 were no longer prescribed TNFi. iIL-6, JAKi were most frequently used to treat.

Keywords: Rheumatoid Arthritis, bDMARDS, JAKi, switching, ineffectiveness

PV063 / #491

POSTER SESSION 06: RHEUMATOID ARTHRITIS
03-07-2025 2:10 PM - 3:10 PM

ANALYSIS OF FUNCTIONAL AND CLINICAL OUTCOMES IN PATIENTS WITH RHEUMATOID ARTHRITIS DURING ABATACEPT THERAPY.

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Background and Aims: The effectiveness of rheumatoid arthritis (RA) therapy is assessed by various indices, which include laboratory and clinical parts. The aim of our study was to analyze the effect of ABA on clinical indices and functional status in patients with RA.

Methods: 91 patients were included in the study, most of them women, with high disease activity of RA (DAS28=5.1±1.0, SDAI=28±13.4, CDAI=25±12). The average duration of the disease was 3.0 (1.4–12) years, most patients were positive for RF 72.5%, ACCP 77%, treated by ABA (IV, 10 mg/kg). Therapeutic effect was evaluated by EULAR criteria, levels of pain were assessed by visual analogue scale (VAS) and the functional state by HAQ.

Results: ABA treatment led to a significant decrease of disease activity. More than half of the patients were in remission and had low disease activity according to the DAS28 (65.7%, n=35) after 12 months of treatment. Clinical improvement according to EULAR criteria after 6 months of treatment was registered in 70.9% (n=56) and after 12 months was 63% (n=47). ABA treatment led to a significant decrease levels of pain according by VAS starting from 3 months of therapy ($p<0.05$). After 12 month of treatment, levels of pain decreased by 50%. ABA significantly improved functional status of patients, after 12 months a marked and moderate improvement in the HAQ was achieved in 39% and 21% of patients, respectively.

Conclusions: Abatacept has shown significant improvement in clinical and functional status in patients with RA.

Keywords: Rheumatoid Arthritis, biologics, abatacept

PV064 / #497

POSTER SESSION 06: RHEUMATOID ARTHRITIS
03-07-2025 2:10 PM - 3:10 PM

TREATMENT WITH FILGOTINIB DOES NOT IMPACT LIPID PROFILE AND ATHEROGENIC INDEX OF PLASMA IN RHEUMATOID ARTHRITIS.

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Background and Aims: The safety profile of filgotinib requires further investigation in real-world settings, particularly regarding its effects on lipid metabolism markers, such as the atherogenic index of plasma (AIP) and the LDL-C/HDL-C ratio, which are important indicators of cardiovascular risk in rheumatoid arthritis (RA) patients. Our aim was to evaluate the safety of filgotinib by analyzing its impact on laboratory parameters, including white blood cell count (WBC), hemoglobin (Hb), creatinine, total cholesterol (TC), HDL, LDL, triglycerides (TG), AIP, and the LDL-C/HDL-C ratio.

Methods: In this prospective multicenter study, we included all RA patients (according to ACR/EULAR 2010 criteria) regularly followed up every 2–6 months and treated with filgotinib (reimbursed in northeast Italy since September 2021) at two referral centers. Laboratory parameters were recorded every 6 months. AIP was calculated as the logarithmic transformation of the TG to HDL-C ratio.

Results: We enrolled 98 RA patients, with a median age of 59 (52–66) years, mean±SD disease duration of 17±10 years, and mean±SD DAS28-CRP of 3.75±1.27 at baseline. Data on WBC, Hb, creatinine, and cholesterol were available for 72, 72, 40, and 35 patients, respectively. No significant changes were observed in WBC, Hb, creatinine, or any cholesterol type during the 18.6±5.6 month follow-up . AIP showed no significant changes (p=0.494), and the LDL/HDL ratio remained stable (p=0.145). One major adverse cardiovascular event (hemorrhagic stroke) was reported.

Table 1.

Total=98 patients	
Age, median (IQR)	59 (52-66)
Sex, females, n(%)	89 (90)
BMI, mean±SD	24.5± 4.3
Disease duration, mean±SD	17±10
Erosions, n(%)	70 (71.4)
RF+/ACPA+, n(%)	87 (89)

Table 2.

	Baseline	6 months	12 months	18 months
Hemoglobin, g/dL	13.40 ± 1.35	13.45 ± 1.35	13.42 ± 1.27	13.43 ± 1.28
WBC, n/mmc	2535.93 ± 3504.47	2902.15 ± 3438.53	2749.75 ± 3381.40	3241.81 ± 3139.87
Creatinine	0.42 ± 0.38	0.38 ± 0.40	0.38 ± 0.42	0.33 ± 0.43
Total Cholesterol, mg/dL	203.66 ± 39.53	213.27 ± 39.06	225.37 ± 45.76	238.80 ± 42.27
LDL, mg/dL	120.05 ± 35.31	114.60 ± 33.94	121.32 ± 32.53	129.14 ± 33.02
HDL, mg/dL	66.82 ± 15.99	67.57 ± 17.45	69.37 ± 14.32	70.92 ± 12.77
Tryglycerides, mg/dL	102.78 ± 40.64	109.19 ± 36.66	107.00 ± 54.55	110.17 ± 35.30
Atherogenic index of plasma	0.187	0.209	0.188	0.191
LDL/HDL ratio	2.08	1.75	1.72	1.90

Conclusions: This prospective multicenter study supports the favorable safety profile of filgotinib regarding metabolic and hematologic markers in RA patients.

Keywords: RA, filgotinib, lipid

PV065 / #520

POSTER SESSION 06: RHEUMATOID ARTHRITIS
03-07-2025 2:10 PM - 3:10 PM

THE BURDEN OF RHEUMATOID ARTHRITIS IN GHANA: ADDRESSING EARLY DIAGNOSIS, ACCESS TO TREATMENT, AND SOCIOECONOMIC IMPACTS IN A RESOURCE-LIMITED SETTING

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Background and Aims: Rheumatoid arthritis (RA) is a chronic, autoimmune disease that causes inflammation, pain, and potential joint damage. Globally, RA is a significant cause of disability, with varying impacts in different regions. This study aims to examine the burden of RA in Ghana by focusing on early diagnosis and access to treatment.

Methods: Methodology: A mixed-methods approach was utilized, combining a cross-sectional survey and in-depth interviews. The survey was conducted among healthcare providers and RA patients across major hospitals in Ghana, assessing knowledge, diagnostic practices, and treatment availability. Semi-structured interviews with patients and healthcare professionals provided qualitative data on the personal, social, and economic impacts of RA. Data analysis involved both descriptive statistics for quantitative data and thematic analysis for qualitative insights.

Results: Preliminary findings reveal a significant gap in early diagnosis, with many patients presenting at advanced stages of the disease. Access to disease-modifying antirheumatic drugs (DMARDs), including biologics, is limited due to high costs and limited supply. Additionally, many patients experience economic strain, as RA affects their ability to work and care for their families. The study also identifies a lack of trained rheumatologists and the insufficient awareness of RA among healthcare workers as major barriers to optimal care.

Conclusions: Rheumatoid arthritis poses a significant healthcare burden in Ghana, exacerbated by challenges in early diagnosis, limited access to effective treatments, and the socioeconomic impact on affected individuals. Strengthening training programs for healthcare workers, improving public awareness of RA, and increasing the availability of affordable medications are crucial steps toward addressing these challenges.

PV066 / #532

POSTER SESSION 06: RHEUMATOID ARTHRITIS
03-07-2025 2:10 PM - 3:10 PM

CORRELATION BETWEEN PERCEIVED STRESS AND DISEASE ACTIVITY IN RHEUMATOID ARTHRITIS: INSIGHTS FROM PSS-10 AND DAS-28 SCORES

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Background and Aims: Background Rheumatoid Arthritis (RA) is a chronic inflammatory disease marked by systemic inflammation and joint destruction. Stress is increasingly recognised as a factor in RA exacerbation, though its quantitative link to disease activity is unclear. The Perceived Stress Scale (PSS-10) and Disease Activity Score (DAS-28) are validated tools to measure stress and RA activity, respectively. **Objective** This study investigates the relationship between PSS-10 and DAS-28 scores in RA patients to inform integrated care approaches addressing physical and psychological health.

Methods: Methods A cross-sectional study examined stress and disease activity in 229 RA patients meeting 2010 ACR/EULAR criteria. DAS-28 scores were assessed by a consultant rheumatologist, and PSS-10 scores by a junior doctor. Statistical analyses explored relationships and controlled for confounders, including demographics, smoking, antibody status, DMARD adherence, and comorbidities. Ethical approval was secured, and informed consent obtained, with data anonymised.

Results: Results Mean PSS-10 and DAS-28 scores were 18.7 (SD = 5.18) and 3.55 (SD = 1.30), respectively. PSS-10 scores ranged from 7 to 32 and DAS-28 scores from 0.71 to 7.53. A significant positive correlation was found (Pearson's $r = 0.498$, $p < 0.001$), with confidence intervals (PSS-10: 18.0–19.4; DAS-28: 3.38–3.72) supporting the findings.

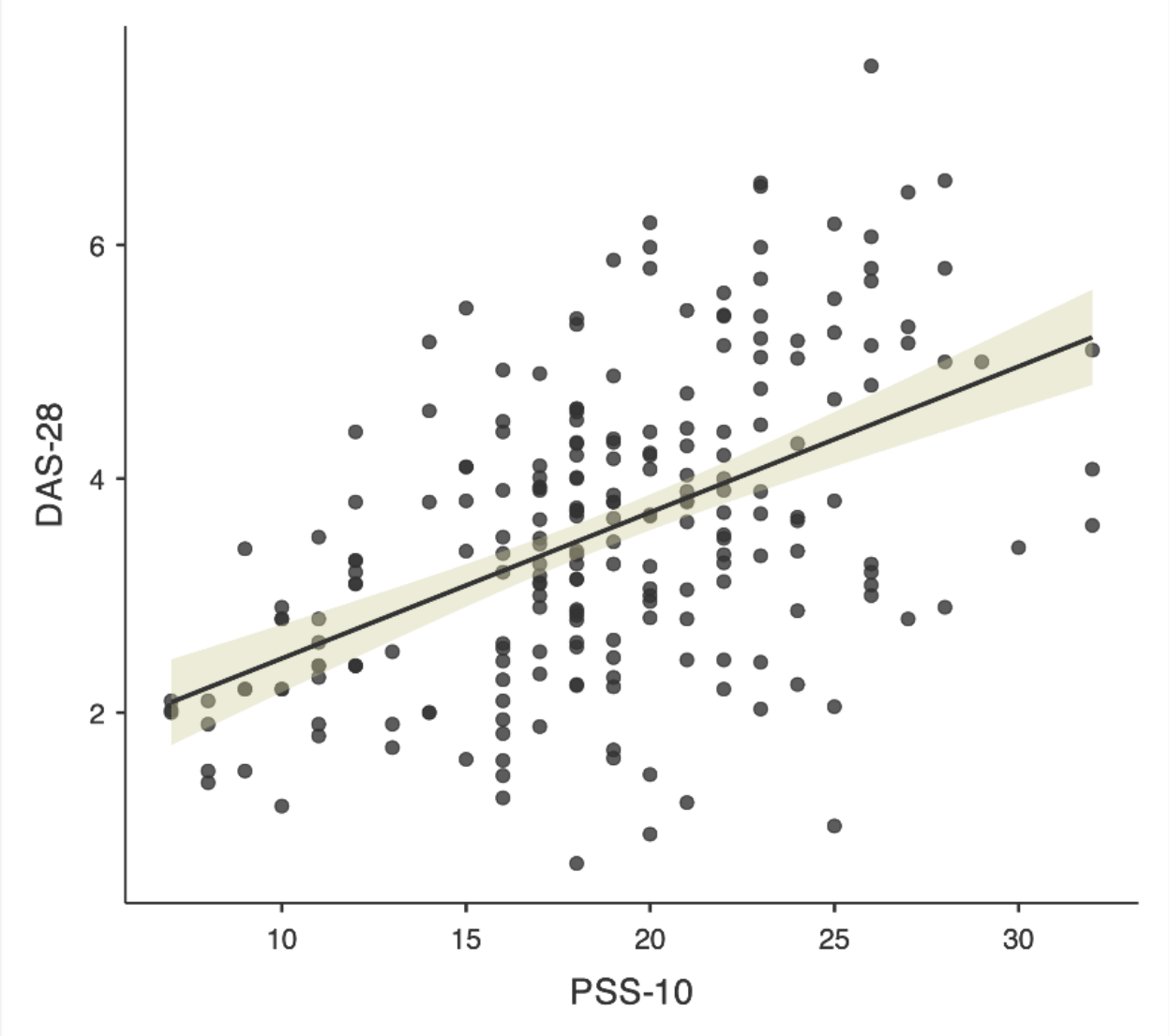

Figure 1 | Scatterplot showing a positive trend between increasing PSS-10 and DAS-28 scores

Conclusions: Conclusion This study highlights a significant correlation between stress and RA activity, underscoring the need for stress management in RA care. Longitudinal studies are recommended to explore causality and the effect of stress reduction on outcomes.

Keywords: Rheumatoid Arthritis, inflammation, stress

PV067 / #554

POSTER SESSION 06: RHEUMATOID ARTHRITIS
03-07-2025 2:10 PM - 3:10 PM

CLINICAL COURSE OF RHEUMATOID ARTHRITIS IN PATIENTS WITH MRI-VERIFIED ATLANTOAXIAL CHANGES

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Background and Aims: Rheumatoid arthritis (RA) affects the cervical spine through the same mechanism as peripheral joints - inflammation of the synovial membrane. However, the underlying risk factors for progression of lesions in the upper cervical spine have not been fully defined. **The Aims:** To evaluate the course of rheumatoid arthritis in patients with MRI-verified atlantoaxial changes.

Methods: Materials and methods: 30 patients with rheumatoid arthritis and various MRI changes in the craniovertebral junction (CVJ) region were included, mean age 53.3±13.51 years. Mean activity score (DAS28 (CRP)) 5,24±0,62. All patients underwent standard general clinical and laboratory-instrumental diagnostic methods, as well as assessment of functional disorders according to HAQ and MRI of the craniovertebral junction with measurement of 5 craniometric parameters for the presence of translocation of the dens axis.

Results: Results: 60% (n=18) had complaints of pain in the neck, mean VAS 6.34±1.84 mm, 40% (n=12) with signs of inflammatory pain in the neck. Median disease duration was 137 [12;660] months. Functional ability (HAQ) carried median values of 1.64±0.61. 36.6% (n=11) were taking glucocorticosteroids (GCS) with a mean prednisolone dose of 4.21±3.83 mg. Correlation between clinical features of RA and changes in CRP was revealed: ESR (rSp=0.470;p=0.018), CRP (rSp=-0.935;p=0.006), number of painful joints (rSp=0.503;p=0.009), age of patients (rSp=0.461;p=0.018).

Conclusions: Conclusion: In the studied cohort of patients with RA, the presence of changes was significantly correlated with inflammatory markers (ESR, CRP), peripheral arthritis, however, there was no association with GCS intake, disease activity and duration, but an association with patient age was detected.

PV068 / #12

POSTER SESSION 06: RHEUMATOID ARTHRITIS
03-07-2025 2:10 PM - 3:10 PM

CLINICAL EVIDENCE OF CHANGES IN CIRCULATING CALPROTECTIN LEVELS AFTER TREATMENT IN RHEUMATOID ARTHRITIS PATIENTS: A SYSTEMATIC LITERATURE REVIEW

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Background and Aims: A differential diagnosis in patients presenting arthritis is needed for an appropriate patient management. Recently, studies published have shown that circulating calprotectin (cCalpro) may be an alternative biomarker of active inflammatory disorders as well as a prognostic or monitoring. We aimed to review and collect all available evidence on the value of cCalpro as a monitoring biomarker in rheumatoid arthritis (RA).

Methods: Electronic, registry and hand searches were conducted to identify studies including cCalpro levels in RA at baseline and after treatment. As Cochrane recommends, the Hedge´s standardized mean difference (SMD) and its 95% confidence intervals were used to synthetize the difference in cCalpro levels. A random-effects model meta-analysis was conducted in STATA MP v17.0. Heterogeneity was investigated by sensitivity analysis, subgroup analyses and meta-regression analyses.

Results: In 15 studies, cCalpro levels of 931 RA patients were significantly higher at baseline than at any timepoint during follow-up (estimated SMD=1.58; 95%CI=1.12-2.03). In 9 studies, cCalpro levels were significantly higher at baseline compared to follow-up in the 522 responders (estimated SMD=2.15; 95%CI=1.59-2.71) but not in the 227 non-responders (estimated SMD=0.44; 95%CI=-0.34-1.21). At baseline there was not statistically significant difference between cCalpro levels in responders vs. non-responders.

Conclusions: In this review and meta-analysis, cCalpro levels were significantly higher at baseline vs. follow-up in responding but not in non-responding RA patients. Pooled Standardized Mean Difference between groups should be interpreted with caution due to substantial heterogeneity. Our meta-data provides further evidence about the potential utility of cCalpro in predicting and assessing treatment response in RA patients.

Keywords: Rheumatoid Arthritis, Calprotectin, systematic review

PV069 / #122

POSTER SESSION 07: VASCULITIDES
03-07-2025 2:10 PM - 3:10 PM

A RARE CONCURRENCE: EXPLORING THE CLINICAL ASSOCIATION BETWEEN TAKAYASU ARTERITIS AND INFLAMMATORY BOWEL DISEASE

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Background and Aims: Takayasu arteritis (TA) and inflammatory bowel disease (IBD) are both relatively rare conditions which share similar autoimmune pathways, populations and systemic immunosuppressive therapies. There are only 150 cases in the literature describing patients with concurrent disease; clinical implications are not fully understood. This study aims to understand how these diseases interact, ultimately improving care for patients with both conditions.

Methods: This is a retrospective study that identified fifteen case patients with both TA and IBD using Advanced Text Explorer and 30 control patients with TA only using SlicerDicer on Epic that were seen from 1/1/2000-7/7/2023. TA diagnoses were confirmed using ACR/EULAR's 2022 Takayasu classification criteria. A 2 to 1 comparison was performed by matching controls to each case by proximity in age and gender. Statistical analyses were performed by Wilcoxon Rank Sum Test (continuous variables) and Fisher's Exact Test (categorical variables).

Results: Both the case and control groups shared similar demographics. Type I Takayasu was more common in the controls at 40% compared to only 6.7% in the cases. Patients with IBD had twice the prevalence of gastrointestinal symptoms. Although not statistically significant, the case group had a higher rate of Takayasu remission at 73% as well as a higher rate of corticosteroid use and pseudoaneurysms.

Conclusions: Our study suggests a close pathogenic relationship between TA and IBD that can result in more disease complications. Both ulcerative colitis and Crohn's disease are implicated in this association. Further research is necessary to understand the interactions of both diseases.

Keywords: Takayasu Arteritis, Vasculiits, Inflammatory bowel disease

PV070 / #263

POSTER SESSION 07: VASCULITIDES
03-07-2025 2:10 PM - 3:10 PM

EFFICACY OF SUBCUTANEOUS INFLIXIMAB [CT-P13] IN MAINTAINING REMISSION IN TAKAYASU ARTERITIS: A MONOCENTRIC CASE SERIES

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Background and Aims: Intravenous (IV) Infliximab is a well-established therapy for Takayasu arteritis (TA). Recently a new formulation of Infliximab, CT-P13, has been developed for subcutaneous (SC) administration. This study aims to assess the efficacy and safety of subcutaneous CT-P13 Infliximab in maintaining remission in patients affected by TA.

Methods: We prospectively enrolled patients affected by TA previously treated with IV Infliximab and we administered CT-P13 120mg every 14 days. Clinical and laboratory data were collected at baseline and during follow-up. Remission was defined according to the Indian Takayasu Clinical Activity Score (ITAS 2010), the Disease Extent Index (Dei.Tak), the National Institutes of Health (NIH) Score and imaging activity, negativity of acute phase reactants and clinical judgement. Safety of the drug was defined as absence of adverse reactions, serious infections and liver toxicity.

Results: Eight patients (100% female, median age 38 years [IQR 27.5-46.8]) in clinical remission were included. Median duration of disease at start was 7.50 years [IQR 4.50-11.3]. No patient was taking steroid therapy, while three patients were on therapy with a DMARD at the start. One discontinued azathioprine during follow-up. Therapy with CT-P13 resulted in the persistence of clinical remission in all patients, according to physician judgment and ITAS 2010, Dei.Tak and NIH scores. No increase in acute phase reactants was observed. No severe systemic adverse drug reactions or liver toxicity were registered. One patient discontinued therapy due to cutaneous side effects.

Conclusions: Subcutaneous infliximab [CT-P13] appears to be effective in maintaining remission in patients with TA. Studies on larger cohorts are needed.

Keywords: Takayasu Arteritis, bDMARD, Anti-TNF

PV071 / #269

POSTER SESSION 07: VASCULITIDES
03-07-2025 2:10 PM - 3:10 PM

PROGNOSTIC VALUE OF 18 FDG-PET AT DIAGNOSIS IN GIANT CELL ARTERITIS: AN OBSERVATIONAL DOUBLE-CENTER RESTROSPECTIVE STUDY

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Background and Aims: Imaging role in giant cell arteritis (GCA) patients is tremendously increased in recent years. However, the role of ^{18}F-FDG PET in predict occurrence of relapse in large vessel (LV)-GCA is still an unmet need.

Methods: Consecutive LV-GCA inpatients and outpatients were prospectively enrolled from 2 vasculitis center. We included all patients who underwent to at least 2 consecutive 18F-FDG PET scans, one at baseline and one during follow-up. Demographic and clinical data as well as disease activity were assessed at each time point. GCA patients were compared according to two treatment regimen: GC monotherapy versus MTX. For each PET scan the vessel's metabolic activity was evaluated using the Meller's grading, the PETVAS score. High arterial FDG uptake was defined as PETVAS ≥ 20, and low arterial FDG uptake was defined as PETVAS < 20, according to Grayson et al 2018.

Results: The study included 53 patients, of whom 38 were treated with GC and 15 with MTX. At baseline, there was no significant difference between these two groups of patients (Table). A total of 41/53 (77.3%) experienced a relapse. In the univariate analysis between the relapse and no relapse groups, no variable was significantly associated with relapse. At baseline, 22/53 (41%) of patients had a PETVAS>20 and no difference was observed in the occurrence of relapse in this group of patients between GC (19/22) and MTX (3/22),

p=0.905.

	Overall (n = 53)	GC (n = 3)	MTX (n = 15)	p GC vs MTX
Demographics, n %				
Female sex, n %	34 (64.2)	23 (60.5)	11 (73.3)	0.381
Age at diagnosis, y, m IQR	67 (60-72)	67 (62 - 72)	62 (56.5 - 72.5)	0.305
Cardiovascular risk factors				
BMI, k/m2, m IQR	24 (21.6-27.4)	24.3 (21.8-28.2)	23.0 (21.1-25.3)	0.303
Previous smoking habits, n %	8 (16.3)	6 (15.8)	2 (13.3)	0.707
Smoking habits, n %	7 (14.0)	5 (13.2)	2 (13.3)	0.929
Arterial hypertension, n %	26 (49.1)	21 (55.3)	5 (33.3)	0.150
Diabetes, n %	4 (7.5)	3 (7.9)	1 (6.7)	0.879
Dyslipidemia, n %	19 (35.8)	17 (44.7)	2 (13.3)	0.032
History of cardiovascular disease, n %				
CKD	14 (26.4)	13 (34.2)	1 (6.7)	0.040
Ischemic cardiopathy	3 (5.7)	3 (7.9)	0 (0)	0.268
Stroke	2 (3.8)	1 (2.6)	1 (6.7)	0.487
Peripheral arterial disease	6 (11.3)	6 (15.8)	0 (0)	0.102
GCA symptoms, n %				
Cranic involvement	22 (41.5)	16 (42.1)	6 (40.0)	0.889
Ocular involvement	4 (7.5)	3 (7.9)	1 (6.7)	0.879
Non ocular involvement	23 (43.4)	18 (47.3)	5 (33.3)	0.353
Weight loss	23 (43.4)	18 (47.3)	5 (33.3)	0.353
Fever	39 (73.6)	28 (73.7)	11 (73.3)	0.979
Headache	24 (45.3)	13 (34.2)	11 (73.3)	**0.010**
Cephalic sign	8 (15.1)	8 (21.1)	0 (0)	0.054
Scalp tenderness	5 (9.4)	4 (10.5)	1 (6.7)	0.665
Jaw claudication	6 (11.3)	5 (13.2)	1 (6.7)	0.502
Visual signs	6 (1.3)	5 (13.2)	1 (6.7)	0.502
Clinical abnormality of tempora artery	5 (9.4)	4 (10.5)	1 (6.7)	0.665
Stroke	2 (3.6)	1 (2.6)	1 (6.7)	0.487
Polymyalgia rheumatica	27 (50.9)	19 (50)	8 (53.3)	0.827
Activity				
CRP, mg/L, m, IQR	79.3 (28-125)	91.3 (16,8-133.3)	62.6 (34.3-100.0)	0.201
VES, mm/h, m IQR	88 (51-100)	80.5 (51.5-95.5)	90 (51-101)	0.813
PETVAS, m IQR	15 (9-23)	15 (8-21)	21 (9-26)	0.575
Dose di prednisone, mg/day, m IQR	50 (35-50)	50 (25 50)	50 (50 - 50)	0.253
Time to first relapse, m, m IQR	13 (8-21)	14.0 (9 - 21)	8 (6.5-15.5)	0.265
PETVAS > 20, n %	22 (41.5)	14 (36.5)	5 (53.3)	0.272
Relapse, n %	41 (77.35)	30 (78.9)	11 (73.3)	0.660
Relapse con PETVAS > 20, n %	22 (53.6)	12 (40)	7 (63.6)	0.905

	RELAPSE (n = 41)	NO RELAPSE (n =12)	p
Demographics			
Female sex	28	6	0.245
Age at diagnosis, y, m IQR	66 (60-72)	69.5 (65.5-74)	0.148
Cardiovascular risk factors			
BMI, k/m2, m IQR	24.3 (21.8-27.8)	23.6 (21.0-25.3)	0.425
Previous smoking habits	7	1	0.461
Smoking habits	4	3	0.151
Arterial hypertension	19	7	0.465
Diabetes	2	2	0.174
Dyslipidemia	17	2	0.115
History of cardiovascular disease			
CKD	11	3	0.899
Ischemic cardiopathy	2	1	0.649
Stroke	1	1	0.346
Peripheral arterial disease	5	1	0.710
GCA symptoms			
Cranic involvement	18	4	0.513
Ocular involvement	4	0	0.260
Non ocular involvment	15	8	0.064
Weight loss	16	7	0.236
Fever	30	9	0.899
Headache	16	6	0.709
Cephalic sign	7	1	0.497
Scalp tenderness	5	0	0.204
Jaw claudication	4	2	0.506
Visual signs	6	0	0.199
Clinical abnormality of tempora artery	4	1	0.882
Stroke	2	0	0.435
Polymyalgia rheumatica	22	5	0.465
Activity			
CRP, mg/L, m, IQR	70.5 (22.3-127.8)	94 (76.6-100.0)	0.986
VES, mm/h, m IQR	79 (50-95)	92 (76.6-100)	0.272
PETVAS, m IQR	18 (8-24)	12.5 (9-18.8)	0.432
Dose di prednisone, mg/day, m IQR	30 (31.2 50)	40 (46.2 50)	0.589
PETVAS > 20, overall (n = 22, 41.5%)	19 /22 (86.5%)	3 / 22 (13.6 %)	0.187
PETVAS > 20, GC (n = 14, 36.5%)	12 (85.7%)	2 / 8 (14.2%)	0.435
PETVAS > 20, MTX (n = 8, 53%)	7/8 (87.5%)	1 / 8 (12.5%)	0.185

Conclusions: PET vascular activity score at diagnosis do not display high performance to predict the occurrence of subsequent relapse in LV-GCA patients.

Keywords: GCA, imaging, Vasculitides

PV072 / #272

POSTER SESSION 07: VASCULITIDES
03-07-2025 2:10 PM - 3:10 PM

A CLINICAL CASE OF EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS AND COEXISTING THROMBOTIC DISORDER

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Background and Aims: A 38-year-old man was admitted to our centre for asthma exacerbation in 2008, with a prior diagnosis of eosinophilic asthma linked to chronic rhinosinusitis with nasal polyps (CRSwNP). During hospitalization, he developed arthromyalgia, hyposthenia, paraesthesia of the left upper limb, and purpura on the lower limbs. Laboratory tests revealed leucocytosis with hypereosinophilia (6000/μL), and positive p-ANCA (100 IU/ml). Chest CT scan showed ground-glass opacities, and skin biopsy of purpura confirmed leukocytoclastic vasculitis. Electroneurography indicated mild sensory-motor axonal polyneuropathy and left ulnar neurapraxic neuropathy. He was diagnosed with Eosinophilic Granulomatosis with Polyangiitis (EGPA) and treated with high-dose corticosteroids, achieving remission. He was discharged on a tapering oral corticosteroid (OCS) and azathioprine but he was lost to follow-up.

Methods: In 2019, he returned to our centre with persistent severe eosinophilic asthma and CRSwNP, both steroid-dependent. After discontinuing azathioprine due to infection, treatment with Benralizumab was started improving asthma but not CRSwNP. He was then switched to Dupilumab, which controlled respiratory symptoms. Although hypereosinophilia persisted, it remains clinical insignificant. After OCS discontinuation, an ANCA-mediated exacerbation (fever and arthromyalgia with ANCA levels of 72.6 UA/ml) occurred, treated with Rituximab.

Results: During lost follow-up, he experienced four transient ischemic attacks (TIA) with left hemiparesis. Investigations revealed antiphospholipid syndrome (APS) with triple positivity, leading to treatment with warfarin and low-dose aspirin.

Conclusions: We propose that the initial left upper limb hyposthenia may have been an early TIA misattributed to neuropathy. In EGPA patients with neurological symptoms, concurrent conditions, especially APS, should be considered.

PV073 / #300

POSTER SESSION 07: VASCULITIDES
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EFFECT OF DAPSONE ON THE TREATMENT OF REFRACTORY IGA VASCULITIS

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Background and Aims: IgA vasculitis (IgAV), previously known as Henoch-Schönlein purpura, is a systemic IgA-mediated vasculitis of the small vessels commonly seen in children. The natural history of IgAV is generally self-limiting. However, one-third of patients experience symptom recurrence and a refractory course. We performed this study to examine the use of dapsone (4,4'-sulfonyldianiline or diaminodiphenyl sulfone) in refractory IgAV cases.

Methods: A literature search of PubMed databases retrieved 16 articles published until Aug 31, 2024. A total of 38 patients with IgAV were recruited.

Results: A palpable purpura persisted in nearly all the patients with refractory course, followed by joint pain (25 of 38 patients; 69.2%). Hematuria and proteinuria were noted in 11 (28.9%) and 7 (18.4%) of the patients, respectively. Treatment response within 1-2 days was obtained in 12 of 38 patients (31.6%), compared to within ≥3 days in 22 patients (57.9%). Relapse after treatment discontinuation was reported in 21 patients (55.3%) but not in 11 patients (28.9 %). Twelve of the 38 patients (31.6%) reported adverse effects of dapsone, including arthralgia (5.3%), rash (5.3%), dapsone hypersensitivity syndrome (2.6%), methemoglobinemia (21%), and hemolysis (15.8%). These findings indicate that dapsone may have a beneficial effect on refractory IgAV.

Conclusions: Despite several side effects, dapsone treatment exerts a beneficial effects in IgAV patients with prolonged, corticosteroid-refractory skin manifestations. Close renal monitoring may be needed to discover renal involvement of IgAV. Multicenter randomized placebo-controlled trials are necessary to determine the standard dosage of dapsone at initial or tapering of treatment in IgAV patients.

Keywords: IgA vasculitis, Henoch-Schönlein purpura, Dapsone

PV074 / #335

POSTER SESSION 07: VASCULITIDES
03-07-2025 2:10 PM - 3:10 PM

THE SPECTRUM OF RENAL INVOLVEMENT IN PATIENTS WITH GRANULOMATOSIS WITH POLYANGIITIS – THE EXPERIENCE OF ONE UNIVERSITY CENTER

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Background and Aims: Granulomatosis with polyangiitis (GPA) is a rare ANCA-associated necrotizing small vessel vasculitis. Renal involvement is observed in the majority of cases. The aim of our study was to evaluate the types of renal involvement in consecutive patients treated in one University Clinic of Nephrology.

Methods: Between September 2017 and August 2024, 27 patients (15 males and 12 females, 21-81 years) with GPA were admitted to a University Clinic of Nephrology. Written informed consent was obtained from all prior to any medical studies. All were taken thorough anamnesis and physical examination, underwent blood and urine investigations, chest X-ray, immunological studies (including ANCA, anti-GBM antibodies and C3+C4 complement levels). Nineteen underwent renal biopsy, 8 were not biopsied: 4 were on dialysis with advanced nephrosclerosis, 4 were treated for acute urinary trat infections without clinical/laboratory data for renal glomerular/tubulo-interstitial disease.

Results: MPO and/or PR3 ANCA were positive in all patients. Renal biopsies revealed crescentic glomerulonephritis (GN) in 12, mesangioproliferative GN in 3, focal and segmental glomerulosclerosis in 2, IgA GN in 1 and hypertensive nephropathy in 1. The histological pattern showed no correlation with the immunological/laboratory parameters.

Conclusions: In a cohort of 27 consecutive patients with GPA in one tertiary center of Nephrology, biopsy-proven renal involvement was detected in 19, with the most common being the crescentic GN in 12. Renal biopsy is crucial for the determination of the type of

renal involvement in GPA, as the latter cannot be predicted only based on the clinical-laboratory and immunological investigations.

Keyword: Granulomatosis with polyangiitis, ANCA, glomerulonephritis, renal biopsy

PV075 / #386

POSTER SESSION 07: VASCULITIDES
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FIGURING OUT GIANT CELL ARTERITIS - WHEN ATYPICAL LOCALIZATION PRECEDES ATYPICAL MANIFESTATION: CASE REPORT.

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Background and Aims: Giant cell arteritis (GCA) is chronic, relapsing inflammatory disease affecting large and medium arteries predominantly in individuals aged ≥ 50 years. Clinical presentation of vasculitis can be versatile and vary during the course of disease making it difficult to diagnose atypical cases.

Methods: A 76-year-old Caucasian female was admitted to hospital with one-month history of mild weight loss, low-grade fevers and headaches in temporal areas without accompanying visual disturbances. Patient showed no signs of arthritis, jaw or extremity claudication. Laboratory tests showed mild anemia and thrombocytosis, elevated inflammatory markers (ESR,CRP). Immunological tests (RF, ANA, ANCA) and blood cultures were negative. Chest and abdominal imaging (X-rays, computed tomography(CT), ultrasonography(US)) demonstrated no evidence of infection, neoplastic process or endocarditis. Brain magnetic resonance(MR) angiography, US of arteries (temporal, carotid, subclavian, axillary), and CT angiography of aorta and its branches didn't reveal any signs of vasculitis. Finally 18-FDG-PET/CT unmasked bilateral inflammation of femoral and popliteal arteries(Figure 1). Diagnosis of GCA was made and patient was successfully treated with glucocorticoids (GCS), tapered down over 18 months, and methotrexate (17,5

mg/week).

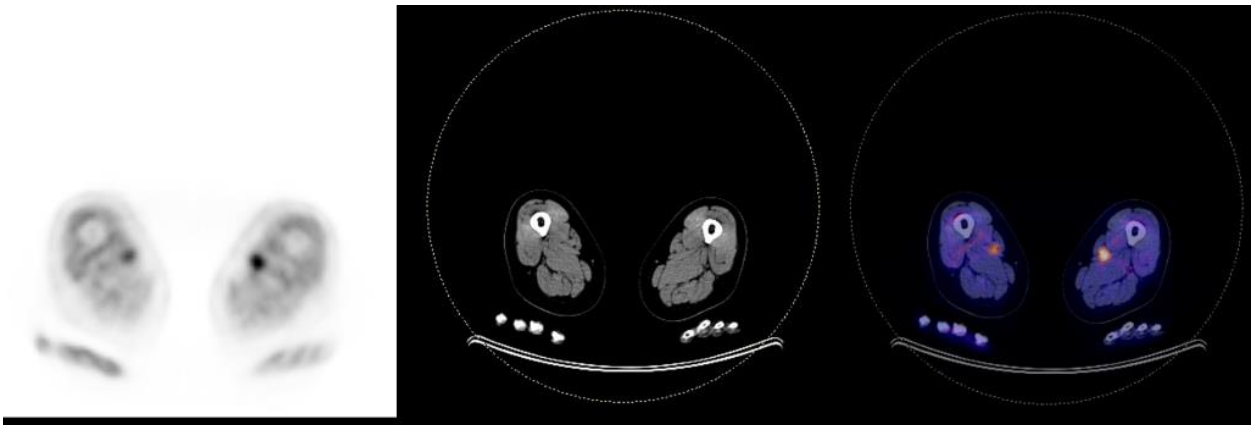

Figure 1. Positron emission tomography (PET)/CT scan (transversal planes). Increased 18-fluorine-2-deoxy-D-glucose(18-FDG) uptake in femoral arteries bilaterally suggesting arterial inflammation. The panel on the left shows PET, the middle panel shows CT, and the panel on the right shows the overlap of the two (PET/CT)
Scan source: Nuclear Medicine Department, Medical University of Warsaw, Warsaw, Poland

Results: A relapse presenting with persistent dry cough, anorexia and nausea occurred 8 months after GCS cessation. Thorax and abdomen CT angiography revealed vasculitis of abdominal aorta, mesenteric and renal arteries(Figure
2).

Figure 2. Abdominal CT angiography (transversal planes). The scan on the left, performed at the time when GCA diagnosis was made, shows no evidence of vasculitis. The scan on the right performed 3 years later (during relapse of GCA) demonstrates mural thickening of aorta, superior mesenteric and renal arteries (arrows)
CT scans source: Department of Radiology, National Institute of Geriatrics, Rheumatology and Rehabilitation, Warsaw, Poland

Conclusions: Giant cell arteritis can be diagnostically fairly challenging especially in less common extracranial localizations or in the absence of ischemic symptoms. Our case demonstrates that relapse symptoms may differ from original manifestations implying other sites of vessel involvement than initially.

PV076 / #418

POSTER SESSION 07: VASCULITIDES
03-07-2025 2:10 PM - 3:10 PM

REVIEW OF A GIANT CELL ARTERITIS COHORT OF A TERTIARY HOSPITAL

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Background and Aims: Giant Cell Arteritis (GCA) is large vessel vasculitis. Clinical manifestations and complementary exams may be unspecific, and relapses have been scarcely addressed. It is paramount to revisit the large cohorts of GCA to understand the phenotype of this disease and to do a retrospective analysis of its clinical and systemic manifestations and therapy options.

Methods: We performed a retrospective analysis of patients with GCA who were followed in the Immunology Clinic of a tertiary hospital between 1999 and 2024. We aimed to investigate the characteristics at baseline, evolution, and characteristics of relapses and to analyze whether a relapsing course is associated with disease-related complications.

Results: 85 patients, with an average age of 78 years old, 78% were female. Headache and scalp tenderness were the most frequent cranial symptoms, and fatigue was the most prevalent extra-vascular symptom (half of the patients). Cranial GCA is the most frequent phenotype, and the most related to PMR. 59% were on a monotherapy regimen, mainly steroids. 41% had a steroid-sparing agent. The relapse rate was 40%. Only a third relapsed with the same phenotype. The majority (85%) of relapses was treated with high doses of steroids, TCZ is the second preferred drug used in almost 40% of the patients. The cumulative steroid dosis is very high, with an average of 30 months, and a rate of steroids-related adverse events around 60%.

Conclusions: GCA should be addressed in a personalized manner. Relapses are a problem in GCA, and treatment can be challenging: better strategies will be the future of GCA.

Keywords: Vasculitides, relapse, Giant Cell Artritis

PV077 / #582

POSTER SESSION 07: VASCULITIDES
03-07-2025 2:10 PM - 3:10 PM

EFFICACY AND SAFETY OF EXTENDED-INTERVAL RITUXIMAB VS. STANDARD MAINRITSAN REGIMEN FOR REMISSION MAINTENANCE IN ANCA-ASSOCIATED VASCULITIS: A MULTICENTER STUDY

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Background and Aims: Rituximab(RTX) demonstrated efficacy in maintaining remission in patients with ANCA-associated vasculitis (AAV). However its cost and potential for adverse effects necessitate optimizing dosing strategies. We aim to compare the effects of Extended-interval of RTX(500 mg once per year) to standard MAINRITSAN regimen RTX (500 mg twice per year) as remission-maintenance therapy in AAV.

Methods: GPA and MPA patients in clinical remission (BVASv3=0), referring to four Rheumatology centers in Italy, were treated with RTX for remission maintenance. Maintenance regimens were classified in the MAINRITSAN (RTX 500-mg every 6 months) and the Extended-Interval(RTX 500 mg once a year) group. After at least two consecutive infusions,we assessed efficacy and complications.

Results: From 2011 to 2024,100 AAV patients (median age 60 [54-70] years, 54% female, 97% ANCA positive), 65 GPA and 25 MPA were included. Eighty-two started maintenance with the standard regimen, 18 with Extended- interval.Groups were homogeneous at disease onset, except for BVASv3, higher in Extended-interval group (p=0.038). 29 patients switched from MAINRITSAN to Extended-interval during maintenance. At the last follow-up, there were no differences between groups in severe hypogammaglobulinemia (3.6% vs. 9.1%), infections (5.4% vs. 9.1%), ANCA negativization (66.1% vs. 56.8%), glucocorticoid discontinuation (51.8% vs. 72.7%). All patients had BVASv3 = 0. One patient in each group experienced a relapse (p = 0.689), both with CD19 repopulation and ANCA positivity. Death rate was 3.6% in the MAINRITSAN and 4.5% in the Extended-Interval group, all caused by SarsCov2 infections.

Conclusions: Remission maintenance with extended-interval of RTX is a safe and more cost-effective option in AAV patients.

Keywords: AAV, Rituximab

PV078 / #119

POSTER SESSION 08: MISCELLANEOUS
03-07-2025 2:10 PM - 3:10 PM

CROWNED DENS SYNDROME, CLINICAL PRESENTATION, IMAGING FINDINGS AND TREATMENT APPROACHES FROM SINGLE CENTER COHORT

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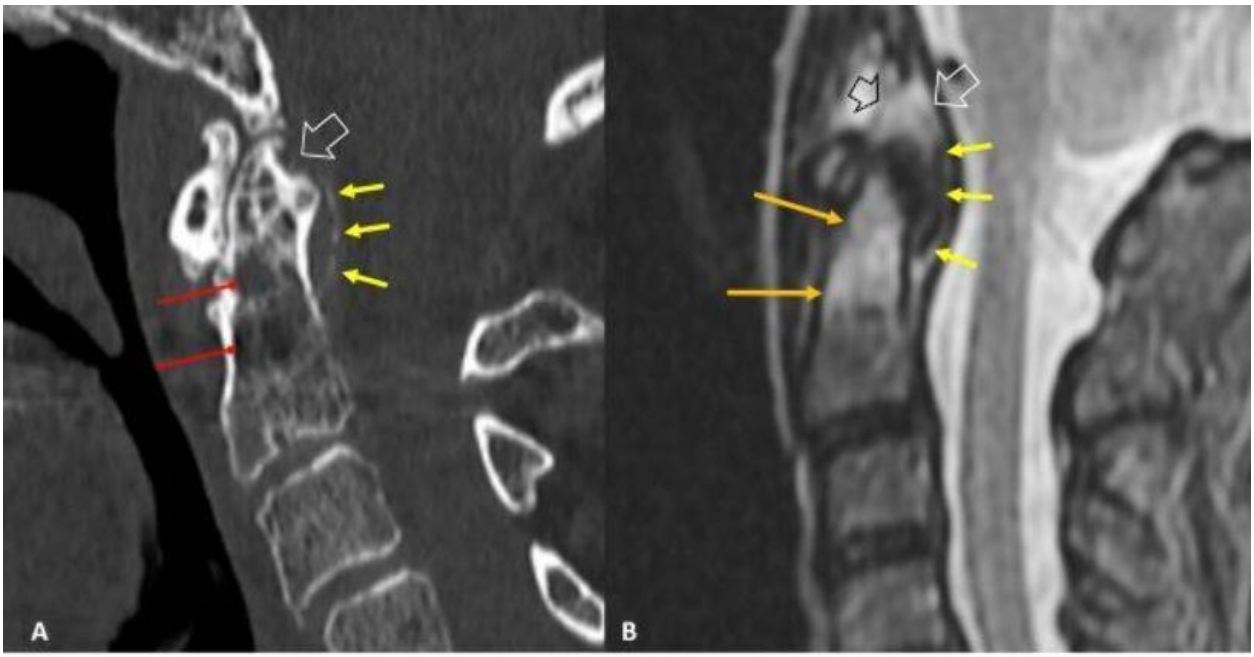
Background and Aims: Crowned dens syndrome (CDS) is rare disease resulting from crystal deposition in the cruciform and alar ligaments surrounding the dens, appearing as a radiopaque "crown" on the top of the dens. Our aim was to identify all patients with such final diagnosis and describe clinical presentation, imaging findings, treatment approaches and outcome.

Methods: We reviewed retrospectively (2016-2023) the medical notes of all patients who after clinical assessment and relevant imaging had as final diagnosis (CDS).

Results: We identified two females and one male. They were 76, 66 and 81 years old respectively. Their main symptoms were acute- subacute neck pain, restrictedrange of motion and low-grade fever. Elevated inflammatory markers were observed in all three patients. The imaging findings are shown in (Figure 1). One female patient had medical history remarkable for multiple myeloma. All patients initially had received moderate to high dose of steroids (0.5-0.75 mg/kg/day) with gradual tapering. The two female patients had excellent clinical response and after stopping steroids were continued with colchicine 0.5mg/day. The male patient had moderate clinical response and also side effects from colchicine and has continued his treatment with IL-1 inhibitor (Anakinra). **Figure**

1.



A .76-year-old female patient with known multiple myeloma. The sagittal CT reconstruction, shows calcifications posterior to dens, in keeping with "crowned dens" syndrome (arrows). An erosion is shown at the upper dens (open arrow). Two multiple myeloma lesions are shown in the base of the dens and the C2 body (red long arrows).
B. 66-year-old female patient with severe neck pain, with marked restriction of rotatory neck motion. Sagittal STIR MR image snowing the bone marrow edema (long arrows), linear low signal calcification posterior to the dens (short arrows), and high signal intensity inflammatory tissue cranial to dens (open arrows).

Conclusions: Due to its rarity and non-specific manifestations, diagnosis of (CDS) is frequently missed. Therefore, physicians when older patients have acute neck pain accompanied by a restricted cervical range of motion and elevated inflammatory markers must consider (CDS) in differential diagnosis.

Keyword: CROWNED DENS SYNDROME

PV079 / #199

POSTER SESSION 08: MISCELLANEOUS
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ATYPICAL ANTI-SAE-1 AUTOANTIBODY MANIFESTATION WITH SPLINTER HEMORRHAGES, AS ONSET, AND RELATED TO UROTHELIAL CANCER

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Background and Aims: Description of a case report of urothelial cancer as onset with paraneoplastic syndrome positive to Anti-SAE-1 autoantibody, directed against a small ubiquitin-like modifier-activating enzyme, that plays a role in regulating transcription, cell cycle, and apoptosis. It is specific for dermatomyositis with skin rash and mild muscle involvement and they can be related to cancer association.

Methods: The anti-nuclear antibodies (ANA) was detected by indirect immunofluorescence (IIF). The confirmatory test was performed by line-blot technology following the manufacturer’s instructions. The test is specific for the following antigens. EUROLINE Myositis: antibodies anti: Mi-2 alpha, Mi-2beta, TIF1g, MDA5, NXP2, SAE1, Ku, PMScl100, PM-Scl75, Jo-1, SRP, PL-7, PL-2, EJ, OJ, Ro-52.

Results: We describe the case of a 50-year-old woman, no smoke, no exposure to toxic substances, but with a familiarity with lung and breast cancer respectively from a father and mother. She developed subungual splinter hemorrhages and acrocyanosis. Infections were excluded and autoimmunity resulted in anti-SAE-1 autoantibody-positivity by IIFest. A cancer screening found a high-grade urothelial carcinoma. The splinter hemorrhages disappeared after the carcinoma enucleation. The peculiarity of our case is the unusual clinical onset of anti-SAE dermatomyositis with splinter hemorrhages. Anti-SAE

autoantibody combined with splinter nail hemorrhage, as the spy of urothelial cancer, has never been described in the literature.

Conclusions: We reviewed the literature and we believe that our case may contribute to the knowledge of atypical dermatomyositis onset and opens the necessity of cancer screening where unusual manifestations are found.

PV080 / #270

POSTER SESSION 08: MISCELLANEOUS
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A COMPLEX CASE OF COMMON VARIABLE IMMUNODEFICIENCY WITH JAK3 VARIANT AND AUTOIMMUNE AND ALLERGIC COMPLICATIONS

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Background and Aims: Common Variable Immunodeficiency (CVID) with concurrent autoimmune disorders and allergic disease could be a challenge, moreover in patients harboring genetic variants. In literature, *JAK3* gene defects result in a type of T cell-negative, B cell-positive, natural killer (NK) cell-negative severe combined immunodeficiency (T-B+NK- SCID), with autosomal recessive heritance[1].

Methods: A 76-year-old Caucasian woman was diagnosed with hypogammaglobulinemia, after screening to initiate venom immunotherapy (VIT) because of an history of two episodes of anaphylaxis following wasp stings. She also reported with a previous episode of pneumonia, adverse reactions (urticaria) to paracetamol and sulfamides and a gastrointestinal stromal tumor (GIST). Moreover, patient exhibited psoriatic arthritis with joint pain in the knees, elbows, shoulders, hands, and lower back.

Results: Initial laboratory tests revealed IgG 323, IgA 68, IgM 19 mg/dl with low IgG1 subclass. Screening for autoimmunity was negative. Following CVID diagnosis, patient received intravenous immunoglobulin (IVIg) replacement therapy at the dose of 20 g/month. Recent clinical issues include candidiasis, conjunctivitis. Imaging also shown bronchial wall thickening, mild emphysema, and aortic dilation. Genetic testing identified a heterozygous VUS in the *JAK3* gene (c.452C>G), which could be linked to her immune dysregulation.

Conclusions: JAK3 c.452C>G mutation carried by our patient was a variant of unknown significance (VUS) related to combined immunodeficiency when associated to another variant. Further clinical research and functional analysis are needed to explore the role

of *JAK3* mutation in complicated CVID. [1]Roberts JL et al, Blood. 2004. Goldberg L et al. Genes Immun. 2020

Keywords: psoriatic arthritis, JAK3 Variant, Common Variable Immunodeficiency

PV081 / #277

POSTER SESSION 08: MISCELLANEOUS
03-07-2025 2:10 PM - 3:10 PM

MULTISYSTEM INFLAMMATORY SYNDROME IN CHILDREN (MIS-C) ASSOCIATED WITH COVID-19 INFECTION IN MOROCCO.

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Background and Aims: Multisystem inflammatory syndrome in children (MIS-C) emerges as a critical, hyperinflammatory condition following COVID-19 infection, presenting a substantial threat to pediatric health. This study aims to describe the epidemiological, clinical and paraclinical characteristics of Multisystem inflammatory syndrome in children (MIS-C).

Methods: We conducted a retrospective descriptive study encompassing 52 Moroccan children diagnosed with MIS-C over a three-year period (March 2020 - March 2023) at the Infectious Diseases and Clinical Immunology Department at Abderrahim Harouchi Hospital. The selection of patients for our study adhered to the MIS-C inclusion criteria as outlined by the World Health Organization (WHO).

Results: The median age was 6 years (IQR : 1-14), with a sex ratio of 1.16. Clinical manifestations were predominantly characterized by fever in all cases (100%), respiratory and gastrointestinal symptoms in 30 cases (58%) and 23 cases (44%) respectively, and shock in 9 cases (17%). The biological tests showed a positive SARS-CoV-2 PCR in 10 patients (19%). Serology was positive in 42 cases (81%), with anti-SARS-CoV-2 Ig M positive in 25 cases (48%) and Ig G positive in 31 cases (60%). We noted an elevated D-dimer levels in 30 cases (58%) with myocarditis in 6 cases (12%). Interleukin 6 levels were elevated in 12 cases (23%). The treatment comprised intravenous human Immunoglobulin combined with methylprednisolone in all patients (100%).

Conclusions: The characteristics of our MIS-C patients were similar to those in the literature, but more studies are needed to confirm these results.

Keywords: COVID-19, MIS-C, Children

PV082 / #317

POSTER SESSION 08: MISCELLANEOUS
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PYODERMA-PYOSTOMATITIS VEGETANS: EXPLORING A RARE MANIFESTATION OF ULCERATIVE COLITIS

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Background and Aims: Inflammatory bowel diseases often present with extra-intestinal manifestations, including dermatological symptoms, which can sometimes correlate with exacerbations or relapses of intestinal inflammation. This report aims to present a case of pyoderma-pyostomatitis vegetans (PV), a rare but pathognomonic skin manifestation of ulcerative colitis (UC).

Methods: A 39-year-old female with a medical history of hypothyroidism and UC, treated with mesalamine, presented with painful mucocutaneous lesions. The lesions developed during a pause in UC treatment due to pregnancy and breastfeeding. Clinical examination, laboratory tests, and microbial cultures were conducted to establish the diagnosis.

Results: The patient exhibited multiple coalescing, fragile, white-yellow pustules on an erythematous base on the gums and oral mucosa, leading to ulcerations. Additionally, vegetative plaques with seropurulent exudate were noted in the nasal mucosa, causing nasal obstruction, and pruritic papulovesicular lesions on the face and extremities. Laboratory findings demonstrated eosinophilia, with negative serological results for hepatitis and HIV, and sterile microbial cultures from the oral lesions. Based on the clinical presentation and medical history, a diagnosis of PV was established. Topical corticosteroids were initiated, and the patient was referred to gastroenterology for UC management. Sigmoidoscopy revealed no exacerbation or relapse of the disease; however, due to the presence of mucocutaneous lesions, adalimumab therapy was administered.

Conclusions: PV is a rare but specific mucocutaneous manifestation of UC that significantly impacts patients' quality of life. Its management remains challenging, as it is often resistant to topical treatments and typically requires appropriate control of the underlying UC.

Keywords: ulcerative colitis, cutaneous manifestation, pustules

PV083 / #417

POSTER SESSION 08: MISCELLANEOUS
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RESIDUAL PAIN AFTER PRIMARY TOTAL KNEE ARTHROPLASTY

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Background and Aims: According to literature above 20% patients after TKA are unsatisfied. The most common patient's complaint is pain.

Methods: In our prospective study 196 patients were examined from 2016 to 2023 with chronic pain after primary TKA. The mean time after TKA was 32 months, mean age was 67 y.o. All patients were examined with the accurate algorithm, in which were included medical history, CT scans, special X-rays and PJI examination tests.

Results: The reason of pain was identified in all patients. PJI was identified in the most part of our cohort – 76 (34%) patients. Among PJI patients the components malposition was founded in12 (15.8%) patients. The second group was with components malalignment – 61 (27%). Aseptic loosening was identified in 38 (17%) and 9 (23.7%) patients in this group has the components malalignment according to early X-rays after TKA. The ligament instability was in 24(11%) patients. Extraarticular reason was identified in 19(9%) patients and among them 5 (2%) patients were with periprosthetic fractures. It's very important, that in 27 (13.7%) patients several reasons of knee pain were identified

Conclusions: It's very difficult to identify the real problem. The most common definition as "arthrofibrosis" is usually only a symptom. According to our research, the most frequent reasons of unsatisfied knee were PJI and component malalignment. Only 19 (9%) patients had an extraarticular reasons. The comprehensive examination can help to identify the real problem. But in different National registries very often reason for revision is pain without its course verification.

Keywords: residual pain, total knee arthroplasty, periprosthetic joint infection

PV084 / #429

POSTER SESSION 08: MISCELLANEOUS
03-07-2025 2:10 PM - 3:10 PM

MAMMARY SARCOIDOSIS MIMICKING BREAST CANCER: A CASE REPORT.

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Background and Aims: Sarcoidosis is an inflammatory disorder, characterized by a multisystemic affection by a noncaseating granuloma formation that affects most commonly the lungs and hilar lymph nodes. [1] The diagnosis is achieved by exclusion, supported by histological, clinical features and/or imaging evidence of noncaseating epithelioid cell granulomas.[2] Processes like neoplasms, may mimic a sarcoid-like granulomatous reaction.[3] Given the prevalence of breast cancer, it should be carefully considered during the diagnostic evaluation for sarcoidosis.

Methods: Case Report

Results: A previously healthy 38-years-old female first presented due to nodular lesions on left breasts with green-like secretion. The family history was negative for neoplastic diseases. She had never been treated for tuberculosis but lives with her mother, who is known to have a positive TBC GeneXpert. Physical examination revealed big and erythematosus left breast, multiple nodules that were immobile with overlying inflammation, non-tender that drains a bloody serous secretion. A mammary ultrasound revealed various nodular images, hypoecoic with good sound transmission**.** A biopsy demonstrated the presence of a granulomatous lesion with giant cells, leading to a preliminary diagnosis of Granulomatous Mastitis.

Conclusions: Mammary sarcoidosis is seen in less than 1% of sarcoidosis cases, in which it presents in very variable ways. In this case, the main goal was to discard the possibility of breast cancer.

Keywords: Sarcoidosis, cancer, mammary

PV085 / #430

POSTER SESSION 08: MISCELLANEOUS
03-07-2025 2:10 PM - 3:10 PM

CLINICAL CHARACTERISTICS OF PATIENTS WITH SARCOIDOSIS ATTENDING A RHEUMATOLOGY CLINIC

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Background and Aims: Sarcoidosis is a systemic disease of unknown etiology, characterized by the presence of non-caseating epithelioid cell granulomas with infiltrates of leucocytes.1,2

Methods: We present a retrospective descriptive study of patients who attended a rheumatology clinic between 2002 and 2019. Only patients with pathology findings consistent with non-caseating granulomas, and in whom other causes of granulomatous disease were ruled out, were included.

Results: Ten patients were included. The female:male ratio was 5:5, with an average age of 47 years (range 29 to 72 years) at the time of diagnosis. Lung involvement present in n=5, diagnosed by chest X-ray or high-resolution CT scan, with 2 patients in stage 2, 2 in stage 3, and 1 in stage 4. Lymph node involvement was observed in n=9. Osteoarticular involvement (n=4) was observed in the form of posterior tibial tendinitis (n=1), vertebral body granuloma (n=1), carpal erosions (=1), and carpal synovitis (n=1). Systemic symptoms were present (n=3). One of these cases had severe hypercalcemia (17 mg/dL) and another had moderate hypercalcemia. A total of n=3 had hypercalciuria. Rare organ involvement (n=3), such as nervous system involvement, large-vessel vasculitis (n=1), small-vessel vasculitis (n=1), myocardial involvement (n=1), ascites, and renal involvement were also present(n=1).

Conclusions: The pleomorphism of its clinical presentation forces us to always consider it in the diagnosis of a patient with involvement of multiple organs and systems.

Keywords: Sarcoidosis, characteristics, clinical

PV086 / #551

POSTER SESSION 08: MISCELLANEOUS
03-07-2025 2:10 PM - 3:10 PM

HISTOLOGICAL CHANGES IN THE ATRIAL AURICLE STRUCTURES IN PATIENTS WITH POSTOPERATIVE ATRIAL FIBRILLATION AFTER CARDIAC SURGERY

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Background and Aims: Atrial fibrillation (AF) is one of the most common types of cardiac arrhythmias in clinical practice and the risk factor for thromboembolism and stroke. The global AF prevalence in elderly patients has increased markedly. This study was performed to make clinical and morphological assessment of postoperative AF using cardiac biopsy specimens taken during surgery.

Methods: Tissues of 53 atrial auricles were removed during cardiac surgery, including 32 auricles of patients with AF and 21 patients without AF (comparison group). Van Gieson's, Mallory's, and Lie-staining were used for histological examination of the paraffin sections, and immunohistochemical analysis – for detecting desmin, S100, S-117.

Results: Patients with AF had significantly more pronounced lesions of cardiomyocytes, including myocytolysis, increased vascular permeability, focal lymphohistiocytic infiltrates, microfocal cardiosclerosis, endocardial sclerosis and myocardial steatosis. The presence of mucoid edema with mural thrombi was reported in some endocardial regions. Immunohistochemistry (IHS) revealed the destruction of cardiomyocyte cytoskeleton in the sites of myocytolysis, and a reduced E-cadherin response in the intercalated disks. Single S-100-positivee telocytes were identified in both patient groups.

Conclusions: The destructive changes of cardiomyocytes dominated in the atrial auricle tissues in patients with postoperative AF. They included myocytolysis with the destruction of cytoskeleton stained positively by desmin. increased permeability of intramural blood vessels, lymphohistiocytic infiltrates, myo- and endocardial sclerosis, and mural thrombi of the endocardium.

Keywords: cardiac arrhythmias, thromboembolism, histological examination, lymphohistiocytic infiltrates, Immunohistochemistry

PV087 / #567

POSTER SESSION 08: MISCELLANEOUS
03-07-2025 2:10 PM - 3:10 PM

PATHOMORPHOLOGICAL EVALUATION OF EOSINOPHILIC ESOPHAGITIS AT FIRST BIOPSY AND AFTER TREATMENT

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Background and Aims: Eosinophilic esophagitis (EoE) is a chronic immune-mediated esophageal disease with predominantly eosinophilic inflammatory infiltration of esophageal mucosa (≥ 15 eos/hpf) and various degree of subepithelial fibrosis. The aim of our study was to analyze histological features of EoE in patient at first biopsy and after treatment with proton pomp inhibitors (PPI).

Methods: Biopsy was performed in 70 patients that fulfilled EREFS criteria for EoE and in 25 patients with previously histologically proven EoE after treatment. Biopsy specimens were fixed in 10%-neutral buffered formalin, stained with haematoxylin and eosin and Mallory for assessment of fibrosis. EoE histology scoring system (EoEHSS) and EoE Histology Remission Score (EoEHRS) were applied for histological evaluation.

Results: Among 70 patients with histologically proven EoE 51 were men (72,86%), median age was 29.5 (21; 42.25). Peak eosinophil count (PEC) ranged from 17 to 222 (Me 52 eos/hpf). Basal zone hyperplasia ranged from 20 to 80%. Eosinophilic abscesses presented in 48,57%, surface layering in 17,14%, dilated intercellular spaces – in 97,14% of patients. Surface epithelial alteration was noticed in 60% and dyskeratotic epithelial cells – in 11,42% of cases. Lamina propria of mucosa was present in 71,43% of biopsy specimens, among them various degree of fibrosis was observed. EoEHSS median grade score comprised 12 (8; 14.5), median stage score was 10 (8; 12). 92% of patients after treatment fulfilled EoEHRS criteria of histological remission.

Conclusions: EoEHSS and EoEHRS are powerful tools to evaluate pathomorphological features of EoE at first biopsy and after treatment. Treatment with PPI was effective in most patients

Keywords: eosinophilic esophagitis, inflammatory, fibrosis, biopsy, epithelial, hyperplasia

PV088 / #568

POSTER SESSION 08: MISCELLANEOUS
03-07-2025 2:10 PM - 3:10 PM

COMPARATIVE CHARACTERISTICS OF THE IMMUNE LANDSCAPE OF THE COLON MUCOSA IN DIVERTICULAR DISEASE

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Background and Aims: Diverticular disease is one of the most common diseases of the gastrointestinal tract. In 15% of cases, complications may arise that require urgent surgical intervention. In elective surgery, the postoperative mortality rate is less than 2%, while in emergency operations this figure already reaches 20%. Thus, it is extremely important to determine the predictors of complicated diverticular disease for timely diagnosis, adequate routing and effective treatment of this category of patients. The aim of the study was to evaluate the local immunity of the colon mucosa in patients with complicated and uncomplicated diverticular disease in order to clarify its pathogenesis and to identify morphological predictors of complicated course, as objective arguments in favor of planned surgery in a number of patients.

Methods: A retrospective comparative analysis of the qualitative composition of local immunity of the colon mucosa was performed in surgical material from 68 patients who underwent left-sided hemicolectomy with diverticular disease of varying course and without diverticula

Results: It was shown that the number of CD4+, CD8+ and C138+ immune cells differed in all compared groups, and the number of CD56+ and CD68+ cells differed in the groups with diverticular disease and without diverticula ($p<0.05$).

Conclusions: The conducted study demonstrates significant differences in the qualitative composition of immunocompetent cells of the colon mucosa in groups of patients with diverticular disease and without colon diverticula, which shows the significance of

changes in local immunity in the development of various variants of the course of diverticular disease.

Keywords: immunocompetent cells, histological remission, Diverticular disease, inflammation, biopsy, basal, immune cells

PV089 / #540

POSTER SESSION 08: MISCELLANEOUS
03-07-2025 2:10 PM - 3:10 PM

CLINICAL FEATURES AND MORPHOLOGY OF COVID-19 IN A PATIENT WITH RHEUMATOID ARTHRITIS COMPLICATED BY SYSTEMIC AA AMYLOIDOSIS. CASE STUDY

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Background and Aims: The COVID-19 pandemic was particularly challenging for patients with rheumatoid arthritis (RA) and secondary systemic AA amyloidosis. This case study reviews the clinical characteristics and morphology of COVID-19 in a 69-year-old female patient with seronegative RA (moderate disease activity; negative AB-CCP; Class III) and AA amyloidosis not diagnosed in her life.

Methods: For performing histopathological examination, the samples were fixed in 10% neutral buffered formalin solution followed by paraffin embedding. All histological sections were stained with H&E. Then, Congo red stain was used for detecting amyloid deposits by polarized microscopy. For IHC we used *GeneTex's* Anti-*ACE2 antibody* [*SN0754*], *Spike RBD antibody* [*HL257*] (*GTX635692*) and an antibody panel for amyloid typing.

Results: The patient was hospitalized to the ICU with severe fatigue, fever, difficulty breathing, dyspnea at rest, and 92% oxygen saturation. SARS-CoV-2 was detected by the real-time PCR method. Despite the provided intensive therapy, her condition deteriorated from the developed multiple organ dysfunction syndrome (MODS). The autopsy revealed that the death was caused by COVID-19-associated viral and bacterial pneumonia. An important postmortem finding was systemic AA amyloidosis with severe kidney alterations not identified during life. As the patient did not receive pathogenesis-based therapy, she developed a rapidly progressing renal failure, significantly aggravating the clinical pattern.

Conclusions: The case study demonstrated the need for personalizing treatment of COVID-19 in patients with RA and AA amyloidosis. These co-existing diseases can impair the immune system and cause MODS, contributing to the severity of COVID-19 and leading to lethal outcome.

Keywords: Rheumatoid Arthritis, COVID-19, amyloidosis

PV090 / #14

POSTER SESSION 09: COMORBIDITIES AND CLINICAL OUTCOMES IN AUTOIMMUNITY
03-08-2025 10:30 AM - 11:30 AM

CETP AND THE LIPID PARADOX IN RA PATIENTS

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Background and Aims: Rheumatoid arthritis (RA) has been associated with changes in the lipid pattern described as the lipid paradox. Active RA is characterized by increased cardiovascular risk despite decreased cholesterol levels. One of the main proteins regulating lipid metabolism and a potential player in the lipid paradox is cholesteryl ester transfer protein (CETP). The aim of this study was to explore potential associations between CETP activity and lipid changes, as well as disease activity in RA patients.

Methods: Ninety-seven RA patients and 97 matched healthy controls were enrolled in the study. Lipid concentrations were measured using enzymatic methods. CETP activity was determined using spectrofluorimetry. RA activity was evaluated using the DAS28-ESR.

Results: RA patients displayed significantly lower CETP activity than healthy controls ($p < 0,001$). CETP activity cut off point of 38,3 had high sensitivity (99-100%), specificity (93-98%) and accuracy (96-99%) in distinguishing between RA patients and controls ($AUC_{CETP} = 0,986$ (95% CI 0,971-1,000; $p < 0,001$). Correlations between CETP activity and total cholesterol concentration ($\beta = 0,4421$, $p < 0,001$); CETP activity and LDL-cholesterol concentration ($\beta = 0,4093$, $p < 0,001$), as well as CETP and non-HDL-cholesterol concentration ($\beta = 0,4295$, $p < 0,001$) were observed in the study group. CETP activity was found to be inversely correlated with DAS28-ESR ($p < 0,05$).

Conclusions: In patients with RA, disease activity and lipid metabolism abnormalities are associated with changes in CETP activity. CETP could be one of the key players on the crossroads between the lipid paradox and inflammation in patients with RA.

Keywords: CETP, RA, Lipid paradox

PV091 / #178

POSTER SESSION 09: COMORBIDITIES AND CLINICAL OUTCOMES IN AUTOIMMUNITY
03-08-2025 10:30 AM - 11:30 AM

LONGITUDINAL MORBIDITY TRAJECTORIES IN PATIENTS WITH RHEUMATOID ARTHRITIS VERSUS COMPARATORS WITHOUT RHEUMATOID ARTHRITIS

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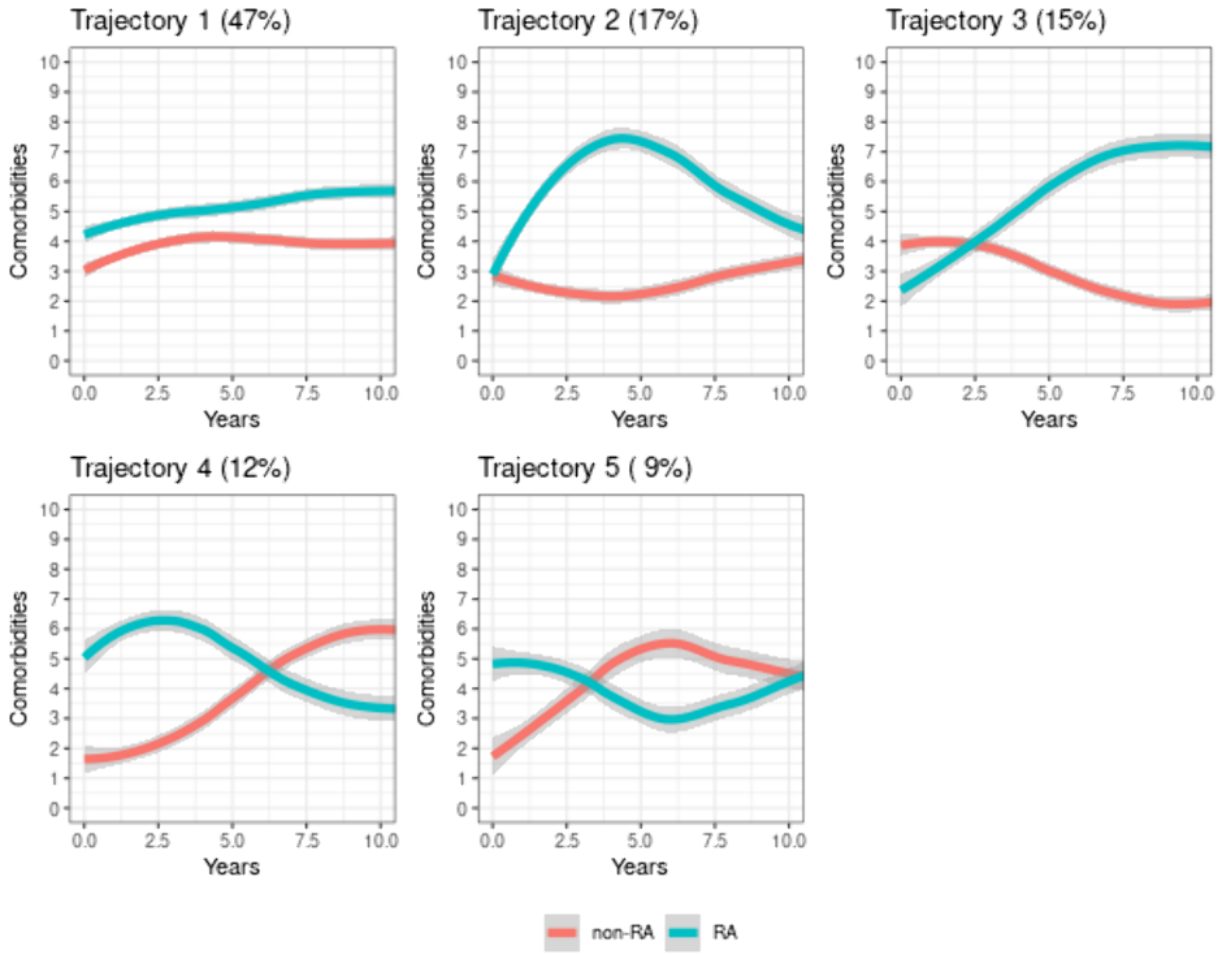
Background and Aims: Multimorbidity (2 or more morbidities) is more common in patients with RA. We determined longitudinal trajectories of morbidity among patients with RA compared to those without RA.

Methods: This population-based study included residents of a geographic area with incident RA in 1999-2019 who fulfilled ACR/EULAR criteria. Each RA patient was matched to a non-RA resident of the same age and sex. Multimorbidity was assessed yearly using 55 chronic conditions. The outcome was the difference in number of morbidities between the RA/non-RA pairs. Latent class mixture models were used to identify similar trajectories.

Results:



The study included 1233 patients with RA and 1233 matched comparators without RA (mean age 55.8 years, 70% female). The median (IQR) number of morbidities at incidence/index was 3 (1-6) in RA and 2 (0-4) in non-RA. We identified 5 trajectories (Figure). Trajectory 1 was the largest (n=585; 47%) with mean age slightly older than the others (57.8 vs 53.0-55.0 years). The number of patients was 207 (17%) in trajectory 2, 187 (15%) in trajectory 3, 146 (12%) in trajectory 4, and 108 (9%) in trajectory 5. There were no differences between the trajectories in sex, seropositivity, erosions, or antirheumatic medication use in the first year after index.

Conclusions: RA patients accumulate morbidities differently than their non-RA counterparts. Half of RA patients have 1 more morbidity on average than matched comparators. A third of RA patients have an accelerated accumulation of morbidities in the first 5 years that attenuates or plateaus subsequently.

Keywords: Rheumatoid Arthritis, comorbidity, multimorbidity

PV092 / #232

POSTER SESSION 09: COMORBIDITIES AND CLINICAL OUTCOMES IN AUTOIMMUNITY
03-08-2025 10:30 AM - 11:30 AM

AUTOIMMUNE-RELATED RENAL INVOLVEMENT IN DYSREGULATION, POLYENDOCRINOPATHY, ENTEROPATHY, X-LINKED (IPEX) SYNDROME: AN UPDATED SYSTEMATIC REVIEW

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Background and Aims: Immune dysregulation, polyendocrinopathy, enteropathy, X-linked (IPEX) syndrome is a systemic autoimmune genetic disorder caused by mutation of the forkhead box protein 3 (FOXP3) gene. We performed this study to analyze the clinical and demographic characteristics of renal involvement in IPEX patients, genotype-phenotype correlations and the effect of renal involvement on patient outcome.

Methods: We performed a literature search (Pubmed and EMBASE) to systematically investigate the case reports of IPEX with renal involvement which were published before Aug 31st, 2024.

Results: A total of 38 patients were identified. All IPEX patients included had FOXP3 mutations. Among 26 patients in whom detailed information is available, 23 showed single presentation (7 proteinuria, 9 nephrotic syndrome (nephrotic range proteinuria), 1 acute glomerulonephritis, 6 renal insufficiency), 3 showed multiple presentations (2 nephrotic range proteinuria, microscopic hematuria and 1 nephrotic range proteinuria, microscopic hematuria, renal insufficiency). Renal biopsy was documented in 20 patients. Membranous nephropathy (n=10) was most common, followed by Interstitial nephritis (n=3), membranoproliferative nephropathy (n=2), tubulopathy (n=2), mesangial proliferative glomerulonephritis (n=2), autoimmune nephritis (n=1). Nephrotic syndrome was more frequent in patients with intron 6 mutation ($p<0.05$). However, renal presentation of IPEX was not related to patient outcome (death).

Conclusions: We found renal involvement is one of the important autoimmune-related manifestations of IPEX syndrome and membranous nephropathy was most common. Close renal monitoring is essential to detect renal involvement in IPEX syndrome. Further studies are needed to compare the course of IPEX-related and unrelated

glomerulonephritis. Response to treatment in IPEX patients should also be elucidated in the future.

Keywords: Autoimmune, IPEX Syndrome, Renal involvement

PV093 / #311

POSTER SESSION 09: COMORBIDITIES AND CLINICAL OUTCOMES IN AUTOIMMUNITY
03-08-2025 10:30 AM - 11:30 AM

CLINICAL PROFILE AND OUTCOME OF CARDIAC SARCOIDOSIS A RETROSPECTIVE SINGLE CENTER STUDY FROM INDIA

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Background and Aims: Cardiac involvement in sarcoidosis is uncommon (<5%). Around one fourth of cardiac sarcoidosis (CS) patients have isolated cardiac Sarcoidosis (ICS). Aim is to characterize the clinical profile and outcome in patients with CS.

Methods: We retrospectively analyzed the clinical profile of 53 cases of CS patients at our unit. Sociodemographic, clinical manifestations, treatment and outcome of these patients were retrieved.

Results: CS was diagnosed by extracardiac biopsy proven granuloma (n=35) and rest by PET scan or late gadolinium enhancement in cardiac MRI. 23 patients had cardiac manifestations. The mean (SD) age of disease onset was 40.21 (11.8) years, age of presentation (SD) was 42.2 (11.8) years, diagnostic delay (SD) was 14.64 (8.4) months. Male to female ratio was 1.5: 1 (Table
1).

Table 1 : Clinical Features of cardiac sarcoidosis patients (n=53)

Age years (mean, SD)	42.9 (12.1)
Gender (M:F)	1.5: 1
Age of onset in years (mean, SD)	40.21 (11.8)
Age of presentation (mean, SD)	42.2 (11.8)
Diagnostic delay (mean, SD)	14.64 (8.4)
Mean follow up in months (SD)	26.7 (28.62)
Diabetes	7 (13.2%)
Systemic Hypertension	18 (34.6%)
Abnormal Chest Xray	14 (84.6%)
Constitutional symptoms	31 (58.4%)
Pulmonary involvement	32 (60.4%)
Skin	3 (5.7 %)
Joint involvement	8 (15%)
Eye	9 (17.5%)
Hepatosplenomegaly	13 (24.5%)
ESR mean SD (mm/Hr)	37 (27-55)
CRP mean SD (mg/L)	11 (4 – 14)
HbA1C (%)	5.7 (5.3-6.1)
Mantoux positivity	10 (18.9 %)
Hypercalcemia (> 10.5 mg/dL)	9 (17.5%)

Cardiac manifestations were palpitation (n=21, 39.6%), chest discomfort (n=19, 35.8%), syncope (n=9, 17%) and presyncope (n=8, 15%), heart failure with low ejection fraction (n=10, 18.8%), ventricular tachycardia (VT)(n=9, 17%), atrial fibrillation (n=6, 11.32%), left ventricular hypertrophy (n=7, 14.3%), and restrictive cardiomyopathy (RCMP) (n=6, 11.32%) (Table
2).

Table 2: Cardiac manifestations among CS patients

Any ECG change	23 (43.4%)
Sinus Tachycardia	1 (2%)
Sinus Bradycardia	1 (2%)
Atrial fibrillation (AF)	6 (11.32%)
Right Bundle Branch Block (RBBB)	4 (8.2 %)
Left Bundle Branch Block (LBBB)	1 (2%)
Left ventricular hypertrophy (LVH)	7 (14.3 %)
Ventricular Tachycardia (VT)	9 (16.98%)
Complete Heart Block (CHB)	4 (8.2%)
LV dysfunction EF< 50%	10 (18.8%)
Restrictive cardiomyopathy (RCMP)	6 (11.32%)
Restrictive cardiomyopathy with AF	3 (5.6%)
Restrictive cardiomyopathy with EF < 50 %	1 (2 %)
Pericardial effusion	3 (5.6%)
MRI Late gadolinium enhancement (LGE)	33 (61 %)
FDG PET uptake	48 (90.6)

Constitutional symptoms (n=9, 39% vs n=22, 73.3%), pulmonary (n=5, 22% vs n=27, 90%) and joint involvement, high ACE (67.3 vs 92.8, p<0.01) and high CRP (8.6 mg/L vs 16.2 mg/L, p<0.02) were more frequent in asymptomatic CS. During a mean follow-up of 26.7 months, 85% patients improved, 7.5% patients worsened. There were four mortality (3 ICS); VT (n=2), RCMP with AF (n=1), sepsis (n=1), all in symptomatic CS group.

Conclusions: CS can present as asymptomatic to life threatening ventricular tachycardia. Symptomatic CS and ICS have high risk of mortality.

Keywords: Sarcoidosis, cardiac, arrythmia

PV094 / #314

POSTER SESSION 09: COMORBIDITIES AND CLINICAL OUTCOMES IN AUTOIMMUNITY
03-08-2025 10:30 AM - 11:30 AM

MYASTHENIA GRAVIS IN INTERSTITIAL LUNG DISEASE DUE TO RHEUMATOID ARTHRITIS: A RARE COMORBIDITY TO CONSIDER.

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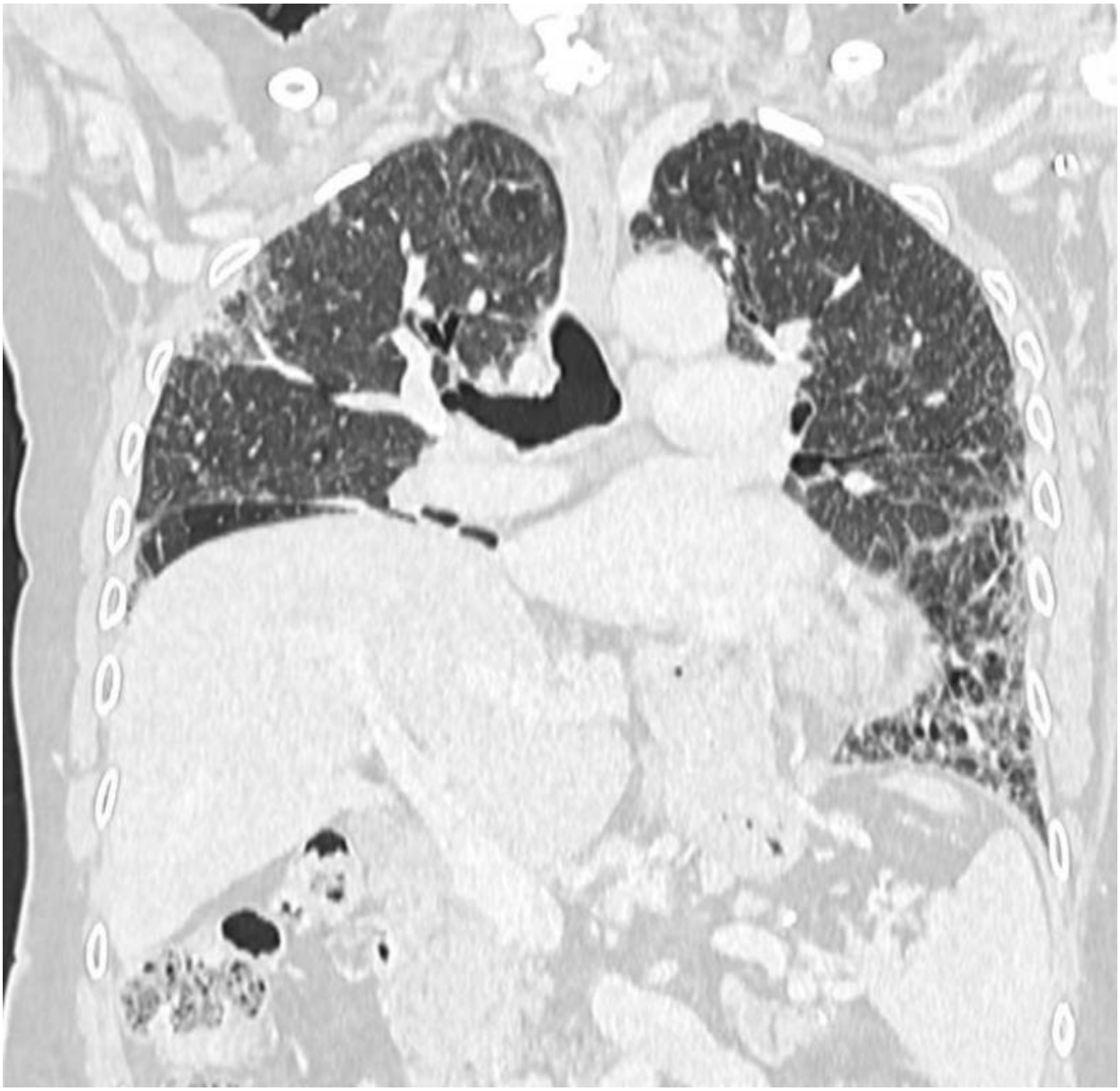
Background and Aims: Myasthenia gravis (MG) is an autoimmune disease that affects the neuromuscular junction of striated muscles. It is often associated with other autoimmune conditions and results from the direct attack of autoantibodies against the acetylcholine receptor (AChR). Patients with rheumatoid arthritis (RA) have an increased prevalence of MG (4%). Our aim is to explore the overlap between autoimmune diseases such as MG and RA, highlighting the potential common pathogenetic mechanisms.

Methods: A 73-year-old female with a history of quadrantectomy for invasive ductal carcinoma of the right breast (2012), non-smoker: in 2023, she was diagnosed with RA (RF+, ACPA+) and initially treated with Plaquenil. During follow-up, the patient developed significant exertional dyspnea. A chest CT revealed pulmonary fibrosis (indeterminate UIP). Plaquenil was prescribed at 200 mg on alternating days with 400 mg. The chest CT also showed marked elevation of the right hemidiaphragm [Fig.1]. Antibody testing for anti-AChR was positive. Measurements of maximal inspiratory pressures (MIP: 46 cmH2O) and maximal expiratory pressures (MEP: 62 cmH2O) confirmed the presence of neuromuscular disease.



[Fig.1]

Results: The patient was diagnosed with MG and specific treatment with pyridostigmine (60 mg twice daily) was initiated. Plaquenil was discontinued. The lack of radiological progression did not justify antifibrotic treatment and dyspnea was considered secondary to MG. For RA rheumatologists recommended biological therapy (abatacept).

Conclusions: The association between MG and RA is a rare condition that should be considered, particularly when RA-associated pulmonary fibrosis does not explain the dyspnea. Evaluating the diaphragms is crucial in identifying concomitant neuromuscular pathology, alongside the rheumatological process.

Keywords: myasthenia, Rheumatoid Arthritis, diaphragm

PV095 / #318

POSTER SESSION 09: COMORBIDITIES AND CLINICAL OUTCOMES IN AUTOIMMUNITY
03-08-2025 10:30 AM - 11:30 AM

THE ROLE OF KARYOTYPE IN AUTOIMMUNE PATHOLOGIES: A CASE OF BULLOUS PEMPHIGOID AND COMORBIDITIES

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Background and Aims: Bullous pemphigoid is an autoimmune disorder characterized by the formation of subepidermal blisters, significantly impacting patients' quality of life. The diagnosis and management of this condition are often complicated by the presence of comorbidities. This study specifically explores the association between Klinefelter Syndrome (KS), genetically defined by the presence of an extra X chromosome (47,XXY), and susceptibility to bullous pemphigoid, a connection that has been insufficiently explored in the literature.

Methods: A 57-year-old male presented with a widespread skin eruption, characterized by tense blisters on an erythematous base and post-lesional ulcers, some covered by hemorrhagic crusts. Additionally, nasal mucosa involvement was observed. The diagnosis of bullous pemphigoid was confirmed histopathologically by the identification of subepidermal blisters and neutrophilic microabscesses. Abnormal hormonal tests(decreased testosterone production), along with symptoms of hypogonadism, gynecomastia, reduced muscle mass and sparse facial and body hair suggested a possible diagnosis of KS, prompting genetic investigations that are still ongoing. We could not prove the infertility. The treatment consists in systemic corticosteroids with good results.

Results: While KS is known to be associated with an increased predisposition to various autoimmune diseases, the specific link between KS and bullous pemphigoid remains poorly documented. Possible role of low testosterone and altered estrogen levels can impact immune system regulation for developing the autoimmunity.

Conclusions: This integrative approach promotes a deeper understanding of the interactions between abnormal karyotype and autoimmune pathologies, facilitating the development of more effective treatments.

Keywords: Autoimmunity, bullous pemphigoid, Klinefelter Syndrome

PV096 / #354

POSTER SESSION 09: COMORBIDITIES AND CLINICAL OUTCOMES IN AUTOIMMUNITY
03-08-2025 10:30 AM - 11:30 AM

A CASE OF RHEUMATOID ARTHRITIS IN A PATIENT WITH HASHIMOTO'S THYROIDITIS, A COMMON COEXISTENCE.

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Background and Aims: Hashimoto hypothyroidism is not a rare condition and the autoimmune etiology is already well known. There are considerable references for the coexistence of HT, rheumatoid arthritis, osteoarthritis, fibromyalgia, polymyositis, and other connective tissue diseases. We present a case with HT under treatment for hypothyroidism which overlaps rheumatoid arthritis.

Methods: The patient was diagnosed with HT many years before, she was under treatment with Levothyroxine 100 mg daily. Her mother was diagnosed with HT and fibromyalgia too. Suddenly she started complaining of fatigue, low-grade fever, morning stiffness, and joint pain, especially in her hands, knees, and both elbows and swollen hands. The first tests showed a high value of erythrosedimentation, anemia, and a high value of PCR. The immunological tests confirmed the presence of a high titer of positive FR(rheumatoid factor), anti-CCP positive, and normalized thyroid function with free thyroxine (free T4) and, thyroid-stimulating hormone (TSH) under normal range, but with elevated values of Anti TPO, and ANA(anti-nuclear antibodies) positive, Extractable Nuclear Antigen (ENA) screening negative. The diagnosis of RA with HT was confirmed. The patient showed significant improvements within four weeks after the therapy with immunosuppressants and corticosteroids.

Results: . The correlation between hashimoto thyroiditis and rheumatoid arthritis or another connective tissue disease is not well documented, but it is seen very often the coexistence of both diseases.

Conclusions: Still, at the same time, we believe there is a link between different presentations of the immune system underlying the "mosaic of autoimmunity".

Keyword: hashimoto thyroiditis, rheumatoid arthritis, autoimmunity, disease activity

PV097 / #358

POSTER SESSION 09: COMORBIDITIES AND CLINICAL OUTCOMES IN AUTOIMMUNITY
03-08-2025 10:30 AM - 11:30 AM

HAND ABILITY EVALUATION IN SYSTEMIC SCLEROSIS - SYSTEMATIC REVIEW

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Background and Aims: Background: Patients with systemic sclerosis (SSc) often experience impaired hand function. However, many assessment methods are developed. The aim of this systematic review was to identify and review the tools used in evaluation of hand ability in SSc while assessing pharmacotherapy interventions.

Methods: Materials and methods: This review was performed according to PRISMA (Preferred Reporting Items for Systematic Reviews and Meta-Analyses) guidelines. Medline and Embase databases were searched for studies that investigated hand ability in SSc participants. Included studies were intervention studies, clinical trials and randomized controlled trials, assessing pharmacotherapy or invasive procedures effect versus control using placebo or no procedure/standard care. Cochrane risk-of-bias tool was used to assess methodological quality.

Results: 2806 results were retrieved, while only 11 studies were included according to inclusion criteria. Quality assessment indicated all studies had low risk of bias. Included studies used hand-specific self-reported questionnaires (Disabilities of the Arm, Shoulder and Hand, Cochin Hand Function Scale (CHFS), modified Hand Mobility in Scleroderma, Kapandji test, grip strength and extension index) and general questionnaires (visual analogue scale (VAS) pain, Scleroderma Health Assessment Questionnaire (SHAQ), Health Assessment Questionnaire Disability Index). Nine studies used 1 hand-specific test (predominantly CHFS) combined with one or two general questionnaire/scale (most often VAS pain or SHAQ), while other two added one more hand-specific questionnaire.

Conclusions: Conclusion: A need exists for systematic evaluation and comparison of all questionnaires/scales developed. However, using a combination of hand-specific and general evaluation in terms of self-reported questionnaires, as well as direct grip strength measurement is generally recommended.

Keywords: systematic review, systemic sclerosis, Hand function

PV098 / #401

POSTER SESSION 09: COMORBIDITIES AND CLINICAL OUTCOMES IN AUTOIMMUNITY
03-08-2025 10:30 AM - 11:30 AM

THE TWO MOST FREQUENT COMORBID AUTOIMMUNE DISEASES IN CHILDREN WITH TYPE1 DIABETES MELLITUS IN ALBANIA

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Background and Aims: Type 1 diabetes mellitus (T1 DM) is the most common type of diabetes in children. T1DM patients are also at higher risk of other comorbid autoimmune diseases, including autoimmune thyroid disease (AITD), celiac disease (CD). The thyroid-specific immune damage of AITD is strongly associated with elevated serum thyroid peroxidase (TPO). Tissue transglutaminase antibody (tTGA) is a specific antibody and a serological marker of CD.This study aimed to evaluate the positivity of anti - TPO and anti - tTGA in children with T1DM after they were diagnosed.

Methods: This study has included 105 children with T1DM.44 children with other diagnoses were taken as control. Anti - TPO and anti - tTGA were carried out by ELISA.

Results: 55.2% of T1DM children were girls. The anti-TPO was positive in 30.5% of T1DM children compared to 4.5% of children in control group. The anti-tTGA was positive in 7.6% of T1DM children compared to 2.3% of children in control group. Risk of Hashimoto's hypothyroidism was more in children older than 10 years old. 21.9% of children 11 - 14 years old were anti - TPO positive, but it was 16.2%, more common in girls. While, anti - tTGA was positive in 3.85% of children 1 - 5 years old with no difference between boys and girls.

Conclusions: The most frequent autoimmune disease resulted Hashimoto's hypothyroidism. Children with T1DM were found to have a lower predisposition to CD. In conclusion we can say that antibodies to other autoimmune diseases must be performed together with diagnostic examinations for T1DM.

Keywords: Celiac Disease, Type 1 diabetes mellitus, Autoimmune Thyroid Disease

PV099 / #498

POSTER SESSION 09: COMORBIDITIES AND CLINICAL OUTCOMES IN AUTOIMMUNITY
03-08-2025 10:30 AM - 11:30 AM

EVALUATION OF HAND ABILITY IN PHYSICAL THERAPY OF SYSTEMIC SCLEROSIS PATIENTS - SYSTEMATIC REVIEW

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Background and Aims: Hand function is frequently compromised in patients with systemic sclerosis (SSc). Finding and reviewing the instruments used to evaluate hand ability in SSc while evaluating physical therapy was the goal of this systematic review.

Methods: The PRISMA (Preferred Reporting Items for Systematic Reviews and Meta-Analyses) standards were followed. Studies that examined hand ability in SSc participants were found using Medline and Embase databases. Intervention studies, clinical trials, and randomized controlled trials evaluating the effects of physical therapy (and all variations) in comparison to controls having no procedure/standard care were included. The methodological quality was evaluated using the Cochrane risk-of-bias technique.

Results: Out of the 2806 results that were obtained, 20 studies met the inclusion criteria. Quality assessment showed low to moderate risk of bias. Included studies used general (visual analogue scale (VAS) pain, Scleroderma Health Assessment Questionnaire (SHAQ), Health Assessment Questionnaire (HAQ) Disability Index) and hand-specific self-reported questionnaires (Cochin Hand Function Scale (CHFS), Disabilities of the Arm, Shoulder and Hand, modified Hand Mobility in Scleroderma (HAMIS), Kapandji test, grip strength, pinch test and extension index). Most of the studies included one or two hand-specific (predominantly CHFS or HAMIS) combined with one or two general questionnaire/scale (most often VAS pain or HAQ/SHAQ), while one study used range of motion measurment.

Conclusions: There is a need for systematic evaluation of scales, tests and questionnaires used for assessment od hand ability in SSc. However, looking at selected studies, use of combination of tests, scales and questionnaires generally increases the quality of evaluation in regard to physical therapy effect.

Keywords: systematic review, systemic sclerosis, Hand function

PV100 / #515

POSTER SESSION 09: COMORBIDITIES AND CLINICAL OUTCOMES IN AUTOIMMUNITY
03-08-2025 10:30 AM - 11:30 AM

THE COMORBIDITIES AND DISEASE FEATURES IN YOUNG WOMEN WITH SLE AND HYPERURICEMIA.

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Background and Aims: Systemic lupus erythematosus (SLE) is a chronic autoimmune disease affecting various organs, mainly in young women. Hyperuricemia (HU) is more common in men and postmenopausal women. There is not enough data on the impact of HU on young women with SLE. The aim is to compare the disease features and comorbidities in young women with SLE and HU.

Methods: 99 women with SLE age 18-45 were involved: 46 with SLE and HU (huSLE) and 53 SLE women with normouricemia (nuSLE). Comorbid status was assessed by Charlson comorbidity index, QRISK ® 3. The demographic and clinical data were studied.

Results: huSLE were older (34.49±7.55 years against 29.56±7.18, p=0,000003), has the same disease activity (SLEDAI2K 5,5±4,87 against 5,06±3,63, p=0,87). Disease duration was 12,37±6,42 and 7,85±7,23 years, p=0,00013. A high incidence of serosites (32.6% against 7.5%, p=0,03) and thrombocytopenia (13% agains 6,5%, p=0,064) was revealed. HuSLE received lower steroid dose (30 [20;40] mg against 50 [30; 60] mg per day, p=0,006). Despite the absence of significant differences in disease features and comorbidities (LN, CKD, arterial hypertension, CAD, stroke, diabetes, etc., CCI 1,46±0,99 against 1,36±0,86, p=0,45), the risk of cardiovascular events (QRISK3) in the huSLE group was higher (4.4% [0; 17] against 1.9% [0; 13], p=0,00005).

Conclusions: Patients with huSLE and nuSLE has comparable disease activity despite that patients with nuSLE receive large doses of GC. Frequent occurrence of serosites in huSLE was revealed, a tendency to thrombocytopenia in this group was noted. Significant differences in the risk of cardiovascular events in huSLE have been revealed.

Keywords: systemic lupus erythematosus, Cardiovascular Disease, hyperuricemia

PV101 / #528

POSTER SESSION 09: COMORBIDITIES AND CLINICAL OUTCOMES IN AUTOIMMUNITY
03-08-2025 10:30 AM - 11:30 AM

FULL RECOVERY FROM LUPUS NEPHRITIS WITH NO SIGNS OF CKD. FOCUSED ANALYSIS

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Background and Aims: . It is not well-defined how common is the development of chronic kidney disease (CKD) in patients with long-term LN.

Methods: Patients and methods Two hundred and sixty biopsy-proven LN patients have been followed for a median of 168.5 (50-287.25) months. At last observation: -- 74 patients (28.5%), after a median follow-up of 148 months (76.4-237), no signs of CKD, based on KDIGO definitions, were present. In these patients, eGFR was >90ml/min/1.73m2, proteinuria <150mg/day, and no red blood cells in urinary sediment were present for the last 110.46 (66.74-190.74) months of the observation. we have compared, with logistic regression analysis, the basal characteristics of these “super-responders” with those of the other LN patients.

Results: Among baseline kidney variables, super-responder patients had significantly lower serum creatinine, higher eGFR, and fewer red blood cells in urinary sediment than patients who did not achieve super response. At one year super-responders had a higher number of complete or complete plus partial responses than the other group. At multivariate analysis, eGFR at LN diagnosis (OR:0.9851,CI:0.9747-0.9956,P 0.0054), the chronicity index at kidney biopsy (OR:1.3144,CI:1.0490-1.6468,P=0.0175) and complete remission at one year after the start of the induction therapy (OR0:4.1500,CI:1.9142-8.9971,P=0.0003) were the independent factors associated with the achievement of super-response in LN.

Conclusions: Our results show that about 50% of patients with LN followed in our center still have normal glomerular filtration rates after an observational period of about 15 years and that half of them do not have any altered variables that may allow the diagnosis of CKD.

Keyword: lupus nephritis, systemic lupus erythematosus, chronic kidney disease

PV102 / #571

POSTER SESSION 09: COMORBIDITIES AND CLINICAL OUTCOMES IN AUTOIMMUNITY
03-08-2025 10:30 AM - 11:30 AM

WHAT WE DON'T TALK ABOUT: FAMILY, FRIENDS, INTIMACY IN CASES OF ARTHRITIS

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Background and Aims: "Health is a state of complete physical, mental and social well-being and not the absence of disease or infirmity." Social health is the totality of developing relationships with others. The effective therapy of arthritides allows to aim not only the disease controll but also at improving the quality of life. Taboo topics such as social and sexual relationships are an integral part of this.

Methods: Using our self-edited questionnaire, we assesed the marital status, number of children, social relationships within the family, social life, friendships, work ebility, sexual habits among patients with rheumatoid and psoriatic arthritis.

Results: Oh the 100 qustionnaries distributed, 62 were suitable for processing. Ninty percent of our patients live in a relationship, Childbearing is lower among them than in the average population. The rheumatology disease causes relationship problems. Twenty three percent of our pateints believe that their social life and friendships changed after the diagnosis of arthritis. Their ability to work has changed for 33% of them, 27% of them feel the disadvantage of their illness at work. Eighty percent of pateints do, 20% do not have an active sexual life. After the diagnosis of arthritis 37% did, 67% did not experience a change in their sexual habits.Fourty seven percent of patients indicated sexual disorder: biological or psychological in origin.

Conclusions: The issue of family planning and childbearing is unavoidable in the daily care of arthritis pateints, but social relationships and sexulity are still taboo topics that we find difficult to discuss with our patients.

Keywords: social relationships, arthritis, sexuality

PV103 / #154

POSTER SESSION 10: SLE, ILD AND NOVEL THERAPEUTIC TARGETS
03-08-2025 10:30 AM - 11:30 AM

DISCOVERY AND CHARACTERIZATION OF MIMOTOPES FOR ANTI-C1Q ANTIBODIES IN SYSTEMIC LUPUS ERYTHEMATOSUS

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Background and Aims: Background and aims Systemic Lupus Erythematosus (SLE) is a complex autoimmune disease, which can develop into severe forms. Essential tolerance mechanisms are increasingly affected during worsening of the disease and anti-double-stranded (ds)DNA antibodies are accused to be involved in the pathologies. They react with nucleic acids but are not strictly specific to it. Via recognizing non-DNA antigens such as the complement component 1 (C1q), inflammatory responses, apoptosis and tissue fibrosis can be triggered. Here we aimed to identify peptides that function as blocking mimotopes for anti-dsDNA autoantibodies and subsequently validate a possible harmful T-cell modulating effect of the mimotopes.

Methods: Methods To identify epitopes of autoantibodies, we designed a peptide microarray that contains non-nucleic acid targets of anti-dsDNA autoantibodies. The microarray contains 4309 overlapping peptides (13 amino acids, overlap 10 amino acids), including 169 peptides of C1q subunit A and B. The microarrays were applied to map the IgG antibody response of 6 SLE patients in comparison to 6 healthy donors. Promising peptides were tested for the specific blocking of antibodies in their soluble form and validated for T-cell activation via ELISpot assays.

Results: Results We observed a significant elevated IgG response in the serum of SLE patients against three peptide sequences in C1q alpha and beta subunit. The soluble mimotopes blocked antibody-binding epitope-specific and showed no alarming activation of effector T-cells.

Conclusions: Peptide microarrays are a powerful tool to investigate the misdirected humoral immune response in autoimmune diseases and can support the development of peptide-based therapeutic strategies.

PV104 / #163

POSTER SESSION 10: SLE, ILD AND NOVEL THERAPEUTIC TARGETS
03-08-2025 10:30 AM - 11:30 AM

CM-101 AS A NOVEL THERAPY TARGETING CCL24-INFLUENCED IMMUNE CELL POPULATIONS IN SYSTEMIC SCLEROSIS

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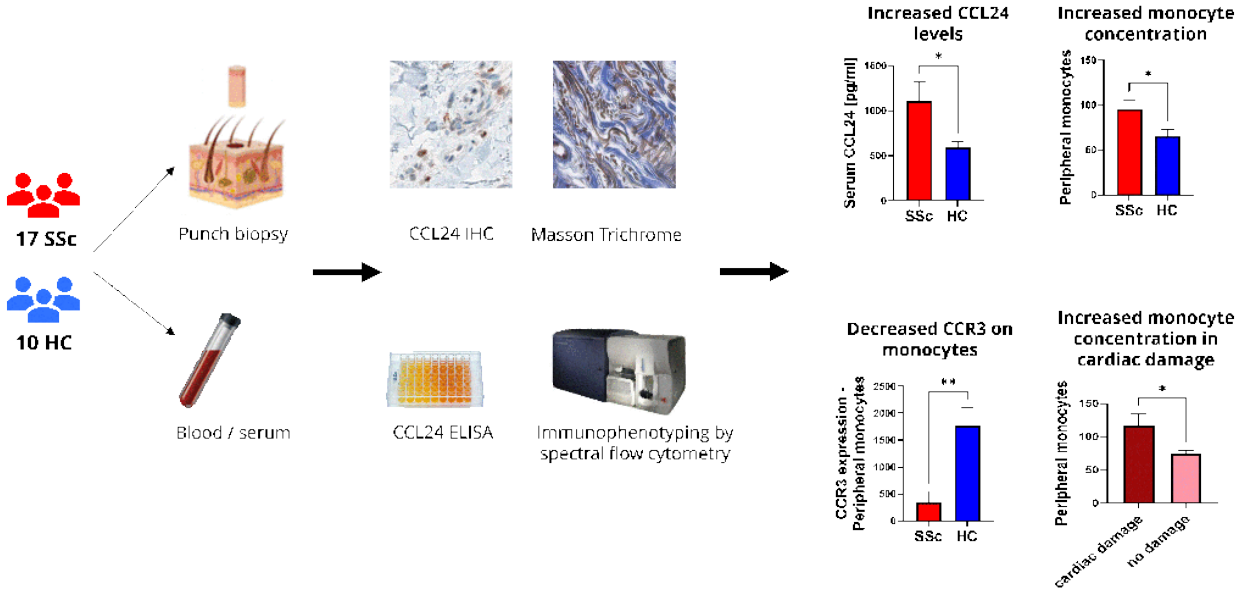
Background and Aims: Systemic Sclerosis (SSc) is a severe autoimmune disorder characterized by skin and organ fibrosis. CCL24, a profibrotic chemokine, is linked to SSc progression. Elevated serum CCL24 levels correlate with disease severity and mortality. Blocking CCL24 with CM-101 reduces fibrosis and inflammation in SSc preclinical models. We present data demonstrating CM-101's therapeutic potential and its possible effects on CCR3-expressing immune cells.

Methods: Changes in bronchoalveolar lavage (BAL) fluid following CM-101 treatment were assessed in a bleomycin-induced fibrosis mice model. CCL24 expression and collagen deposition were assessed in skin tissue biopsies from healthy controls (HC) and SSc patients. Peripheral blood cells from HC and SSc patients were classified by spectral flow cytometry (Cytek® 25-Color Immunoprofiling Assay). Serum and plasma CCL24 levels were quantified by ELISA.

Results: In bleomycin-treated mice, CM-101 significantly reduced mononuclear cell counts in BAL fluid. In SSc patients, elevated serum CCL24 levels were associated with increased skin CCL24 expression. Blood immunophenotyping revealed specific changes in SSc patients, including increased neutrophils, plasmacytoid dendritic cells (pDC) and monocytes, whereas innate lymphoid and T cell concentrations decreased. Interestingly, altered CCR3 expression was observed in these populations. pDC and monocyte concentrations correlated with SSc severity, inflammation activity and organ fibrosis, as measured by joint contractures, DLCO, heart involvement, serum ferritin concentration and gastrointestinal complications.

Conclusions: This study sheds light on CCL24's role in bleomycin-induced fibrosis and SSc pathogenesis. We identified two immune cell populations with altered CCR3 expression linked to SSc, underscoring the role of CCL24 in SSc and the therapeutic

potential of targeting CCL24.



Keywords: Chemokines, systemic sclerosis, Biomarkers

PV105 / #202

POSTER SESSION 10: SLE, ILD AND NOVEL THERAPEUTIC TARGETS
03-08-2025 10:30 AM - 11:30 AM

TREATMENT OF INTERSTITIAL PNEUMONIA WITH AUTOIMMUNE FEATURES (IPAF), SYSTEMIC REVIEW

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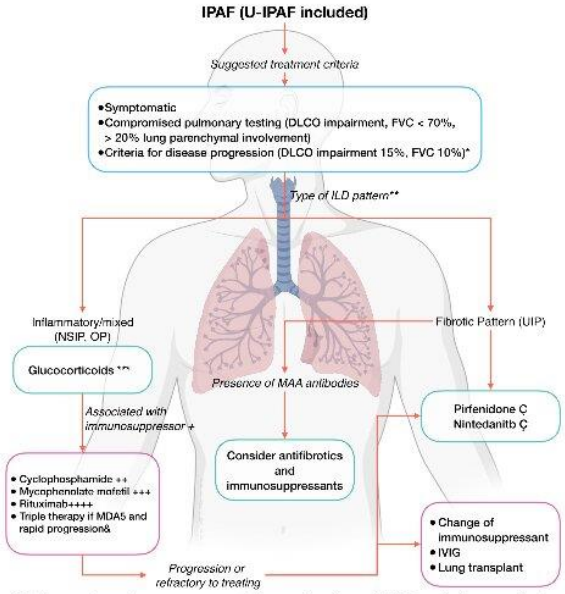
Background and Aims: Interstitial Pneumonia with Autoimmune Features (IPAF) is a subset of Interstitial Lung Disease (ILD) characterized by autoimmune features without fulfilling connective tissue disease (CTD) criteria. Despite the 2015 ERS/ATS classification, optimal therapeutic strategies remain unclear. This systemic review evaluates treatment in IPAF patients and identifies key gaps in clinical management.

Methods: A systematic review, registered with PROSPERO (CRD42023443823), was conducted using PubMed and EMBASE databases up to December 2022. We included cohort studies, case series, and clinical trials assessing IPAF treatments. Data were extracted on treatment modalities, lung function outcomes (FVC, DLCO), clinical domains, and comorbidities. Study quality was assessed using Joanna Briggs Institute tools

Results: 32 studies, encompassing 1500 patients (62.8% female, mean age 57.4 years), were included. Glucocorticoids were the primary treatment (69.4% of patients), followed by mycophenolate mofetil (9.7%) and cyclophosphamide (2.7%). Patients with non-specific interstitial pneumonia (NSIP) showed a mean FVC improvement of 8-10% with immunosuppressive therapy. Conversely, those with usual interstitial pneumonia (UIP) had poorer outcomes, with only minimal FVC gains (<3%), resembling idiopathic pulmonary fibrosis (IPF). Smoking (27%) and comorbidities such as hypertension (30%) and gastroesophageal reflux disease (27%) were common and impacted prognosis. Based on these results, we propose a diagnostic algorithm (Figure

1).



Conclusions: IPAF treatment responses are highly variable, with immunosuppressants being more effective in no fibrotic patterns. Antifibrotics may benefit UIP patients. There remains a significant need for randomized controlled trials to standardize IPAF treatment and address variability in clinical, serologic, and morphologic responses.

Keywords: autoimmune diseases, Interstitial Lung Diseases, Immunosuppressive Therapy

PV106 / #271

POSTER SESSION 10: SLE, ILD AND NOVEL THERAPEUTIC TARGETS
03-08-2025 10:30 AM - 11:30 AM

THE ONSET OF COMMON VARIABLE IMMUNODEFICIENCY AFTER RITUXIMAB INFUSION IN A PATIENT WITH SYSTEMIC LUPUS ERYTHEMATOSUS AND ABSOLUTE IGA DEFICIENCY

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Background and Aims: Selective IgA deficiency (SIgAD) is a common primary immunoglobulin production disorder. It is associated with autoimmune diseases, including systemic lupus erythematosus (SLE). The association of SIgAD with SLE is relevant due to the high morbidity and mortality rates of infections in patients with SLE. Rituximab is often used to treat refractory SLE but can induce hypogammaglobulinemia and increase the risk of recurrent infections, potentially unmasking an underlying common variable immunodeficiency (CVID).

Methods: In December 2023, a 37-year-old woman with SLE, with hemolytic anemia, pericarditis and arthritis, who also had a childhood history of SIgAD, came to our attention. At her initial visit, she exhibited severe hypogammaglobulinemia (IgG 265, IgA 5, IgM 15 mg/dL), first identified after Rituximab treatment in 2015. She reported recurrent upper respiratory infections and frequent antibiotic use. Flow-cytometric analysis showed a deficiency in B cell maturation. Genetic analysis is underway.

Results: The patient started replacement therapy with intravenous immunoglobulin at 20 g every three weeks. She was later transitioned to subcutaneous immunoglobulin 2 g every three days using the manual push technique. Following treatment, she has not experienced further infections and has achieved good IgG levels (IgG 792 mg/dl).

Conclusions: We aim to highlight the need for caution when administering immunosuppressive drugs, particularly Rituximab, in patients with absolute SIgAD. The use of these medications could potentially unmask a primary antibody deficiency.

Keywords: Common Variable Immunodeficiency, Rituximab in refractory SLE, Selective IgA deficiency (SIgAD)

PV107 / #424

POSTER SESSION 10: SLE, ILD AND NOVEL THERAPEUTIC TARGETS
03-08-2025 10:30 AM - 11:30 AM

NINTEDANIB IN THE TREATMENT OF INTERSTITIAL LUNG DISEASE IN SJÖGREN'S SYNDROME – A CASE REPORT

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Background and Aims: The incidence of pulmonary lesions associated with primary Sjögren's syndrome ranges from 9 to 24%, and involvement of the respiratory system is the main cause of mortality in these patients. The optimal regimen for the treatment of interstitial lung disease associated with Sjögren's syndrome (pSS-ILD) has not yet been defined. Glucocorticoids and immunosuppressive drugs are most commonly used in the therapy of active pSS-ILD lesions. In recent years, antifibrotic drugs have emerged for chronic pSS-ILD. The last line of treatment for pSS-ILD patients is lung transplantation.

Methods: In this study, we present a case of a 46-year-old man diagnosed with pSS-ILD and treated with nintedanib.

Results: The initial symptoms were cough and exertional dyspnea, which were preceded by symptoms of sicca. Rheumatological investigation was initiated due to the diagnosis of ILD of unclear etiology one year after the onset of symptoms. The patient was diagnosed with primary Sjögren's syndrome based on the presence of typical p/SS-A antibodies and a positive Schirmer test. He was initially treated with glucocorticosteroids, mycophenolate mofetil and cyclophosphamide. Eight months from diagnosis, nintedanib was added, slowing down disease progression, but due to respiratory failure, the patient was started on home oxygen therapy. He is currently on an active waiting list for lung transplant.

Conclusions: Learning points for clinical practice 1. ILD may be the first manifestation of pSS. 2. pSS-ILD is burdened with delayed diagnosis. 3. Nintendanib may be used as a bridge therapy slowing down pSS-ILD progression in patients who will require a lung transplant.

Keywords: Sjögren syndrome, interstitial lung disease, nintedanib

PV108 / #464

POSTER SESSION 10: SLE, ILD AND NOVEL THERAPEUTIC TARGETS
03-08-2025 10:30 AM - 11:30 AM

RAISING ANTI-PHBSPHORYLCHOLINE ANTIBODIES THROUGH VACCINATION TO PROTECT AGAINST AUTOIMMUNITY, ATHEROSCLEROSIS AND CHRONIC INFLAMMATION?

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Background and Aims: Chronic inflammatory conditions is a major health issue, obviously including rheumatic diseases but also several other common conditions. atherosclerosis which is a major cause of cardiovascular disease (CVD). Further to this, atherosclerosis and CVD are increased in several rheumatic diseases, where SLE is a striking example. In addition to these conditions, also dementia, osteoarthritis and obesity are related to chronic inflammation. Even in cancer, to a varying degree, inflammation may play an important role. Phosphorylcholine (PC) is a small lipid-related hapten, which is both a danger associated molecular pattern (DAMP), and a pathogen associated molecular pattern (PAMP). Antibodies against PC (anti-PC) are ubiquitous constituting 5-10% of circulating IgM ,

Methods: We combined cohort studies including nested case cohort studies, with experimental studies, ex vivo and in vivo, and also in silico.

Results: especially IgM and IgG1 anti-PC, is associated with protection in the chronic inflammatory conditions mentioned above. Anti-PC is almost absent at birth, in contrast to other natural antibodies like those against malondialdehyde, and increase during the first years of life. Associations were especially strong in SLE among rheumatic diseases and also in CVD in many studies, where associations are independent of and often much stronger than established risk factors. Animal experiments with immunization to raise anti-PC ameliorate atherosclerosis and other chronic inflammatory conditions. Potential mechanisms include anti-inflammatory, immune modulatory, clearance of dead cells and protection against infectious agents.

Conclusions: Anti-PC levels could be a new risk marker and raising anti-PC through immunization, may prevent and ameliorate chronic inflammation.

Keyword: antibodies, vaccination, atherosclerosis, autoimmunity, Phosphorylcholine

PV109 / #500

POSTER SESSION 10: SLE, ILD AND NOVEL THERAPEUTIC TARGETS
03-08-2025 10:30 AM - 11:30 AM

UNVEILING THE TRANSFORMATIVE POWER OF SPIRITUAL DISPOSITION GROUP INTERVENTION IN ALLEVIATING STRESS AND ENHANCING COPING STYLES AMONG SYSTEMIC LUPUS ERYTHEMATOSUS PATIENTS FROM INDIA.

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Background and Aims: Systemic lupus erythematosus (SLE) is a chronic systemic rheumatic autoimmune disorder wherein the body's immune cells start attacking vital organs and connective tissues. Patients who are diagnosed with SLE are faced with a lot of psychological challenges. The study is based on a theoretical framework of biopsychosocial-spiritual models for holistic person-centered care.

Methods: A novel spiritual dispositional intervention was developed and validated by experts in the field. The holistic spiritual intervention combining spiritual practices and Indian classical music therapy, known as Raga Chikitsa. Using an experimental pretest and posttest-design, recruited 30 participants (female = 28 and male = 2) diagnosed with SLE for the past 6 months and randomly assigned them to the experimental and control waitlist groups for the 8 weeks online intervention.

Results: An independent T test revealed significant mean differences between the groups in stress ($t = -5.25$, $p = <.001$), problem-focused coping ($t = 2.55$, $p = 0.016$), and emotional-focused coping ($t = 4.37$, $p = <.001$). There was no significant difference in the avoidant style of coping between the groups. The paired t test reveals significant differences in stress ($t = 4.61$, $p = .001$), avoidant coping ($t = 2.88$, $p = 0.012$), problem-focused coping ($t = -3.46$, $p = .004$), and emotional-focused coping ($t = -3.46$, $p = 0.007$) .

Conclusions: The research concedes that the spiritual disposition intervention was successful in reducing stress and improving coping styles post intervention. The intervention is focused on whole person-centered care and can be used for managing other chronic diseases as well.

Keyword: spirituality, health, systemic lupus erythematosus, behavioral medicine, stress, coping styles

PV110 / #549

POSTER SESSION 10: SLE, ILD AND NOVEL THERAPEUTIC TARGETS
03-08-2025 10:30 AM - 11:30 AM

MACROPHAGES FROM SYSTEMIC LUPUS ERYTHEMATOSUS PATIENTS EXHIBIT A THROMBOINFLAMMATORY PHENOTYPE

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Background and Aims: Thrombosis in SLE remains a major cause of morbidity and mortality in these patients. Inflammation in SLE is driven by interferon (IFN)α which is key to aberrantly activating immune cells, including macrophages. By examining the cross-talk between macrophages with the coagulation cascade, we aim to uncover critical insights into the complex network of interactions that lead to thromboinflammation in SLE.

Methods: Peripheral blood was collected from consented SLE patients at St. James's Hospital and age-and-sex matched healthy controls (HC). Monocytes were differentiated into macrophages using plastic adherence in 10% human serum for 7 days. On day 7, macrophages were either stimulated with LPS (10ng/ml) or IFNα (100U/ml) for 3h after which the thrombin generation assay (TGA) was used to assess the thrombin production over time.

Results: Unstimulated macrophages from SLE patients not only produced significantly higher concentrations of thrombin, but released higher concentrations into the supernatant compared to HC. Although the time to release thrombin (lagtime) was the same in SLE and HC macrophages, the velocity index suggests that a higher concentration of thrombin over a shorter amount of time was released from SLE macrophages at rest.

Conclusions: These results show for the first time that SLE macrophages at rest, contribute to the thrombotic state observed in SLE and that these macrophages are likely proinflammatory which directly influences this myeloid cell-driven hypercoagulability. This data opens new avenues for assessing thrombosis in SLE as well as puts forward a new contributor to SLE pathogenesis.

Keywords: SLE, Macrophages, thrombin

PV111 / #569

POSTER SESSION 10: SLE, ILD AND NOVEL THERAPEUTIC TARGETS
03-08-2025 10:30 AM - 11:30 AM

THE POTENTIAL ROLE OF INTRAVENOUS IMMUNOGLOBULIN FOR THE THERAPY OF RECURRENT ABORTION IN PATIENTS WITH IGG SUBCLASS DEFICIENCY

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Background and Aims: Recurrent pregnancy loss remains an important reproductive health concern in women. There are previous reports of IgG subclass deficiency as a possible cause of pregnancy loss. IgG subclass deficiency is amenable to therapy with intravenous immunoglobulins. We describe a case of IgG subclass deficiency treated with IVIG during pregnancy.

Methods: This is a 29-year-old woman who had had four miscarriages and no children. Clinical history revealed asthma and recurrent bacterial respiratory infections. Immunological studies performed to identify potential causes of recurrent pregancy loss detected low levels of IgG2 (230 mg/dL) and IgG4 (3.6 mg/dL). The level of anti-pneumococcal antibodies was low (0.65 mg/dL). The percentage of NK cells (8%) was normal. The patient did not have antiphospholipid antibodies. The levels of total IgG (885 mg/dL), C3 (138 mg/dL) and C4 (40 mg/dL) were normal. No other causes had been detected that would explain the recurrence of gestational losses in the patient.

Results: During the following pregnancy, the patient used a low-dose protocol of intravenous immunoglobulin (KIOVIG 10%) at a dose of 100 mg/kg every 30 days. No other therapy was used. Due to mild to moderate infusion related adverse reactions we used a slow infusion IVIG rate. IgG subclass reconstitution was demonstrated during follow-up. At 40 weeks the patient had a vaginal birth and a healthy live newborn.

Conclusions: Although the scope of our observation is limited to only one case, we suggest that low-dose intravenous gammaglobulin therapy may have been an important factor in the reproductive success obtained in this case.

Keywords: immunemodulation, Recurrent pregnancy loss, IgG subclass

PV112 / #572

POSTER SESSION 10: SLE, ILD AND NOVEL THERAPEUTIC TARGETS
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THE POTENTIAL ANTI-INFLAMMATORY ROLE OF MUCOSAL BACTERIAL VACCINES IN PATIENTS WITH RHEUMATIC DISEASES AND RECURRENT INFECTIONS

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Background and Aims: In patients with systemic autoimmune rheumatic diseases (SARDs), the prevalence of bacterial infections is a frequent complication. As co-therapy, the use of mucosal bacterial vaccines (mucosal trained immunity–based vaccines) has been described as a possible effective tool for therapy of recurrent infections in SARDs. However, these vaccines have also demonstrated an immunomodulatory role. In an experimental study, MV130 (a preparate of bacterial mucosal vaccine), disclosed therapeutic potential for the treatment of ulcerative colitis. The possible role of this vaccine has not been specifically evaluated as anti-inflammatory strategy in SARDs patients with recurrent infections.

Methods: We performed an exploratory preliminary study to assess the potential protective role of bacterial mucosal vaccination indicated to control recurrent bacterial infections on the number of clinical flares of SARDs in a restrospective observational study performed in a small series of 10 cases. Sublingual vaccines were indicated during 3 months. Clinical follow-up was performed during 3 months after therapy. We compared the mean number of clinical flares observed in clinical records during the previous 6 months before therapy with those observed during 6 months after first administration of the mucosal vaccines. Mann-Whitney test was used to analyse if there was a significative difference.

Results: After follow-up a significant decrease in clinical flares was observed in treated patients (mean 1+/-0.8 vs 2+/-0.9, p=0.028).

Conclusions: We hypothesized that the use of sublingual mucosal bacterial vaccines could help to better control the underlying disease in patients with SARDs treated for recurrent infection. We plan an extended observational study to confirm this observation.

Keywords: Bacterial mucosal vaccines, immunotherapy, immunemodulation

PV113 / #24

POSTER SESSION 11: TARGETED THERAPIES
03-08-2025 10:30 AM - 11:30 AM

SERUM SICKNESS INDUCED BY RITUXIMAB INFUSION

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Background and Aims: A recent study described the epidemiological and clinical characteristics of rituximab induced serum sickness as more than 20%. Most frequent manifestations were rheumatologic symptoms (joint pain) (92%), fever (87%), skin lesions (78%). We present a case of 70-year-old female with serum sickness after three-weeks post-rituximab infusion.

Methods: Patient was diagnosed with Sjogren's syndrome December 2022, planned for rituximab infusion in view of poor response to pulse dexamethasone and oral steroids. Following rituximab infusion, 3 weeks later patient developed fever, chills, arthralgia, skin rashes.

Results: WC: 3.22, Neutrophils 54.7, CRP 50.3, Procalcitonin 0.09, Blood culture no growth, UFEME bland, chest x-ray and CTTAP clear. Treated empirically with IV piperacillin/tazobactam for presumed infection but fever persisted for 10 days. PET scan and bone marrow aspiration was suggested in view of pyrexia of unknown origin but not performed. Eventually, a clinical diagnosis of serum sickness was made in view of her signs and symptoms. Patient started on high dose prednisolone and responded well.

Conclusions: Serum sickness typically presents with a classic triad (fever, rash, myalgia/arthralgia) and is a common event after receiving monoclonal antibodies specifically with rituximab. Risk factors for developing serum sickness include Sjogren's syndrome, polyclonal hypergammaglobulinemia and the administration of rituximab alone. The differential diagnosis of serum sickness should be considered for patients developing fever post-rituximab infusion. It is not necessary to investigate extensively with blood tests, imaging modalities and bone marrow aspirations, if there is no clinical and biochemical evidence of infection. A trial of steroids might be an appropriate course of management.

Keywords: Rituximab, Serum sickness

PV114 / #180

POSTER SESSION 11: TARGETED THERAPIES
03-08-2025 10:30 AM - 11:30 AM

BIOLOGICAL THERAPY IN PSORIATIC ARTHRITIS: WHEN CLINICAL EFFICACY IS OVERSHADOWED BY PARADOXICAL REACTIONS

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Background and Aims: Psoriatic arthritis (PsA) is a chronic inflammatory disease associated with skin psoriasis. The management of the disease is complex and requires a multidisciplinary team. The presented case emphasizes the importance of careful monitoring and treatment adjustment according to the therapeutic response and the occurrence of possible paradoxical reactions.

Methods: The 60-year-old patient, diagnosed with polyarticular PsA in July 2019, followed different synthetic conventional treatment schemes (Methotrexate, Leflunomide) without a satisfactory therapeutic response. From May 2021, the biological therapy with IL-17A inhibitor - Secukinumab was initiated. After 6 months of treatment, the psoriasiform palmar skin lesions resolved and the DAPSA disease activity score revealed low disease activity. However, in October 2022, the patient developed facial skin lesions of papulopustular rosacea, which required a reevaluation of the treatment.

Results: After a rheumatological and dermatological assessment, it was decided to change the therapy to IL-23 inhibitor, Guselkumab. The subsequent therapeutic response was a favorable one, materialized by the remission of rosacea skin lesions and a significant improvement of the DAPSA score, the patient fulfilling the remission criteria.

Conclusions: Recently, blocking IL-17A has become a point of interest in the treatment of papulopustular rosacea, thus we consider skin lesions as a paradoxical reaction. The use of the IL-23 blocker in the case of our patient proved to be beneficial both for the joint injury and for rosacea. So far there are no published data on the effectiveness of Guselkumab in rosacea, the beneficial skin effect being supported by the presentation of several cases of SAPHO syndrome.

Keywords: psoriatic arthritis, Secukinumab, paradoxical reaction

PV115 / #330

POSTER SESSION 11: TARGETED THERAPIES
03-08-2025 10:30 AM - 11:30 AM

TREATMENT WITH INTERFERON-ALPHA IN BEHÇET'S DISEASE: A REVIEW OF THE PUBLISHED DATA

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Background and Aims: Despite the availability of conventional immunosuppressive therapies for Behçet's disease (BD), some cases are resistant to standard treatments. Interferon-alpha (IFN-α) is a promising therapeutic option for refractory BD. This review aims to evaluate the features of IFN-α in managing BD.

Methods: PubMed/Medline, EMBASE, and the Cochrane Central Register of Controlled Trials (CENTRAL) were searched from their inception to 23 July 2024. Only reports presenting data on BD patients treated with IFN-α were included in the study.

Results: We identified 63 articles including 1153 BD patients treated with IFN-α (Table 1). The median (min-max) age at diagnosis of these patients was 28 (6-58) years and the majority (68.7%) were male. Previous or concurrent immunosuppressive treatments in most patients included corticosteroids (92.4%), cyclosporine A (48.2%), and azathioprine (43.5%). The most common indication for IFN-α use was resistant uveitis, followed by mucocutaneous involvement (Table 2). The improvement rate was 87.6% for treatment initiated for uveitis and 83.1% for mucocutaneous involvement. The overall improvement

rate was 87.9%. Although adverse events were observed in 82.4% (n=821/996) of patients, most (95.5%, n=784/821) were flu-like
symptoms.

Table 1. Overview of characteristics and treatment of patients with Behçet's disease treated interferon-alpha

Number of patients, n	1153
Age, years, median (min-max)	28 (6-58)
Sex, male, n (%)	772/1123 (68.7)
Clinical manifestations, n (%)	
Oral aphthous	727/729 (99.7)
Uveitis	831/1119 (74.3)
Skin involvement	463/729 (63.5)
Genital aphthosis&ulcers	399/729 (54.7)
Arthralgia&arthritis	254/761 (33.4)
Thromboembolism	71/761 (9.3)
CNS involvement	64/761 (8.4)
Epididymo-orchitis	32/761 (4.3)
GIS involvement	10/761 (1.3)
Cardiac involvement	3/761 (0.4)
Positive pathergy test, n (%)	106/380 (27.9)
Positive HLA-B5/51, n (%)	202/307 (65.8)
Other immunosuppressive treatment, n (%)	
Corticosteroid	797/863 (92.4)
Cyclosporine A	378/780 (48.5)
Azathioprine	337/780 (43.2)
Colchicine	106/780 (13.6)
Cyclophosphamide	82/780 (10.5)
Anti-TNF-alpha agents	43/810 (5.3)
Mycophenolate mofetil	30/780 (3.8)
Chlorambucil	28/780 (3.6)
Methotrexate	25/780 (3.2)
Levamisole	19/780 (2.4)
Leflunomide	4/780 (0.5)
Tocilizumab	2/780 (0.3)
Tacrolimus	2/780 (0.3)
Others*	7/780 (0.9)
Duration of interferon-alpha treatment, months, median (min-max)	10.1 (0.08-191)
Disease duration, months, median (min-max)	48 (1-396)
Relapse under interferon-alpha, n (%)	217/585 (37.1)
Adverse event due to interferon-alpha, n (%)	821/996 (82.4)
Outcome, n (%)	
Improvement	871/991 (87.9)
No improvement	120/991 (12.1)

CNS, central nervous system; GIS, gastrointestinal system; HLA, human leucocyte antigen; TNF, tumor necrosis factor
**pentoxifylline (n=1), vincristine (n=1), doxorubicin (n=1), bleomycin (n=1), vinblastine (n=1), dacarbazine (n=1), thalidomide (n=1)*

Table 2. Indications and treatment responses of interferon-alpha in patients with Behçet's disease

Indications for interferon-alpha treatment	Response to interferon-alpha treatment
Uveitis (n=919)	Improvement (n=738) No improvement (n=104) NI (n=77)
Mucocutaneous involvement (n=117)	Improvement (n=49) No improvement (n=10) NI (n=58)
GIS involvement (n=23)	Improvement (n=21) No improvement (n=2)
Vascular involvement (thromboembolism, n=23)	Improvement (n=22) No improvement (n=1)
CNS involvement (n=19)	Improvement (n=18) No improvement (n=1)
Cardiac involvement (n=3)	Improvement (n=2) No improvement (n=1)
GIS involvement (n=19)	Improvement (n=2)
CS-dependent disease (n=1)	Improvement (n=1)
NI (n=66)	Improvement (n=60) No improvement (n=6)

CNS, central nervous system; CS, corticosteroid; GIS, gastrointestinal system; NI, not indicated

Conclusions: The present systematic review showed that IFN-α is widely used in BD, especially in uveitis refractory to multiple immunosuppressive agents, and is effective in BD. However, further controlled trials are needed to analyze the indications for IFN-α in BD.

Keywords: Behçet's disease, interferon-alpha, uveitis

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POSTER SESSION 11: TARGETED THERAPIES
03-08-2025 10:30 AM - 11:30 AM

ASSESSMENT OF FIBRINOLYTIC SYSTEM PARAMETERS IN PATIENTS WITH RHEUMATOID INFLAMMATION AND CONCOMITANT PERIODONTAL DISEASE.

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Background and Aims: The objective of the study was to assess the periodontal health of patients with rheumatoid arthritis (RA) and its influence on inflammatory markers, fibrinolytic parameters, and red cell indices, with a focus on understanding the risk of cardiovascular disease development.

Methods: In this cross-sectional study, 50 RA patients undergoing biological therapy and exhibiting gingivitis or periodontitis were included. Serum levels of erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) served as inflammatory markers, while plasminogen activator inhibitor-1 (PAI-1), vascular endothelial growth factor (VEGF), intercellular adhesion molecule (ICAM), and red cell distribution width (RDW) were examined to evaluate cardiovascular disease risk.

Results: No statistically significant correlation was observed between periodontal status and RA activity. RA patients exhibited significantly higher concentrations of PAI-1 ($p = 0.0006$) and VEGF ($p < 0.0001$) compared to the control group. There was no correlation between RA and ICAM levels or between fibrinolytic parameters and RA disease activity. Notably, PAI-1 concentration showed significant differences with periodontal screening index (PSI) scores ($p = 0.025783$), as did CRP results ($R=-0.264510$, $p=0.048837$) and periodontal indexes. RDW values exhibited significant differences with ESR ($R=0.369398$, $p=0.008289$) and CRP ($R=0.367405$, $p=0.008672$), as well as with VEGF ($R=-0.352027$, $p=0.025903$). RDW values correlated with RA activity ($R=0.286387$, $p=0.043769$) and disease duration ($R=0.339425$, $p=0.015889$). No statistically significant relationships were found between RDW and periodontal indices or PAI-1 and ICAM parameters.

Conclusions: In conclusion, elevated CRP levels and PAI-1 concentrations in RA patients with periodontitis suggest a negative impact on overall clinical condition and an increased risk of cardiovascular disease.

Keyword: rheumatoid arthritis, PAI-1, VEGF, periodontitis, cardiovascular diseases

PV117 / #363

POSTER SESSION 11: TARGETED THERAPIES
03-08-2025 10:30 AM - 11:30 AM

WILL JAKI BE SAFER THAN ANTI TNF BIOLOGICS IN THE SETTING OF ANA POSITIVITY

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Background and Aims: To shed light on the relative safety of JAKis in RA patients with ANA positivity and to compare that with the safety of Anti TNF biologics in a similar setting.

Methods: Two clinical cases will be presented reflecting the objectives of this presentation

Results: Positivity of ANA is a risk factor for the appearance of anti-drug antibodies during infliximab or adalimumab therapy in patients with rheumatoid arthritis Unfortunately, these agents have been documented to cause lupus-like syndrome with the appearance of new onset ANA and anti-dsDNA. This is unlike the case of JAKis use in a similar setting

Conclusions: Though ANA positivity should be considered as a relative contraindication on initiating Anti TNF biologics and close monitoring for development of drug induced lupus needed, however, this doesn't seem to be the case with initiating JAK inhibitors therapy. On the contrary such therapy with ANA positivity in RA could be beneficial. The international rheumatology organizations guidelines need to include a clear precaution regarding the use of Anti TNF therapy in the setting of ANA positivity as such patients are more prone for development of anti- drug antibodies and are less responsive to therapy and more at risk of drug induced lupus.

Keyword: Anti TNF, JAKi, ATIL

PV118 / #416

POSTER SESSION 11: TARGETED THERAPIES
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MALIGNANT DISEASES IN PATIENTS WITH RHEUMATIC DISEASES TREATED WITH DMARDS

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Background and Aims: The study objective is to analyze the incidence of malignancies in patients with rheumatic muscle-skeletal disorders (RMDs) receiving biologic or targeted synthetic DMARDs (b/tsDMARDs)

Methods: Patients treated with b/tsDMARDs who were diagnosed with rheumatoid arthritis according to the 2010 EULAR/ACR classification criteria and spondyloarthritis according to the ASAS criteria were included in the study. Data from medical records were analysed.

Results: A total of 674 patients is currently being treated with bDMARDs (anti-TNFα, anti-IL6, anti-IL17, anti-IL23, anti-IL1, anti-CD20, abatacept) and 115 patients with tsDMARDs. The number of patients with malignancies during treatment with b/tsDMARDs was 17 (2.5%), eight were women and nine were men. The patients average age was 64.5 years (48-81). The average time of illnes was 20 years (2-38). The average onset of the malignancy was eight years after the start of active treatment (2-14). Five patients were treated with b/tsDMARDs, twelve patients were treated with b/tsDMARDs and csDMARDs. 14 patients were treated with TNFα inhibitors. Types of malignancies were breast cancer (six patients), colon cancer (two patients), papillary thyroid cancer (two patients), vocal cord/oropharynx cancer (two patients), basal cell carcinoma (two patients) and one patient each with lung cancer (this patient is one of two with basal cell carcinoma), ovarian cancer, GIST and melanoma.

Conclusions: The frequency of malignancies in RMDs patients is highest in patients who were treated with TNF inhibitors. The most common type of malignancy is breast cancer.

Keywords: biological therapy, neoplasms, Arthritis, Rheumatoid

PV119 / #428

POSTER SESSION 11: TARGETED THERAPIES
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EVALUATING THE EFFECTIVENESS AND SAFETY OF UPADACITINIB IN MANAGING RHEUMATOID ARTHRITIS: A POST-MARKETING ANALYSIS

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Background and Aims: Background: While biologics (bDMARDs) dominate rheumatoid arthritis (RA) treatment landscape, JAK inhibitors have rapidly emerged as a promising alternative, with strong efficacy and safety profiles across various clinical scenarios. **Objectives:** We conducted a prospective, longitudinal, multicenter study to assess the real-world application of upadacitinib (UPA) in managing RA, focusing particularly on difficult-to-treat cases. Our objectives included mapping prescription patterns for JAK inhibitors within a Romanian cohort, evaluating effectiveness as both monotherapy and in combination with conventional synthetic DMARDs (csDMARDs), and identifying potential predictors of treatment response.

Methods: Methods: RA patients prescribed UPA under local guidelines were monitored biannually for efficacy and safety, using both standard and clinical assessments.

Results: Results: Out of 512 RA patients in our rheumatic disease register, 75 (60 women) were included in this study. At baseline, 50 patients had a history of at least one b/tsDMARD, with 42 classified as difficult-to-treat. After six months, significant improvements were observed, with reductions in Disease Activity Score (DAS28-ESR) and Simplified Clinical Disease Activity Index (SDAI) ($p < 0.05$). ACPA serology and previous b/tsDMARD use were identified as predictors of response. Cycling within the JAK inhibitor class showed no negative impact on outcomes, although seven patients discontinued UPA due to suboptimal efficacy. Importantly, no cardiovascular events, such as deep vein thrombosis or pulmonary embolism, were reported, and there was only one case of herpes zoster infection.

Conclusions: Conclusion: Our data support the efficacy and safety of upadacitinib in real-world settings, emphasizing its value in managing difficult-to-treat RA cases.

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POSTER SESSION 11: TARGETED THERAPIES
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RISK OF MALIGNANCIES ASSOCIATED WITH BDMARDS AND JAK INHIBITORS THERAPY IN PATIENTS WITH RHEUMATIC DISEASES

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Background and Aims: The growing use of bDMARDs and tsDMARDs therapy causes necessity of risks evaluating taking into consideration the contradicting data on malignancy risk. **Objectives** The objective of this study is to analyze the incidence of malignancies in rheumatic diseases patients treated with bDMARDs and JAK inhibitors.

Methods: Data on 3,827 bDMARDs and JAK inhibitors patients with rheumatic diseases from the registry of the I. I. Mechnikov North-West State Medical University (2005-2024) were analyzed. 28 patients were included (mean age: 56.93±9.78 years, 19 (67.86%) females): 13 with rheumatoid arthritis (46.43%), 9 with ankylosing spondylitis (32.14%), 4 with psoriatic arthritis (14.29%), 1 with systemic scleroderma (3.57%), and 1 with Churg-Strauss syndrome (3.57%). Therapy duration median at the time of malignancies was 10 [0.083–16] years. The disease's mean duration before malignancies debut was 15.57±9.4 years.

Results: The total observation period was 72,713 patient-years. The treatments included: TNF-alpha inhibitors, IL-6, IL-17, IL-23 inhibitors, JAK inhibitors, rituximab, and abatacept. A total of 28 solid tumors were identified (0.03 per 100 patient-years), including 6 cases of breast cancer, 3 cases each of thyroid, kidney, and skin cancer, 2 cases each of lung and ovarian cancer, and 1 case of cancer in other locations. 16 cases (57.14%) reported in TNF-alpha inhibitors patients.

Conclusions: The data obtained demonstrates 0.73% of malignancies incidence with bDMARDs and JAK inhibitors that does not exceed the incidence in general population of Russian Federation (2.84%) and indicates a positive safety profile and no increased risk of malignancies de novo in rheumatic disease patients.

Keywords: Malignancies, JAK inhibitors safety profile, bDMARDS safety profile

PV121 / #492

POSTER SESSION 11: TARGETED THERAPIES
03-08-2025 10:30 AM - 11:30 AM

WHY JANUS KINASE INHIBITOR (JAKI) DRUGS ELIMINATE SOME BUT NOT ALL AUTOIMMUNE CD8 T CELLS: A CLUE FROM TCR/CD3 AND FAS-DEPENDENT MECHANISMS

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Background and Aims: How are CD8 T cells inhibited by JAKi, a drug class used for several autoimmune disorders? Initially, we hypothesized that via cytokine receptor inhibition, JAKi would directly block CD8 T cell growth and clonal expansion, but results did not match this expectation.

Methods: OT1.RAG-/- T cells were stimulated with various peptide antigens followed by flow cytometry, cell division, and viability analysis. In vivo, the C3H/HeJ inbred mouse model of T cell-mediated autoimmune Alopecia Areata (AA) was used, grafting healthy skin onto full-disease recipient mice, thence measuring hair loss-versus growth on the new grafts.

Results: JAKi inhibited OT1 CD8 T cell clonal expansion, as expected. However, in the presence of anti-Fas ligand (FasL) blockade, clonal expansion was rescued, because JAKi was not inhibiting basal survival, cell cycle progression, and cell division, but was rather allowing those to proceed acting instead on Fas-dependent activation-induced cell death (AICD). In vivo, JAKi (Tofacitinib) inhibited the autoimmune reaction of AA and favored hair regrowth in ~90% of mice, but adding anti-FasL blocking mAb reduced drug responder frequency to ~50%. These unanticipated results were a function of TCR/CD3 stimulation. Whereas CD8 T cells bearing unengaged TCR/CD3 died from JAKi treatment without requiring Fas death signaling, engaging TCR/CD3 required Fas activation for JAKi to induce death.

Conclusions: When TCR/CD3 reactions were active, JAKi drugs blocked CD8 T cell responses by Fas-dependent death rather than by inhibition of growth. Some T cell subsets are resistant to Fas-mediated death, and might be better eliminated upon pharmacological targeting of these additional pathways.

Keywords: Janus Kinase Inhibitor, CD8 T cell, Fas

PV122 / #563

POSTER SESSION 11: TARGETED THERAPIES
03-08-2025 10:30 AM - 11:30 AM

EFFICACY OF RISANKIZUMAB IN ACTIVE PSORIATIC SPONDYLARTHRITIS IN CLINICAL PRACTICE IN RUSSIA

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Background and Aims: Risankizumab, an interleukin-23p19 inhibitor, is indicated for the treatment of moderate to severe plaque psoriasis and active psoriatic arthritis in adult patients in Russia. However, data on the efficacy of risankizumab in clinical practice in patients with psoriatic arthritis, and particularly in psoriatic spondylitis, are limited. **Aim** To evaluate the effectiveness of risankizumab in patients with active psoriatic arthritis and spondylitis.

Methods: The prospective analysis included 22 patients with active psoriatic arthritis who received risankizumab for at least 4 months and completed >3 injections at 0, 4, 16, 28 weeks. Gender: 14 men (64%) and 8 women (36%). The average age was 39 years. The most common manifestations of musculoskeletal involvement were arthritis (90%) and sacroilitis (75%). HLA-CW6 positivity was 30%, and HLA-B27 positivity was 25%. Bio-naïve patients - 35%, most of the patients were bio-experienced (iTNF-alpha, n=7 (35%), iIL-17, n=10 (50%), iIL-23, n=2 (10%), and iIL-12/23, n=2 (10%)).

Results: All 22 patients were with high activity of psoriatic arthritis and psoriasis (DAPSA = 33 (avg), PASI = 33.2 (avg)), active sacroiliitis in 12 patients (BASDAI = 4.9 (avg), ASDAS = 3.6 (avg). All patients demonstrated disease activity reduction on >3 injections of risankizumab with treatment target achieved (DAPSA=7.14 (avg), BASDAI=1.1 (avg), ASDAS=1.5 (avg), PASI=2.9).

Conclusions: Risankizumab demonstrates high efficacy in all populations of patients with active psoriatic arthritis and particularly spondylitis in real clinical practice regardless of previous therapy with csDMARDs and bDMARDs.

Keywords: IL23 inhibitors, Psoriatic spondyloarthritis, Risankizumab

PV123 / #538

POSTER SESSION 11: TARGETED THERAPIES
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MALIGNANCIES RISK IN JAK INHIBITORS THERAPY

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Background and Aims: JAK inhibitors are now widely used in real clinical practice in rheumatic patients. However, safety and efficacy data are still limited, especially with malignancies incidence. Most studies demonstrate a good safety profile and no significant risk of adverse events, including malignancies. However, some data contradict these findings. **Objective** The objective of this study is to analyze the incidence of malignancies in rheumatic patients treated with JAK inhibitors.

Methods: Data on 3,827 biologics-treated rheumatic patients from the registry of the I. I. Mechnikov Nort-West State Medical University (2005-2024) were analyzed. 372 received targeted therapy: upadacitinib (n=227), tofacitinib (n=75), and baricitinib (n=70).

Results: The study included 3 JAK inhibitors patients with confirmed malignancy (mean age: 52±10.44 years, 3 males): 2 with rheumatoid arthritis (66.67%) and 1 with ankylosing spondylitis (33.33%). The median therapy duration by malignancy diagnosed was 2 [1–2] years. The disease's duration mean was 8±8.6 years. 3 cases of solid tumors were identified (0.81%): 1 case of testicular cancer and 1 case of laryngeal cancer - on tofacitinib therapy (2.6%), and 1 case of papillary thyroid carcinoma (0.44%) - on upadacitinib therapy.

Conclusions: The data obtained demonstrates no increased incidence of malignancies de novo on JAK inhibitors therapy. It is comparable to the rate of the general population (2.84%) in Russia. The study results demonstrate the positive safety profile of JAK inhibitors, especially upadacitinib.

Keywords: Malignancies, JAK inhibitors safety profile, upadacitinib safety profile

PV124 / #555

POSTER SESSION 11: TARGETED THERAPIES
03-08-2025 10:30 AM - 11:30 AM

HERPES ZOSTER RISK ON UPADACITINIB IN CLINICAL PRACTICE IN RUSSIA

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Background and Aims: Upadacitinib is a selective JAK1 inhibitor effective in rheumatic diseases. However, safety data from clinical practice is limited, especially the rate of Herpes zoster. **The aim.** To determine rate of Herpes zoster on upadacitinib in patients with rheumatic diseases.

Methods: Registry data of the North-West State Medical University n.a. I.I. Mechnikov (2005—2024) on bDMARDs and JAK inhibitors (n= 3,827) was analyzed.

Results: 227 patients from a total of 3827 received Upadacitinib with 2 cases of Herpes zoster (0.9%) in rheumatoid arthritis patients with localization in the intercostal space (n=1) and upper limbs (n=1). Gender and age did not increase the incidence of Herpes zoster. The risk factors were: high disease activity (HR 1.3; 95% CI 1.1-1.5)), concomitant therapy with glucocorticosteroids >10 mg per day (HR 1.4; 95% CI 1.1-1.7), cyclophosphamide (HR 3.9; 95% CI 1.6—7.3), leflunomide (HR 1.3; 95% CI 1.1—1.9), azathioprine (HR 1.9; 95% CI 1.3—1.9) —3.4), methotrexate (HR 1.29; 95% CI 0.37—4.47), while Upadacitinib (HR 0.9; 95% CI 0.78-1.04) was not associated with Herpes zoster risk. Herpes zoster cases were non-serious with full recovery in 10-14 days on acyclovir. These cases were reported in the first 2-6 months of Upadacitinib therapy with no relapses after antiviral treatment. Upadacitinib was discontinued during antiviral treatment, then re-initiated.

Conclusions: There was no increase in herpes zoster rate on Upadacitinib; it does not exceed herpes zoster rate in the general population. Our data demonstrates a favorable safety profile for upadacitinib with herpes zoster risk.

Keywords: JAK inhibitors, herpes zoster, JAK inhibitors safety profle

PV125 / #478

POSTER SESSION 11: TARGETED THERAPIES
03-08-2025 10:30 AM - 11:30 AM

DESCRIPTIVE ANALYSIS OF BIOLOGICAL THERAPY OPTIMIZATION IN PATIENTS WITH AUTOIMMUNE DISEASES: EXPERIENCE FROM A COLOMBIAN CENTER

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Background and Aims: Optimizing biological therapy is crucial for improving clinical outcomes and reducing costs in patients with autoimmune diseases. This study evaluates the effectiveness of biological therapy optimization in terms of economic savings, treatment strategies, and failure rates.

Methods: A retrospective analysis was conducted on 578 patients with autoimmune diseases, including rheumatoid arthritis (n=413), juvenile idiopathic arthritis (n=89), systemic lupus erythematosus (n=35), and spondyloarthropathies (n=14). The types of biologics used, costs before optimization, and savings after optimization were documented. Biologic failure rates and current status in the program were also analyzed.

Results: Sixteen biologics were analyzed. The highest savings were achieved with etanercept ($202,647 USD), certolizumab ($137,616 USD), and abatacept ($101,220 USD), with a total savings of $767,245 USD. The overall failure rate across biologics was 36.9% (Table 1). In terms of optimization, 413 patients were on first-line biologics, with 89 on second-line and 35 on third-line. The majority of patients (n=329) remain on the optimization programme. Table 1. Savings and Failure Rates in Biological Therapy

Biologic	Savings in
Abatacept	$101,220
Adalimumab	$43,513
Baricitinib	$13,000

Biologic	Savings in
Belimumab	$10,008
Certolizumab	$137,616
Dupilumab	$17,398
Etanercept	$202,647
Golimumab	$48,021
Infliximab	$15,737
Ixekizumab	$11,149
Risankizumab	$1,941
Rituximab	$70,534
Secukinumab	$6,761
Tocilizumab	$65,715
Tofacitinib	$57,510
Upadacitinib	$6,165
Ustekinumab	$37,552

Conclusions: Biological therapy optimization in autoimmune diseases generates substantial savings, with most patients successfully managed on first-line biologics. Optimization strategies are crucial for long-term sustainability in healthcare.

Keywords: autoimmune diseases, Biological Therapy Optimization

PV126 / #209

POSTER SESSION 12: AUTOINFLAMMATORY DISEASES
03-08-2025 10:30 AM - 11:30 AM

THE PREVALENCE OF HLA-B27 IN FAMILIAL MEDITERRANEAN FEVER RELATED SACROILIITIS

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Background and Aims: Familial Mediterranean fever (FMF) is an autoinflammatory disorder characterized by recurrent fever, serositis, articular syndrome with some reported cases of sacroiliitis. In the majority of cases FMF sacroiliitis is not HLA-B27 associated, there is limited data on the prevalence of HLA-B27 antigen among FMF patients and its influence on the severity of FMF associated sacroiliitis. This study **aims** to investigate the prevalence of HLA-B27 in FMF patients with inflammatory low back pain and its role in disease severity.

Methods: 47 Armenian patients with a mean age of 33,53±11,73 years (76,6%-male, 23,4%-female) who met the criteria for FMF (Tel-Hashomer) and inflammatory low back pain (ASAS) were included. MEFV mutations and HLA-B27 presence were evaluated. Schober's test was measured, sacroiliac joints and lumbar spine X-ray and radiological grading according to the New York criteria was performed. Functional ability of sacroiliac joints and disease activity were measured by BASDAI and BASFI scores.

Results: The mean age of FMF diagnosis was 13.71±10.56 years, with a duration of 6.35±10.39 years. Low back pain onset was at 22.29±11.47 years (mean duration 9.65±5.86 years). HLA-B27 was found in 65.96% of FMF patients with sacroiliitis. Unilateral sacroiliitis was present in 8 cases, while bilateral in 39. Correlation analyses revealed a weak association between HLA-B27 and BASFI scores ($p<0.05$), but no significant link with sacroiliitis severity.

Conclusions: HLA-B27 is more frequently present in FMF patients than in the general population, suggesting potential overlap with ankylosing spondylitis. Further studies with larger groups can potentially explore these findings.

Keywords: HLA-B27, Familial Mediterranean fever(FMF), Sacroiliitis

PV127 / #242

POSTER SESSION 12: AUTOINFLAMMATORY DISEASES
03-08-2025 10:30 AM - 11:30 AM

TWO CASES OF CHILDHOOD LINEAR IGA BULLOUS DERMATOSIS FOLLOWING MULTIVISCERAL TRANSPLANT

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Background and Aims: Linear IgA Bullous Dermatosis (LABD) is a rare blistering disorder that has been associated with various triggers, including medications, gastrointestinal conditions, and autoimmune diseases. However, its connection to organ transplantation remains poorly understood. This report highlights two pediatric cases of LABD following multivisceral transplantation (MVTx) from our institution.

Methods: Clinical data were collected from the patients' medical records, including demographic information, timing of LABD onset, immunosuppressive regimens, and clinical course. The diagnosis of LABD was confirmed by skin biopsy and direct immunofluorescence, which revealed linear IgA deposition at the basement membrane zone. Relevant laboratory results, histopathological findings, and treatment responses were also reviewed.

Results: Patient 1, a 16-month-old female, developed biopsy-confirmed LABD three months after MVTx for megacystis microcolon intestinal hypoperistalsis syndrome. Despite prophylactic dapsone and immunosuppressive therapy, her condition worsened with infections. She initially received dapsone and prednisone but required rituximab, IVIG, and omalizumab for disease control. Patient 2, an 11-month-old female, developed LABD three months after MVTx for biliary atresia. Managed with tacrolimus, mycophenolate mofetil, and prednisone, she initially responded to dapsone. However, new vesicles appeared, attributed to Koebnerization, and remission was achieved with oral and topical dapsone.

Conclusions: Both patients were younger than the typical LABD onset age of 4.5 years, with disease appearing three months post-transplant and no clear triggers. Most transplant-related LABD cases in the literature are idiopathic or drug-related. Possible mechanisms include graft-versus-host disease or immune dysregulation due to

immunosuppressive therapy. Further research is needed to understand LABD's pathogenesis in transplant patients and optimize management.

Keywords: pediatric, Linear IgA bullous dermatosis, Transplant

PV128 / #278

POSTER SESSION 12: AUTOINFLAMMATORY DISEASES
03-08-2025 10:30 AM - 11:30 AM

UTILIZATION OF POSITRON EMISSION TOMOGRAPHY IN DIAGNOSING ADULT-ONSET STILL'S DISEASE: A SYSTEMATIC REVIEW

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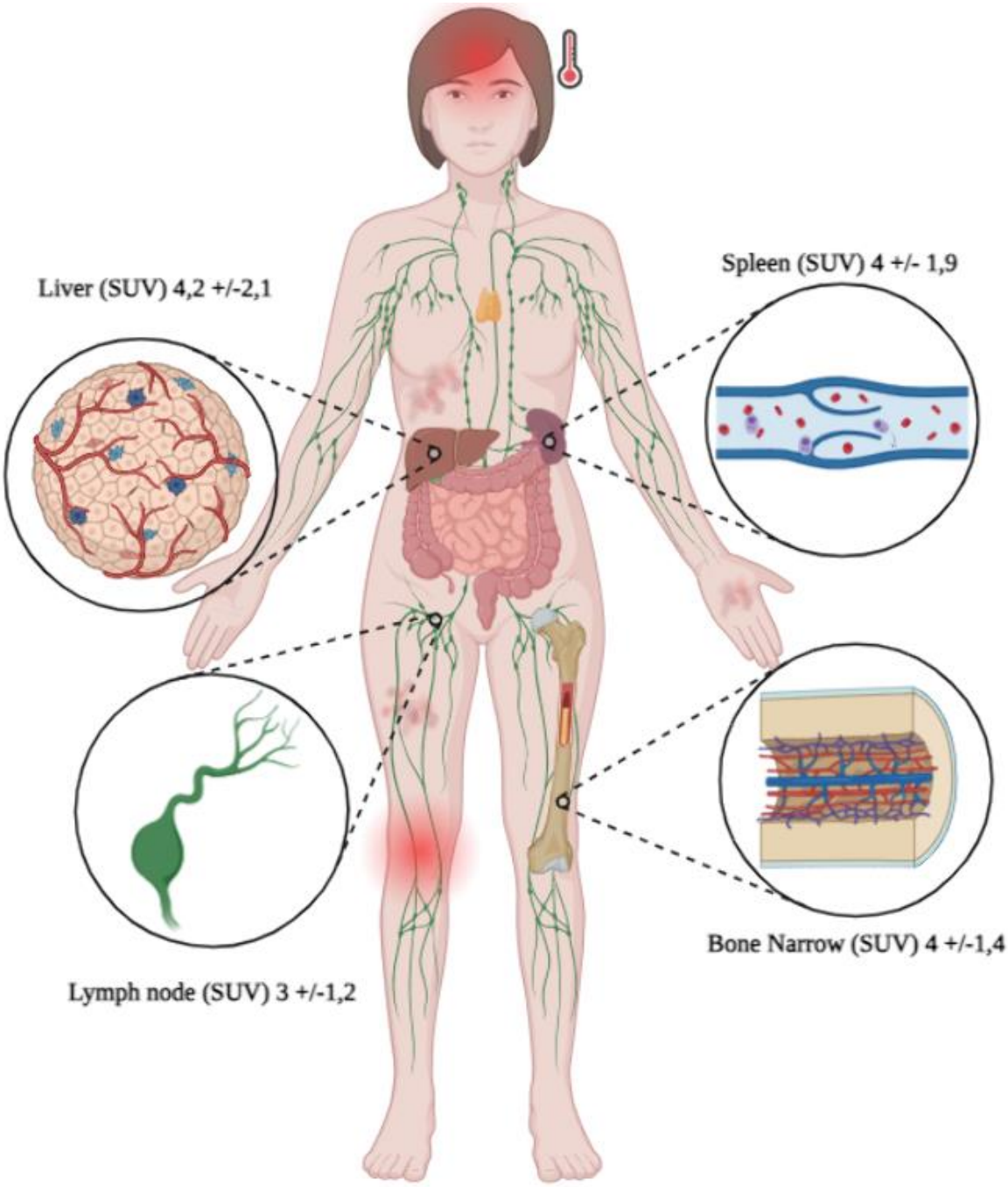
Background and Aims: Adult-onset Still's disease (AOSD) is a systemic inflammatory disorder that is difficult to diagnose due to its nonspecific symptoms and the need to exclude other conditions. Positron Emission Tomography (PET) with 18F-fluorodeoxyglucose (FDG) is emerging as a diagnostic tool. This systematic review evaluates the role of PET in diagnosing AOSD.

Methods: A systematic review was conducted using PRISMA guidelines. We searched PubMed, Ovid, and EMBASE databases for studies published in the last decade that utilized PET in adult patients diagnosed with AOSD. A total of 54 studies involving 545 patients were included. Data were synthesized using the "Synthesis without meta-analysis" (SWiM) framework, and quality was assessed using Joanna Briggs Institute (JBI) tools.

Results: PET was used for diagnosis in 77% of studies. FDG was the primary tracer in 85% of cases, with doses ranging from 3 to 5.5 Mbq/kg. The highest standardized uptake values (SUV) were observed in bone marrow (mean SUV: 4 ± 14), followed by lymph nodes, spleen, and liver (Image 1). The Yamaguchi criteria were used in 63% of cases for diagnosis. PET aided differential diagnosis in 85% of cases, especially in fever of unknown origin and suspected

neoplasms.



Conclusions: PET, particularly with FDG, shows diagnostic utility in AOSD, helping differentiate it from other inflammatory and neoplastic conditions. Further research is needed to standardize its use in AOSD diagnosis, particularly in cases of fever of unknown origin.

Keywords: Adult-Onset Still's Disease, Positron-Emission Tomography, Diagnostic Imaging

PV129 / #372

POSTER SESSION 12: AUTOINFLAMMATORY DISEASES
03-08-2025 10:30 AM - 11:30 AM

PREFERENCES OF IMMUNOSUPRESSIVE TREATMENT OF CHRONIC RECURRENT MULTIFOCAL OSTEOMYELITIS (CRMO) IN ADULTS

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Background and Aims: Data about therapeutic treatment of CRMO in adults are limited due to orphan nature of the disease, rare onset of the disease in adults and extreme rarity of monofocal forms. Aim of the current study is to show the efficacy of one-year non-surgical treatment of CRMO in adults.

Methods: Data collection was carried out in St. Petersburg Clinical Rheumatology Hospital No. 25, North-Western State Medical University named after I.I. Mechnikov, Tyumen Regional Clinical Hospital №1. From 1790 patients 12 patients aged from 19 to 69 years with a morphologically confirmed diagnosis of “CRMO” were involved in the study. The follow-up period for the patients with CRMO was 12 ± 1 months. Clinical, demographic and laboratory data of the patients were recorded.

Results: Gender and age structure of the studied group of patients (n, %): female 7 patients (58.33%), male 5 patients (41.67%). Characteristics of the treatment and results of the non-surgical treatment of CRMO in adults are presented in the tables 1-3. Table 1. Drug therapy of adult CMNO,
n=12

Name of the drug with dose or pharmaco-therapeutic group	Patients, n (%)
Non-steroid anti-inflammatory drugs (NSAIDs)	12 (100)
Glucocorticoids (GCS), 5-10 mg / day, per os	3 (25.0)
Sulfasalazine, 2–3 g/day	4 (33.33)
Methotrexate, 15–25 mg/week	1 (8.33)
Mycophenolate mofetil, 1–3 g/day	1 (8.33)
Leflunomide, 20 mg/day	1 (8,33)
Zoledronic acid, 5 mg/6 months	2 (16.67)
Pamidronic acid, 90 mg/ 12 months	1 (8.33)
Adalimumab, 40–80 mg/2 weeks	2 (16.67)
Infliximabe, 5 mg/kg / 8 weeks	2 (16.67)
Golimumab, 50 mg/4 weeks	1 (8.33)

Table 2. Surgical treatment of CRMO,
n=12

Type of the syrgery	n (%)
Bone resections before the immunosuppressive treatment	6 (50)
Bone resections before the immunosuppressive treatment	6 (50)
Surgical treatment after initiation of the immunosuppressive therapy (table 1)	0 (0)

Table 3. The results of the immunosuppressive treatment of CRMO,
n=12

Parameters of activity	Baseline	Month 12	p-value
Pain, VAS (0-10), Mean±SD	5.2±1.64	1.25±0.96	0.004
Number of relapses*	12	3	NA
CRP, mg/L, Mean±SD	57.1±35.12	5.61±2.22	0.02
ESR, mm/h, Mean±SD	36.91±30.16	18.64±21.39	0.2

VAS: visual analogue scale, CRP: C-reactive protein, ESR: Erythrocyte Sedimentation Rate

*Relapses was only in first six months, no one relapses after initiation of immunosuppressive therapy.

Conclusions: The immunosuppressive treatment in the most perspective option to control of the disease activity. Surgical treatment could be used only in diagnostic approaches and is ineffective as treatment option in CRMO.

Keywords: “sterile” osteomyelitis, chronic recurrent multifocal osteomyelitis in adults, CRMO treatment

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POSTER SESSION 12: AUTOINFLAMMATORY DISEASES
03-08-2025 10:30 AM - 11:30 AM

DOUBLE-NEGATIVE T-LYMPHOPROLIFERATION AND SYSTEMIC INFLAMMATION ASSOCIATED WITH A HETEROZYGOUS PRKCD MUTATION

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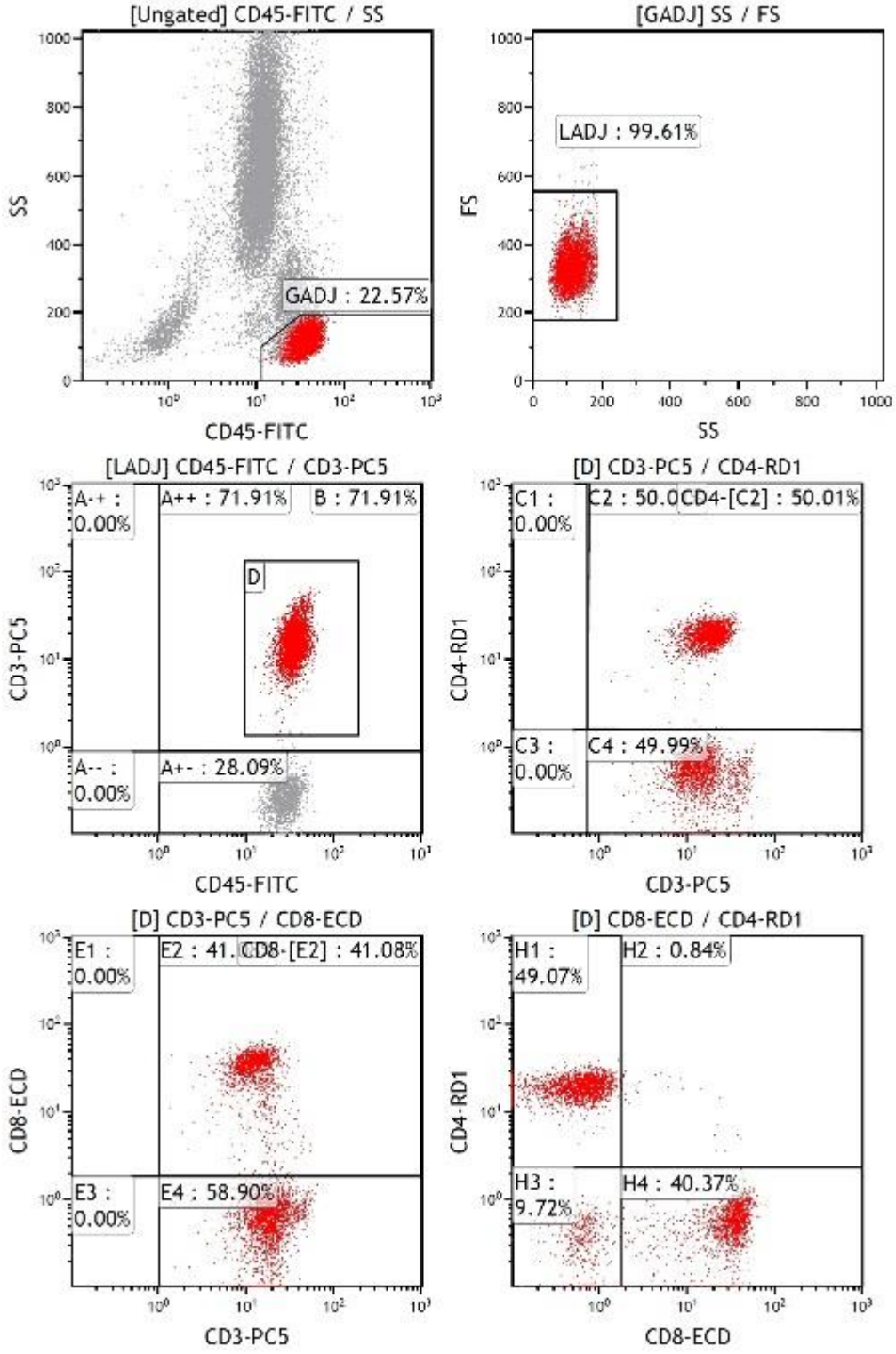
Background and Aims: Homozygous mutations in the *PRKCD* gene, which encodes protein kinase C delta, are associated with early-onset autoimmune lymphoproliferative syndrome and immunodeficiency. However, the clinical implications of heterozygous mutations are poorly understood.

Methods: We report the case of a 21-year-old woman with a heterozygous *PRKCD* mutation, presenting with painful, tender lymphadenopathy and systemic inflammation.

Results: The patient developed intermittent spiking fever, malaise, and right-sided neck lymphadenopathy one year prior to presentation. Physical examination revealed palpable, hard, and tender right supraclavicular lymph nodes. Computed tomography showed numerous conglomerated lymph nodes with perinodal infiltration in the right neck. A core biopsy revealed lymphadenitis with dense eosinophilic infiltration and fibrosis, with no evidence of lymphoma or mycobacterial infection. Laboratory tests showed elevated inflammatory markers. Rheumatoid factor and a panel of autoantibodies were negative, except for an antinuclear antibody titer of 1:320. Positron-emission tomography revealed low-to-moderate uptake in the affected lymph nodes. Flow cytometry of peripheral blood detected an expanded population of double-negative T-cells (9.72% of total T-cells, Figure 1). Given the unexplained and persistent lymphadenopathy and lymphoproliferation, the patient underwent targeted gene panel-based testing, including sequencing of 374 genes associated with primary immunodeficiencies. This testing revealed a heterozygous PRKCD c.931G>A, p.Gly311Arg missense mutation, absent from population databases

(https://gnomad.broadinstitute.org/).



Conclusions: This case highlights a potentially broader spectrum of immune dysregulation associated with heterozygous *PRKCD* mutations, which may present later in life with

clinically significant, though milder, features compared to homozygous mutations. Further research is needed to understand the pathogenetic and clinical consequences of heterozygous *PRKCD* mutations.

PV131 / #433

POSTER SESSION 12: AUTOINFLAMMATORY DISEASES
03-08-2025 10:30 AM - 11:30 AM

THE FIRST FAMILY MALIGNANT CASE OF SINGLETON-MERTEN SYNDROME (DDX58 C.1181A>T (P.LYS394LLE) MUTATION), WHICH HAS PREVIOUSLY BEEN DESCRIBED AS BENIGN

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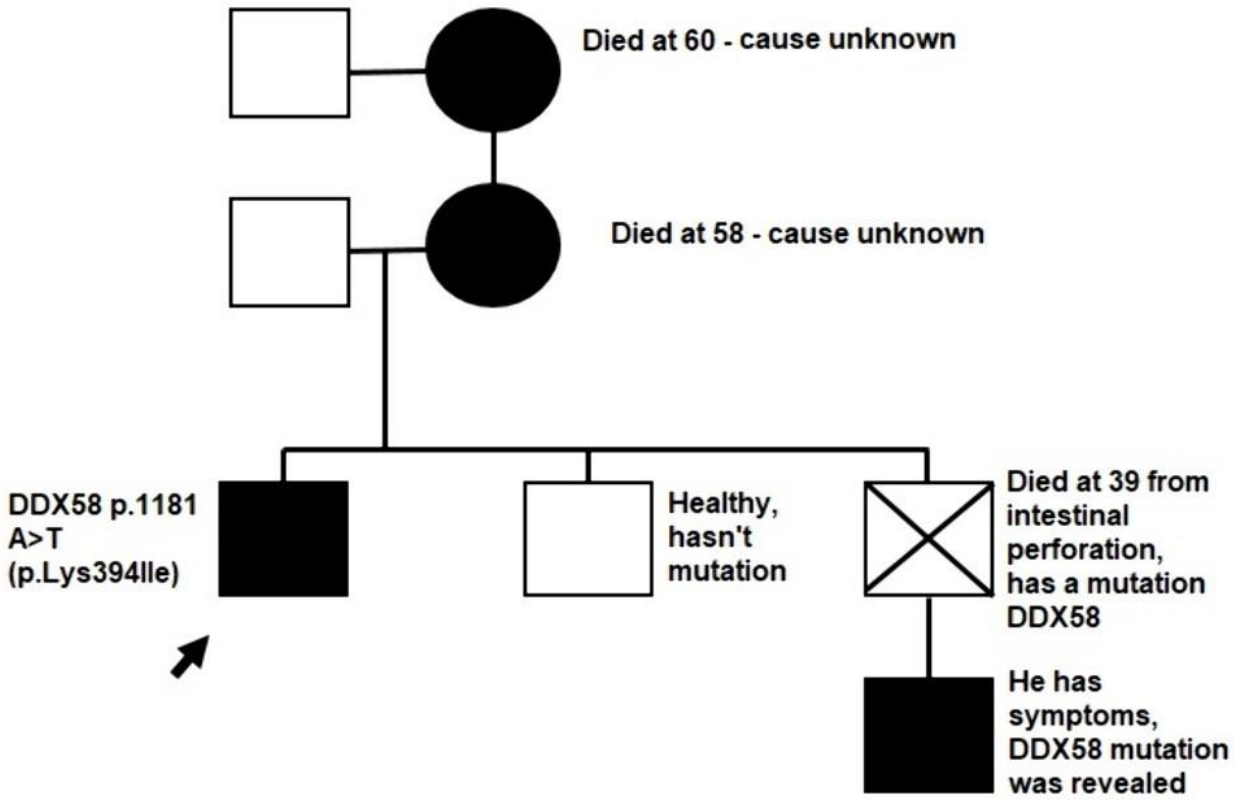
Background and Aims: Background: Singleton-Merten Syndrome (SMS) is a rare autosomal-dominant disorder belonging to the type 1 interferonopathies group caused by DDX58 gene mutation, which was previously considered as benign. Below we discribe the first familial malignant case of SMS.

Methods: The 38-years old man admitted to the clinic. Sickness debuted since age 10, with manifestations of erythematous psoriasis-like rashes on the face and scrotum, urticaria rashes, episodes of abdominal pain and flatulence, accompanied by belching and liquid stool for several hours (which brought relief), blepharoconjuctivitis, fever up to 38C and episodic joint pain. The examination excluded infectious, autoimmune, inflammatory bowel diseases, lactose intolerance, oncohematological diseases. The examination revealed synovitis of large joints, ulcers and hemorrhages of the ileum and rectum, osteoporosis, calcification of the aorta and femoral arteries (patient never smoked). Glucocorticoid therapy was effective, mesalazine wasn't.

Results: Taking into account the family history of the patient (see Figure 1), a genetic study was conducted (patient with complete genomic sequencing and his brothers and nephew with Sanger sequencing). It confirmed the presence of a heterozygous missense-mutation in the gene DDX58 p.1181 A>T (p.Lys394lle) in the affected family members. An increased

expression of interferon genes was determined by real-time PCR in the patient's blood.

Figure 1: Family history



Conclusions: Despite the non-classical picture (in addition to the osteoporosis, aortic calcification, psoriasis-like rashes, DDX58 mutation, and an increase in IFN genes in the blood the presence of intestinal lesions) the patient was diagnosed with SMS.

Keywords: genetics, autoinflammation, mutation

PV132 / #470

POSTER SESSION 12: AUTOINFLAMMATORY DISEASES
03-08-2025 10:30 AM - 11:30 AM

ADENOSIN DEAMINASE 2 DEFICIENCY

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Background and Aims: Adenosine deaminase 2 deficiency (DADA2) is a complex systemic autoinflammatory disorder (autosomal recessive) in which vasculopathy/vasculitis, dysregulated immune function and hematologic abnormalities may predominate. Inflammatory features include intermittent fevers, rash and musculoskeletal involvement. Dysregulation of immune function can lead to immunodeficiency or autoimmunity of varying severity, lymphadenopathy may be present. Hematologic disorders may begin early in life or in late adulthood, and can include lymphopenia, neutropenia, pure red cell aplasia, thrombocytopenia, or pancytopenia. Interfamilial and intrafamilial phenotypic variability can be observed, also some individuals may remain asymptomatic until adulthood or may never develop clinical manifestations of DADA2. A 63-year-old man was hospitalized in Internal Medicine because of persistent fever, pericardial and pleural effusion and chest pain, with microbial culture negative and without response to empiric microbial therapy. In several blood test we found high level of ferritin, C-Reactive Protein and VSG. This pacient blood test showed neutropenia, low IgM levels. Several image test were done in order to rule out infectious and neoplastic diseases. Once infections and malignant tumors were excluded corticotherapy and colchicine were initiated. Clinical and blood test disorders disappeared and the patient became asympthomatic.

Methods: Autoimmune, microbial and image test were done

Results: Our subject presented suggestive clinical and laboratory findings and low ADA2 catalytic activity in plasma.

Conclusions: Molecular confirmation is being ruled. These patients deserve investigation for possible structural variants so an appropriate follow-up and support familial counseling can be done.

Keywords: ADA2, autoinflammatory disorders, fever

PV133 / #536

POSTER SESSION 12: AUTOINFLAMMATORY DISEASES
03-08-2025 10:30 AM - 11:30 AM

THE TWO FACES OF VEXAS SYNDROME: FROM CHRONIC MANIFESTATIONS TO LIFE-THREATENING MACROPHAGE ACTIVATION SYNDROME.

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Background and Aims: VEXAS is a severe, progressive autoinflammatory disease with clinical features that mimic various rheumatologic and hematologic conditions. Systemic inflammation involving the skin, the mucosal membranes, the muscoloskeletal system and the vascular system is associated with myelodysplastic syndrome potentially evolving to hematologic malignancy or macrophage activation syndrome.

Methods: A 65-year-old male referred to our Unit for a long clinical history characterized by fever, arthromyalgias, auricular chondritis, scalp painful nodules, splenomegaly, widespread lymphadenopathies and weight loss. In his medical record a tranfusion-dependent myelodysplastic syndrome and a previous episode of ocular thrombosis. A diagnosis of VEXAS Syndrome was made with confirmatory genetic study showing c.118-1G>C variant in UBA1 gene. The patient was initially treated with prednisone 0,5 mg/kg, with partial clinical response.

Results: After 3 months, the patient presented to the ER for refractory fever and further decline of the known bilinear cytopenia. Initially, he was treated with broad-spectrum antibiotic and antimycotic therapy, with no clinical response. No infectious evidence was found on blood samples. A second bone marrow biopsy showed hemophagocytosis, without elements of immature clonality, confirming the diagnosis of Macrophage activation syndrome on VEXAS Syndrome. The patient was treated with high dose steroids, immunomodulatory iv-Ig, iv anakinra and iv cyclosporine with only initial clinical and laboratory response. However, the patient relapsed quickly and lastly died because of sudden cardiac death.

Conclusions: Macrophage activation syndrome is not widely reported in current literature as a complication of VEXAS Syndrome and future studies will help the clinicians to define the long-term prognostic role of this pathogenetic variant.

Keywords: VEXAS, Macrophage activation syndrome, autoinflammatory disease

PV134 / #544

POSTER SESSION 12: AUTOINFLAMMATORY DISEASES
03-08-2025 10:30 AM - 11:30 AM

MUCKLE-WELLS SYNDROME IN RHEUMATOLOGY PRACTICE: A CASE STUDY

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Background and Aims: Cryopyrin-associated periodic syndromes (CAPS) represent a group of rare, autosomal dominant hereditary autoinflammatory disorders with varying severity. Muckle-Wells syndrome (MWS), a subtype of CAPS, manifests as recurrent fever, urticaria-like skin rash, arthritis, and ocular inflammation. Sensorineural hearing loss develops later in the disease course and is common in approximately 70% of cases. Late complications such as amyloidosis develop in about 25% of patients, further increasing morbidity.

Methods: A Case Study

Results: We present the case of a 42-year-old male patient under rheumatology care since childhood for juvenile idiopathic arthritis (JIA). Despite biologic therapy with adalimumab, persistent elevation of acute phase reactants and recurrent urticaria were noted. Molecular-genetic testing identified a pathogenic variant in the NLPR3 gene, confirming MWS diagnosis. Therapy adjustment to interleukin-1β inhibitor (canakinumab) resulted in significant improvement, including normalization of acute phase reactants and resolution of skin symptoms.

Conclusions: This case underscores the necessity of considering autoinflammatory diseases, especially MWS in the differential diagnosis of patients with JIA who exhibit persistent, unexplained inflammatory activity and skin manifestations, given the substantial overlap in clinical features between MWS and JIA. Timely recognition and targeted therapy significantly improve patient outcomes and reduce the risk of severe complications.

Keyword: Muckle-Wells Syndrome, CAPS, autoinflammatory disease, interleukin-1β inhibitor, canakinumab, juveni

PV135 / #545

POSTER SESSION 12: AUTOINFLAMMATORY DISEASES
03-08-2025 10:30 AM - 11:30 AM

FROM GENETICS TO THE CLINIC: CAN VARIANTS OF UNKNOWN SIGNIFICANCE (VUS) PROVE AN AUTOINFLAMMATORY PHENOTYPE?

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Background and Aims: Targeted gene panels for Next-Generation Sequencing (NGS) are a cornerstone in supporting the diagnosis of systemic autoinflammatory diseases, especially when the clinical phenotype seems unclear. We aimed to correlate the found Variants of Unknown Significance (VUS) with the clinical phenotype of a group of suspected autoinflammatory patients.

Methods: Patients with clinical and biological autoinflammatory features were retrospectively collected. NGS sequencing was performed using the Custom “Fever & Autoinflammatory Disease” panel (*SOPHIA Genetics*) in Illumina MiSeq, analysing coding regions of 17 selected genes. Variant calling and data analysis were performed by the Sophia-DDM-V6.5 bioinformatics programme. Variants were interpreted according to the 2015 ACMG standards and guidelines.

Results: Clinical and demographical data are displayed in Figure 1. Among 103 patients screened, 80 (77.7%) displayed at least one retained non-synonymous variant with a minor allele frequency (MAF) <= 0.05. Of these, 34 carried at least one VUS, and 6 had pathogenic variants confirming the phenotype. Two distinct clusters of patients were identified with VUS on NOD2 (p.Arg702Trp and p.Leu1007Profs*2) and NRLP12 (p.F402L); patients carrying two VUS across the same or different genes generally experienced a higher burden of inflammatory symptoms and responded well to colchicine or IL-1 inhibitors.

Conclusions:

Demographical data (n=40)	
Age population (mean ± SD)	36.1 ± 14.2
M:F	14:26
Clinical features	
Recurrent Fever	35 (87.5%)
Skin rash	17 (42.5%)
Musculo-skeletal involvement (arthritis, arthralgias/myalgias)	24 (60%)
Serositis (Pleuritis, pericarditis) and/or peritonitis (eg. Recurrent chest/abdominal pain)	16 (40%)
Headache	7 (17.5%)
Lymph nodes enlargement	5 (12.5%)
Aphthosis	8 (20%)
Other inflammatory symptoms	16 (40%)
Therapy	
Colchicine	11 (27.5%)
Anakinra	2 (5%)
Canakinumab	3 (7.5%)
Combination th (IL-1i + colchicine)	5 (12.5%)
Combination th (PDN + colchicine)	1 (2.5%)
Combination th (ruxo + anakinra)	1 (2.5%)
Other	4 (10%)
Off therapy	13 (32.5%)

Although patients with VUS remain classified as “Systemic Undifferentiated Recurrent Fever”, emerging clusters of patients sharing the same variant and clinical profiles suggest the possibility of “novel” gene-related pathologies. Further investigations are essential for enhancing the diagnostic framework for VUS carriers.

Keywords: genetics, autoinflammation, recurrent fevers

PV136 / #546

POSTER SESSION 12: AUTOINFLAMMATORY DISEASES
03-08-2025 10:30 AM - 11:30 AM

A WINDOW OF OPPORTUNITY IN STILL'S DISEASE: THE EARLY INTRODUCTION OF IL-1 INHIBITORS

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Background and Aims: According to the recent EULAR/Pres Recommendations for Still's disease (SD), the employment of IL-1 inhibitors(i) is crucial to reduce the glucocorticoids intake (GC), prevent flares and achieve Clinical Inactive Disease (CID). We gathered patients with SD diagnosed between 2010 and 2024 to observe the efficacy and safety of early IL-1i introduction.

Methods: One-hundred and nine SD patients (Yamaguchi's criteria) were screened; CID achievement at six months (6M) was set as primary outcome. Chi square test was employed as statistical test and $p<0.05$ was considered significant. GraphPad Prism8 was used for statistical analysis.

Results:

Demographical data (n=42)	
Age population (mean ± SD)	46,7 ± 16,8
Age at disease onset (mean ± SD)	39 ± 16,4
Age at diagnosis (mean ± SD)	39 ± 16,4
M:F	20:22
Median time from symptoms onset to IL-1 treatment start, in months (IQR)	6 (IQR 15)
Median time from diagnosis to treatment start, in months (IQR)	1 (IQR 8)
Clinical features	
Pyrexia (>38°TC)	42 (100%)
Skin rash (transient pink-salmon or maculopapular)	23 (55%)
Pharyngodynia	21 (50%)
Arthritis	15 (36 %)
Arthralgia/myalgia	40 (95,2%)
Hepato/splenomegaly	13 (31,7%)
Lymph nodes enlargement	8 (19 %)
Serositis (Pleuritis, pericarditis) and/or myocarditis	5 (3 pleuropericarditis, 1 myopericarditis, 1 abdominal and chest serositis) (18.5%)
MAS at onset	6 (23%)
Laboratory biomarkers	**Value at diagnosis (mean±SD)**
Haemoglobin (g/L)	96 g/L
White blood cells (mmc3/L)	18649±9276.89 mmc3/L
PCR (g/L)	159,45 ±142,5
Ferritin (mcg/L)	27 267 mcg/L±57603
Glycosylated ferritin (%)	20,4%
Transaminases (AST/ALT)	230/216 U/L
PET-CT or PET-MR hyper-uptakes	**Patients (n=25) (59%)**
Bone marrow	13 (52%)
Lymph nodes	15 (60%)
Spleen	6 (24%)
Joints	4 (16%)
Current therapy	
Anakinra	27 (64%)
Anakinra + MTX	4 (9,5%)
Anakinra + Ruxolitinib	1 (2%)
Canakinumab	6 (14%)
Other	1 (2%)
Off therapy	4 (9,5%)

We obtained data from 42 of the sixty-four SD IL-1i treated as shown in Figure 1. 27/42

(64%) received the diagnosis within 6M from symptoms start. In 24/42 (57%) patients, IL-1i was given within 6M from disease onset, whereas in 18 (43%) patients, it was started >6M. Overall, CID at 6M was achieved by 23/42 (55%) patients; despite not significant (p=0.073), of the 24 patients that introduced IL-1i within 6M, 16 (67%) achieved CID at 6M, while of those treated later than 6M from disease onset, CID was achieved by 7 (38%); severe flares recurred after IL-1i discontinuation in three patients, while two had a refractory form. No serious adverse events raised during IL-1i therapy.

Conclusions: CID was achieved at 6M by a higher percentage of those early-treated than those late-treated with IL-1i. We did not observe serious adverse events related to therapy and flares occurred mostly after IL-1i discontinuation

Keywords: autoinflammation, Interleukin 1, anakinra

PV137 / #575

POSTER SESSION 12: AUTOINFLAMMATORY DISEASES
03-08-2025 10:30 AM - 11:30 AM

IDENTIFYING PREDICTORS OF THE NEED FOR IN-DEPTH EXAMINATION OF PATIENTS WITH STILL'S DISEASE

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Background and Aims: Genetic testing plays an important role in the diagnosis of autoinflammatory diseases (AIDs), but it isn't universally available method. We compare groups of patients with monogenic (mAIDs) and polygenic (pAIDs) AIDs to identify markers of the need for a deeper examination.

Methods: The study included patients admitted to our department from 02.2023 to 12.2024, who were diagnosed with mAIDs based on complete genomic sequencing, pAIDs according to internationally accepted criteria.

Results: 40 patients were diagnosed with pAIDs (Figure 1

Figure 1: Polygenic autoinflammatory diseases

1		Previous diagnosis	Duration before diagnosis	Age of debut (year)	Fever: not intermittent 1, intermittent 2	Arthritis: poly 1, oligo	Sacroiliitis	Skin	Ulcers	Sore thro	Lymph adenopathy	Serositis	Lungs	Intestinal	Uveites	Leukocytosis	Count (10x1 *9/l)	Cytopenia	High CR	High AST, ALT	Ferritin (µg/L
2	Still	0	2	25	1	2	n/d	0	0	1	1	0	0	0	0	1	11	0	1	1	28
3	Still	0	2	36	1	1	n/d	1	0	1	1	0	0	0	0	1	12	0	1	1	58
4	Still	0	0	32	1	1	n/d	0	0	1	1	0	0	0	0	1	18	0	1	0	287
5	Still	0	0	24	1	1	n/d	1	0	0	1	0	0	0	0	1	12	0	1	1	122
6	Still	0	0	45	1	1	n/d	1	0	1	0	1	0	0	0	1	10	0	1	1	76
7	Still	0	0	27	1	1	n/d	1	0	1	1	0	0	0	0	1	10	0	1	1	68
8	Still	SpA	3	35	1	1	n/d	1	0	1	1	0	0	0	0	1	21	0	1	1	1890
9	Still	SpA	2	55	1	2	n/d	1	0	1	1	1	0	0	0	1	9	0	1	1	19
10	Still	0	0	38	1	2	n/d	1	0	1	1	1	0	0	0	1	22	0	1	1	2860
11	Still	0	1	65	1	1	n/d	1	0	1	1	1	0	0	0	1	12	0	1	0	1802
12	Still	0	0	33	1	1	n/d	1	0	1	1	1	0	0	0	1	19	0	1	1	1809
13	Still	0	0	26	1	1	n/d	1	0	0	1	0	0	0	0	1	16	0	1	1	329
14	Still	0	1	23	1	1	n/d	1	0	1	1	0	0	0	0	1	14	0	1	1	1768
15	Still	0	0	27	1	1	n/d	0	0	0	1	1	0	0	0	1	11	0	1	0	112
16	Still	SpA	4	22	1	2	0	1	0	0	1	0	0	0	0	1	11	0	1	0	497
17	Still	SpA	7	32	1	1	n/d	1	0	1	1	1	0	0	0	1	10	0	1	0	125
18	Still	0	0	39	1	1	n/d	1	0	1	1	1	0	0	0	1	18	0	1	1	1726
19	Still	0	0	39	1	0	n/d	1	0	1	1	0	0	0	0	1	13	0	1	1	3577
20	Still	PA	1	58	1	1	n/d	1	0	0	0	0	0	0	0	1	15	0	1	1	613
21	Still	0	0	26	1	2	n/d	1	1	0	1	1	0	0	0	0	6	1	1	1	5561
22	Still	0	0	37	1	1	n/d	1	0	1	1	0	0	0	0	1	10	0	1	1	2744
23	Still	0	0	55	1	2	n/d	1	0	1	0	0	0	0	0	1	10	0	1	0	474
24	Still	0	0	34	1	2	n/d	1	0	1	1	0	0	0	0	1	14	0	1	0	648
25	Still	0	0	75	1	1	n/d	0	0	1	0	1	0	0	0	1	14	0	1	0	2362
26	Still	0	2	28	1	1	n/d	1	0	0	1	0	0	0	0	1	9	0	1	0	1598
27	Still	0	1	38	1	2	n/d	1	0	1	1	0	0	0	0	1	9	0	1	0	210
28	Still	0	7	43	1	1	n/d	1	0	1	0	0	0	0	0	1	18	0	1	1	315
29	Still	0	0	43	1	1	n/d	1	1	1	1	1	0	0	0	1	11	0	1	1	98
30	Still	0	1	54	1	1	n/d	1	0	0	1	0	0	0	0	1	10	0	1	0	427
31	Schnitzler	urticaria	5	52	2	1	n/d	1	0	0	0	0	0	0	0	1	12	0	1	0	112
32	Schnitzler	urticaria	7	45	2	0	1	1	0	0	1	0	0	0	0	1	10	0	1	0	76
33	Schnitzler	urticaria	22	38	2	0	1	1	0	0	0	0	0	0	0	0	7	0	1	0	48
34	Schnitzler	Still	10	40	2	2	1	1	0	0	1	0	0	0	0	1	11	0	1	0	120
35	Schnitzler	SpA	18	31	2	1	1	1	0	0	0	0	0	0	0	0	6	0	1	0	162
36	Schnitzler	Still	6	55	2	1	1	1	0	0	0	0	0	0	0	1	14	0	1	0	672
37	Bechchet	SpA	38	5	2	1	n/d	1	1	0	1	0	0	0	0	1	11	0	1	0	48
38	Bechchet	RA	4	15	0	2	1	1	1	1	1	0	0	0	0	1	10	1	1	0	58
39	Bechchet	SpA	17	5	0	2	1	0	1	0	0	0	0	0	0	1	12	0	1	0	14
40	Bechchet	RA	15	15	1	0	n/d	0	1	0	0	0	0	1	1	0	8	0	1	0	409
41	Bechchet	vasculitis	1	52	1	2	1	1	1	0	1	0	0	1	1	1	10	0	1	0	404
42	SpA - spondylarthritis, RA - rheumatoid arthritis, n/d - no data																				

): 29 - Still's disease (SD), 6 - Schnitzler (SS), 5 - Bechchet (BD); 22 were mAIDs (Figure 2

Figure 2: Monogenic autoinflammatory diseases

Diagnosis	Previous diagnosis	Duration before diagnosis (year)	Age of debut (year)	Fever: not intermittent 1, intermittent 2	Arthritis: poly 1, oligo 2	Sacroiliitis	Skin	Ulcers	Sore throat	Lymphadenopathy	Serosites	Lungs	Intestinal	Uveites	Leukocytosis	Count (10x1*9/l)	Cytopenia	High CRP	High AST, ALT	Ferritin (µg/L)
PAMI	SpA	40	7	0	2	1	1	0	0	1	1	0	0	0	0	5	1	1	0	53
TRAPS	Still	6	18	2	0	n/d	1	0	1	0	0	0	0	0	1	11	0	1	0	375
MEFV	Gout	26	50	2	2	n/d	1	0	0	0	1	0	0	0	0	7	0	1	0	40
NLRP3	0	17	26	2	2	n/d	1	1	1	1	1	0	0	0	0	6	0	0	0	23
VEXAS	SLE	20	34	1	2	1	1	0	0	0	1	1	0	0	0	5	1	1	0	186
MEFV	SpA	24	25	2	2	1	0	0	0	1	1	0	0	0	0	5	0	1	0	64
STAT	Bechchet	0,5	30	1	0	n/d	1	1	1	0	0	0	0	0	0	6	0	1	0	348
CYBB	0	7	30	0	0	1	1	1	0	1	0	1	1	1	0	8	0	1	0	122
VEXAS	SLE	3	56	1	0	1	1	0	0	1	0	1	0	0	0	3	1	1	0	455
MEFV	Gout	20	28	0	2	n/d	0	0	0	0	1	0	0	0	0	5	0	1	0	311
MEFV	SpA	0	6	2	2	1	0	0	0	0	1	0	0	0	0	7	0	1	0	175
MEFV	Still	4	1	2	2	n/d	0	0	0	1	1	0	0	0	0	4	0	1	0	38
NLRP3	SpA	20	26	2	1	n/d	1	0	0	0	0	0	0	1	0	4	0	1	0	122
NLRP3	RA	33	1	2	2	n/d	1	0	0	0	0	0	0	1	0	6	0	1	0	105
MEFV	0	10	5	2	0	n/d	0	0	0	0	1	0	0	0	0	5	0	1	0	255
MEFV	Still	10	14	2	2	n/d	0	0	1	1	0	0	0	0	1	10	0	1	0	122
MEFV	0	2	6	2	2	n/d	0	0	0	0	1	0	0	0	0	7	0	1	0	24
MEFV	Gout	21	5	2	0	n/d	0	0	0	1	1	0	0	0	0	6	0	1	0	204
NOD2	0	10	28	0	1	n/d	0	0	0	0	0	0	1	0	0	4	0	0	0	18
SAVI	SLE	25	18	1	2	0	1	1	0	0	0	0	0	0	0	5	0	0	0	48
NLRP3	Bechchet	20	5	2	2	1	1	1	0	0	1	0	1	1	0	6	0	1	0	77
NLRP3	Bechchet	35	10	2	2	1	1	1	0	0	1	0	0	1	1	11	0	1	0	133

SpA - spondylarthritis, RA - rheumatoid arthritis, SLE - systemic lupus erithematosis, n/d - no data

). The mean time to diagnosis was (years): 1 SD, 11 SS, 15BD, 16 mAIDs. For clinical characteristics of patients see figures. According to the analysis to the Whitney mana criterion ($p<0,01$) between groups (SD, SS, BD, mAID), it was noted for patients with SD have: a higher average value of ferritin (1110/198/186/149 µg/L), leukocytes (13/10/10/6 x10*9/L), polyarticular syndrome (66/0/20/9%), pain in throat (72/0/20/18%), NOT intermittent fever (100/0/40/18%), shorter delay time of diagnosis (1/11/15/16 years), increase in AST, ALT (60/0/0/0%). All SS have signs of sacroiliitis according to X-ray or MRI, intermittent fever, oligoarthritis, but we have too small a sample of patients to talk about a reliable difference.

Conclusions: Chronic course of the disease, lack of sore throat, modest elevation ferritin, leukocytosis, absence of elevated AST, ALT, presence of extra-articular manifestations, sacroiliitis, intermittent fever, oligoarthritis is likely a reason for thorough examination, including consideration of genetic testing patients with signs of AIDs.

Keywords: genetics, autoinflammation, Still's disease

PV138 / #146

POSTER SESSION 13: AUTOIMMUNITY AND BIOMARKERS
03-08-2025 1:55 PM - 2:55 PM

NATURAL AUTOANTIBODIES TARGETING G PROTEIN-COUPLED RECEPTORS: KEY REGULATORS OF IMMUNE RESPONSE AND AUTOIMMUNE DISEASES

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Background and Aims: G protein-coupled receptor (GPCR) autoantibodies play a critical role in regulating immune responses. We propose that GPCR autoantibodies influence the location, intensity, and composition of immune responses pivotal in various conditions. Assessing their levels and functions is essential for understanding inflammatory diseases and advancing pharmaceutical innovation.

Methods: This review synthesizes our recent findings on autoantibodies targeting GPCRs, specifically the angiotensin receptor type-1, chemokine receptor CXCR3, and thrombin receptor PAR1. We also present data on IgG antibodies to explore disease-specific pathways that may uncover new therapeutic targets.

Results: GPCR autoantibodies appear tightly regulated, with both low and high levels linked to diseases. Low levels correlate with vascular diseases, supporting their role in modulating immune responses at barrier sites. High levels are associated with inflammation in organs with abundant GPCR expression. These antibodies resist conventional immunosuppressive therapies, such as Rituximab and stem cell transplantation, suggesting they reflect long-term memory of environmental factors. They not only modulate receptor-ligand interactions but also exhibit unique effects on their targets. The impact of GPCR autoantibodies is most extensively studied in systemic sclerosis (SSc), pulmonary arterial hypertension (PAH), and atherosclerosis, but abnormal levels are also implicated in cancer, fibromyalgia, and chronic fatigue syndrome. IgG function studies demonstrate disease-specific signaling, offering insights into therapeutic pathways and patient responses.

Conclusions: GPCR autoantibodies are essential for understanding the mechanisms of inflammatory and autoimmune diseases. Their study should be prioritized in both basic and pharmacological research to identify novel therapeutic targets and develop more precise treatments.

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POSTER SESSION 13: AUTOIMMUNITY AND BIOMARKERS
03-08-2025 1:55 PM - 2:55 PM

COMPARATIVE EVALUATION OF TWO COMMERCIALLY AVAILABLE ANTI-DOUBLE-STRANDED DNA IMMUNOASSAYS FOR THE DIAGNOSIS OF SYSTEMIC LUPUS ERYTHEMATOSUS

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Background and Aims: Background and aim: The determination of anti-dsDNA antibodies plays a significant role in diagnosis and management of SLE. According to the 2019 EULAR/ACR Classification Criteria for SLE, assay specificity ≥ 90% is required for use in the classification scoring system. In this study, we evaluate and compare the diagnostic performance of two available anti-dsDNA immunoassays.

Methods: Methods: Serum samples from 50 SLE patients and 100 disease controls were obtained from a sample bank. Within the SLE group, 11 patients had a diagnosis of lupus nephritis. Disease controls included cases of RA, SjS, SScl, IIM, MCTD, infections, and tumors. Samples were evaluated using the EliA™ dsDNA (Thermo Fisher Scientific, Germany) and Quanta Flash® dsDNA (Werfen, San Diego) assays. Pre-defined manufacturer cut-offs of 15 IU/mL for EliA™ and 35 IU/mL for the Quanta Flash® assays were used.

Results: Results: The two assays demonstrated 91% sensitivity in the lupus nephritis sub-population, and the overall sensitivity was 30% for EliA™ and 38% for Quanta Flash®. Specificities were 99% for EliA™ and 89% for Quanta Flash®. False positive results on the Quanta Flash® assay clustered to patients with RA, MCTD, or SjS, and the EliA™ assay reported a false positive result for a SjS case.

Conclusions: Conclusions: Analysis of predefined patient samples and disease controls indicates that the EliA™ dsDNA assay is more specific but less sensitive than the Quanta Flash® dsDNA. However, according to these results, the Quanta Flash® dsDNA assay does not meet the specificity threshold required by the ACR/EULAR for classification purposes.

Keywords: systemic lupus erythematosus, anti-dsDNA antibodies, comparison

PV140 / #210

POSTER SESSION 13: AUTOIMMUNITY AND BIOMARKERS
03-08-2025 1:55 PM - 2:55 PM

ENHANCED DETECTION OF AUTOANTIBODIES IN IDIOPATHIC INFLAMMATORY MYOPATHIES: A COMPARISON OF MICROBLOT ARRAY AND STANDARD DIAGNOSTIC METHODS

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Background and Aims: Idiopathic inflammatory myopathy (IIM) encompasses dermatomyositis (DM), polymyositis (PM), overlap myositis (OM), anti-synthetase syndrome (ASS), sporadic inclusion body myositis (IBM), and immune-mediated necrotizing myopathy (IMNM). These autoimmune conditions cause muscle inflammation, organ dysfunction, and increased morbidity and mortality. Autoantibodies are crucial for diagnosis and pathogenesis. Traditional ANA tests like immunofluorescence (IFA) and BLOT have limitations in antigen distribution, sample volume, and accuracy. The Microblot-Array (MBA) ANA enables simultaneous analysis of 44 markers, offering a comprehensive view of autoimmune diseases (AIDs).

Methods: The study evaluated the diagnostic accuracy of the MBA in ANA diagnostics. Antibody distribution was assessed in sera from 423 patients (319 female, 104 male) with idiopathic inflammatory myopathies, stored at the Institute of Rheumatology. Routine methods included IIF (IMMUNOCONCEPT) and Myositis Blot (Euroimmun). The goal was to determine the correlation between MBA, traditional methods, and the clinical presentation.

Results: The study demonstrated a strong concordance between myositis-specific (MSA) and myositis-associated (MAA) antibodies detected by MBA and traditional diagnostic methods, confirming MBA's reliability. In ASS, antibodies were found in 26 patients, correlating strongly with IIF results. Additionally, anti-HMGCR antibodies were detected by MBA in 100% of the 53 IMNM patients. These results were fully confirmed by ELISA (Werfen), highlighting the test's high sensitivity.

Conclusions: MBA aligns well with BLOT and IFA, significantly enhancing ANA diagnostics. Its ability to detect multiple antibodies in a single test improves diagnostic accuracy and

efficiency, leading to faster, more personalized treatment options and better long-term patient outcomes. **Acknowledgements** BBMRI-CZ LM2018125

PV141 / #252

POSTER SESSION 13: AUTOIMMUNITY AND BIOMARKERS
03-08-2025 1:55 PM - 2:55 PM

NON-CRITERIA ANTIPHOSPHOLIPID ANTIBODIES IN PATIENTS WITH CLINICAL CRITERIA FOR ANTIPHOSPHOLIPID SYNDROME: MULTICENTER STUDY

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Background and Aims: International consensus for antiphosphiolipid syndrome (APS) classification considers clinical diagnostic criteria and the presence of lupus anticoagulant, IgG and IgM anticardiolipin antibodies and IgG and IgM anti beta-2-glycoprotein I antibodies. Over the past few years, much attention has been focused on the prevalence of non-criteria antiphospholipid antibodies (aPL) in association with different clinical categories of APS. We aimed to conduct a preliminary multicenter study of the prevalence of non-criteria aPL in cohorts of patients with clinical classification criteria of APS.

Methods: Patients from 2 centres were evaluated: University Hospital Puerta de Hierro Majadahonda and University Hospital Gregorio Marañon. aPL were analysed by ELISA technique, with a fully automated Alegria 2 (ORGENTEC by Sebia) instrument that offers an autoimmune panel where each test is validated by its own integrated calibrator and control for accuracy and reliability.

Results: Sixty five patients were evaluated with the following distribution: 45 with obstetric criteria (mean age 36 years, range 26-42 years), 20 with thrombotic criteria (mean age 60 years). Non-criteria aPL were detected in 13 patients (20%): anti-phosphatidylserine IgG antibodies in 8 (12.3%), anti-cardiolipin IgA in 1 (1.5%), beta-2-glycoprotein I IgA in 3 (4.6%) and annexin V IgG in 2 (3.1%). In 6 patients non-criteria aPL were not associated with any classical aPL specificity (9.2%).

Conclusions: Non-criteria aPL are detected in about 10% of patients with APS clinical criteria in the absence of classical aPL. The positioning of this aPL is very important for the eventual prophylactic or therapeutic coverage that these patients may require.

Keywords: antiphospholipid antibodies, Antiphospholipid Syndrome, laboratory criteria

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POSTER SESSION 13: AUTOIMMUNITY AND BIOMARKERS
03-08-2025 1:55 PM - 2:55 PM

MACHINE LEARNING AND ARTIFICIAL INTELLIGENCE IN ADULT AUTOIMMUNE DISEASES: APPLICATIONS, CHALLENGES, AND FUTURE PERSPECTIVES

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Background and Aims: Autoimmune diseases (ADs) in adults, including rheumatoid arthritis, systemic lupus erythematosus, and multiple sclerosis, present significant diagnostic and therapeutic challenges due to their multifaceted and often overlapping clinical manifestations. Machine learning (ML) and artificial intelligence (AI) are emerging as powerful tools capable of analyzing extensive datasets to identify clinically relevant patterns that may aid in diagnosis and treatment. This study aims to explore the current applications of AI and ML in diagnosing, managing, and discovering biomarkers for adult autoimmune diseases, while also addressing the challenges faced in their effective clinical implementation.

Methods: A comprehensive review was conducted by searching PubMed, Scopus, and Web of Science for studies published between 2010 and 2024. Inclusion criteria focused on original research articles, systematic reviews, and meta-analyses investigating AI and ML in adult ADs. Key applications, such as diagnostic imaging, disease activity prediction, and treatment response, were analyzed. Data extraction followed PRISMA guidelines.

Results: Forty-five studies met the inclusion criteria. AI and ML applications significantly improved diagnostic accuracy, particularly in early-stage rheumatoid arthritis and lupus nephritis, with models showing sensitivities and specificities exceeding traditional methods. Predictive models for disease activity and personalized treatments showed promise, though challenges such as data heterogeneity, algorithm transparency, and clinical workflow integration persist.

Conclusions: AI holds great potential in managing adult autoimmune diseases by enabling early diagnosis, individualized treatments, and novel biomarker discovery. Further research is necessary to overcome limitations and ensure AI's integration into clinical practice.

Keywords: autoimmune diseases, Machine learning, Artificial Intelligence

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POSTER SESSION 13: AUTOIMMUNITY AND BIOMARKERS
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REFRACTORY ANTIPHOSPHOLIPID ANTIBODY SYNDROME, OTHER RISK FACTORS FOR THROMBOSIS. A CASE REPORT.

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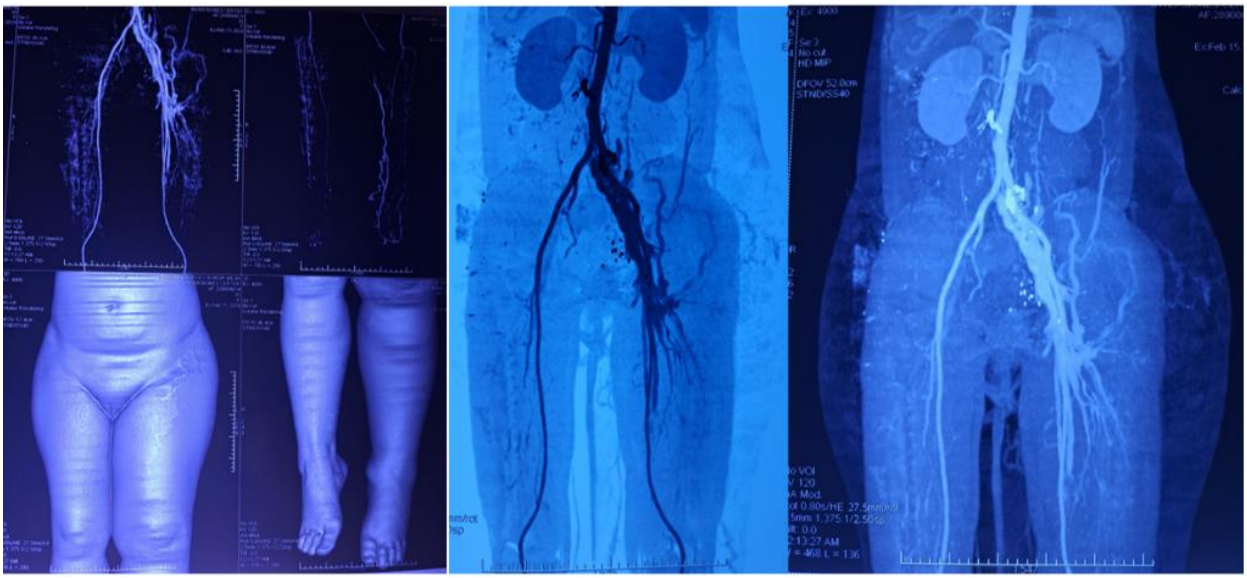
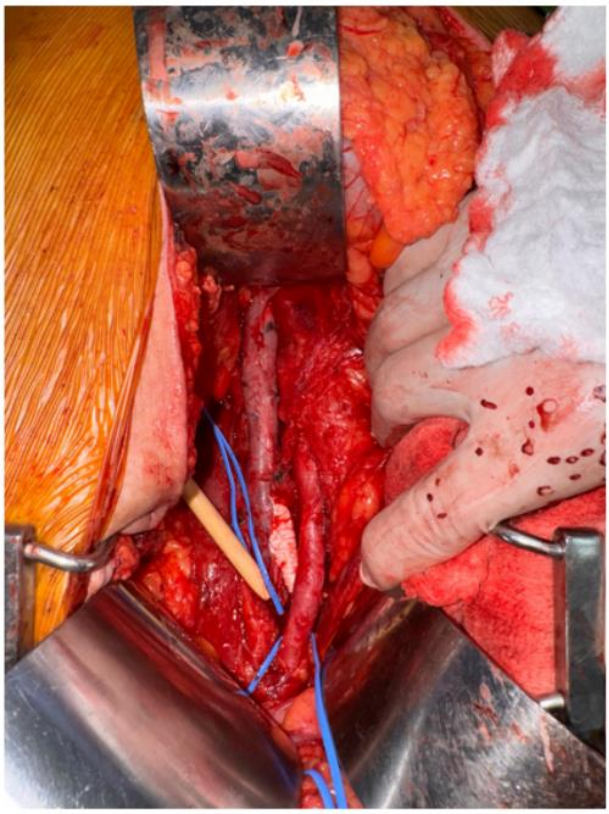
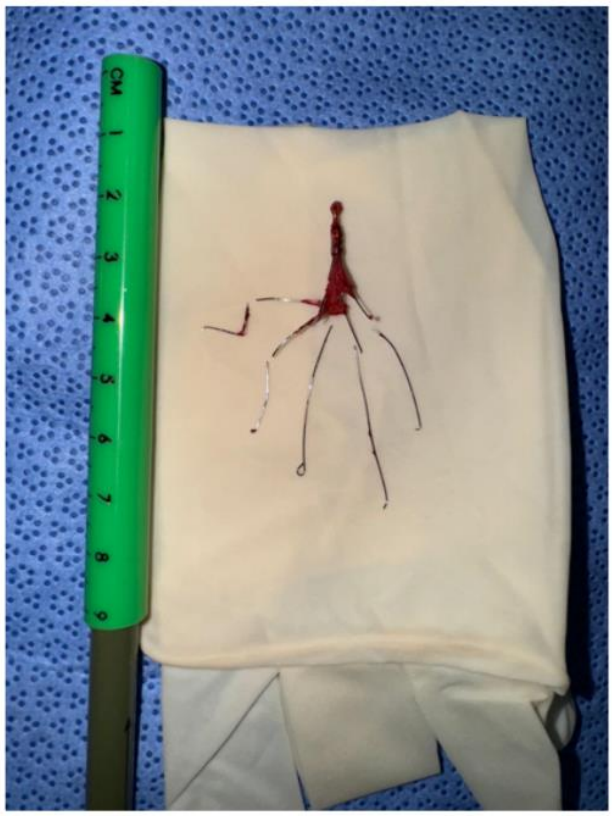
Background and Aims: Refractory APS can be defined as a breakthrough thrombosis during standard oral anticoagulant therapy. Management is challenging and is conducted by extrapolating the management of similar cases.

Methods: A 35-year-old female patient with APS, with a history of obstetric losses, triple positivity for aPL, three thrombotic episodes of the left common femoral vein, May Turner syndrome with stent placement at the left lower limb, and the presence of an inferior vena cava filter; she received LMWH, warfarin, and rituximab. She presented to the emergency with a history of edema, pain, and an increase in the diameter of the left leg. A CT angiography was performed, which showed a thrombotic event (Image 1). It was considered as a refractory APS.



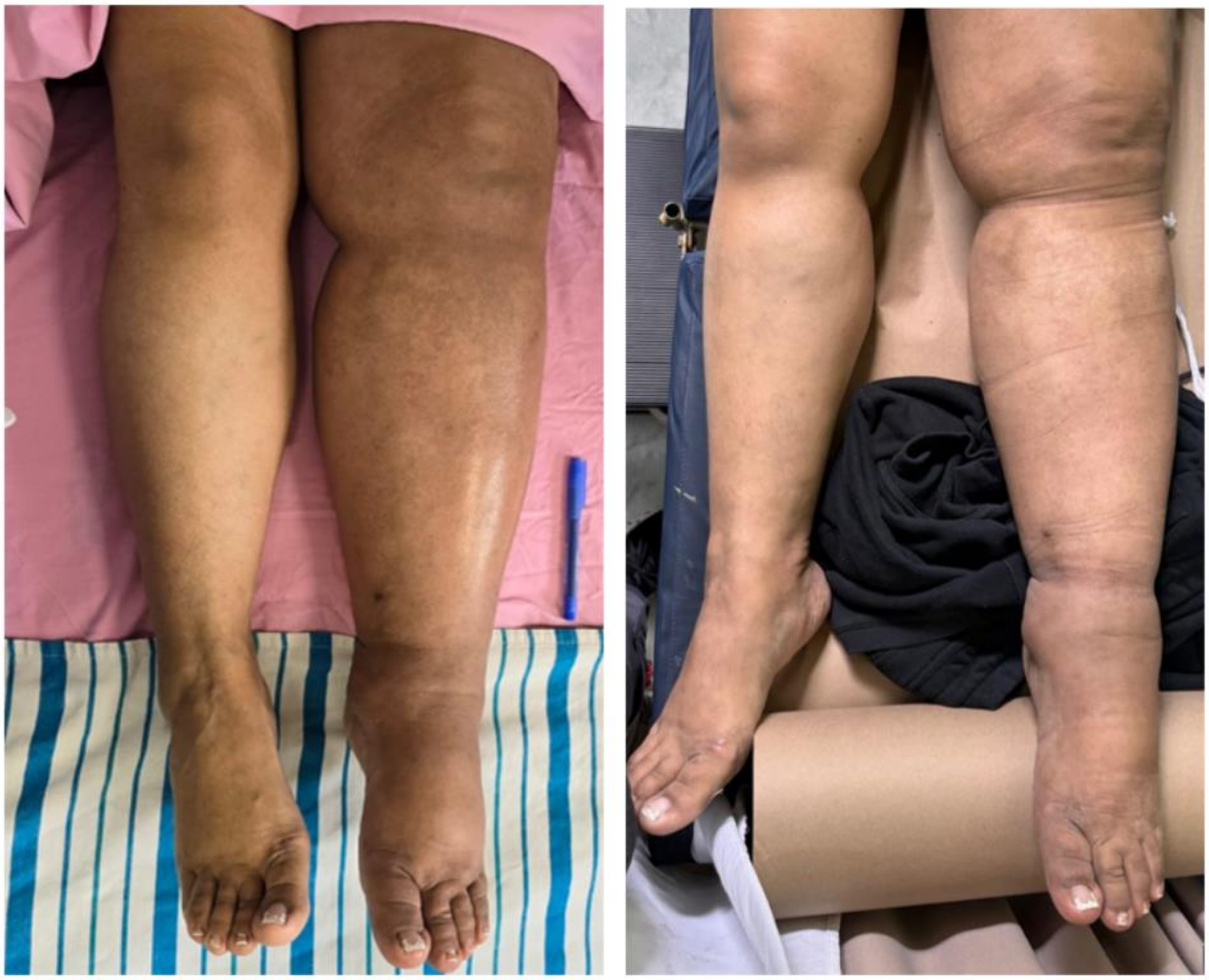
Results: During the Vascular Surgery assessment, migration of the vena cava filter towards the aorta was established; there was no passage of contrast medium from the left common femoral vein to the inferior cava, with dilated accessory venous pathways. She was taken to exploratory laparotomy to extract the vena cava filter, primary cavorrhaphy, and iliocaval reconstruction with a PTFE graft, with adequate passage of contrast medium. **(Image 2)**





A diagnosis of refractory APS with mechanical obstruction was made. After five months, the patient's left lower limb diameter decreased, she experienced no pain, and his mobility

improved. **(Image 3)**



Conclusions: The mechanisms leading to the thrombotic phenotype in APS suggest that anticoagulation alone may not control thrombosis. Therefore, establishing other risk points, such as mechanical associations, can represent up to 6% of thrombosis in these cases.

Keywords: Refractory, Antiphospholipid antibody syndrome, vascular surgery

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POSTER SESSION 13: AUTOIMMUNITY AND BIOMARKERS
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ASSOCIATION OF IL-17 WITH LUNG AND SKIN FIBROSIS IN PATIENTS WITH SYSTEMIC SCLEROSIS AND INTERSTITIAL LUNG DISEASE.

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Background and Aims: Try to evaluate the relationship of level IL-17 with lung and skin fibrosis in pts with SSc and interstitial lung disease (ILD).

Methods: Serum levels of IL-17,IL-6 and TGF- β were examined by ELISA and were measured in two points (mean age at the time of inclusion in study was 49.5±13.1,fem 49(79%),with diffuse form 64,5%, the average follow-up duration was18.7±14 months).All of the pts underwent HRCT.HRCT patterns were presence of reticulations, honeycombing, ground-glass opacity.The pts underwent spirometry and measurement of DLCO.

Results: We didn't find correlation between levels of TGF- β and IL-17.We found a negative correlation level of IL-6 with mean dates of DLco(R=-0.28($p<0.05$)) in first point(FP) and IL-6 in FP with mean dates of FVC in second point(SP)(R=-0.28($p<0.05$)).We didn't find any correlations of IL-17 with dates of FLT, but found correlation level of IL-17 in FP with level of IL-6 in both point(R=0.248 and R=0.397($p<0.05$) accordingly) and IL-17 with level of IL-6(R=0.57($p<0.05$)) in SP.It's worth noting that level of IL-17 had better correlations with level of IL-16 in pts with diffuse form, than in limitation form.Also we found correlation level of IL-17 in both points with digital ulcers and necrosis in SP(R=0.306, R=0.414 and R=0.358 and R=0.267($p<0.05$) accordingly).

Conclusions: Thus the level of IL-17 was indirectly association with lung fibrosis via level of IL-6, but we found better association with skin fibrosis in pts with SSc-ILD.It needs to

further explored in longitudinal studies of SSc-ILD patients to assess in validity as a biomarker and future treatment target.

Keyword: systemic sclerosis, lung fibrosis, IL-17

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POSTER SESSION 13: AUTOIMMUNITY AND BIOMARKERS
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ROLE OF SERUM LEVELS IL-13 IN SYSTEMIC SCLEROSIS PATIENTS WITH INTERSTITIAL LUNG DISEASE.

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Background and Aims: Our aim was to test change of IL-13 in patients with SSc and various patterns of interstitial lung disease (SSc-ILD). the role of IL-13 relevant to specific organ system disease in SSc would be discussed.

Methods: Serum levels of IL-13 were measured in 62 patients with SSc in two points (mean age at enrollment was 49.5 ± 13.1, fem 49 (79%), with diffuse form 64,5%, the average follow-up duration was18.7±14 months). All of the pts underwent HRCT on 1.0-1.5 mm thick overlapping sections using a high -spatial-frequency reconstruction algorithm, which were taken during a single breath hold using various computed tomography scanners. The HRCT images were evaluated by experienced radiologist. HRCT patterns were presence of reticulations, honeycombing, ground-glass opacity.

Results: Mean dates of serum levels of IL-13 were 5.74 ± 25.12, med 0,001 [0.001;0.01] and 2.0 ± 8.82, med 0,001 [0.001;0.001]. Serum level of IL-13 at the first point correlated with level of TGF-β at the second point (R=0.292 ($p<0.05$)) and was significantly decreased with reticulations (p=0.04), ground-glass opacity (p=0,04) and tended to decrease without honey combing (p=0.064). We also found a negative correlation level of IL-13 with total dose of cyclophosphamide (R=-0.248 ($p<0.05$)).

Conclusions: Our dates indirectly confirm that IL-13 indirectly induces fibrosis by activating macrophages that produce TGF-β. A molecular understanding of pathogenic pathways is expected to lead to the development of novel therapeutic strategies aimed at

targeting these cells and the pathways governing abnormal expression of the cytokines they produce.

Keyword: systemic sclerosis, lung, IL-13

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POSTER SESSION 13: AUTOIMMUNITY AND BIOMARKERS
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REVOLUTIONIZING GRD DIAGNOSIS: NOVEL EPITOPES AND MULTIPARAMETRIC DIAGNOSTICS TO ELIMINATE UNNECESSARY BIOPSIES

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Background and Aims: Autoantibody profiles are a powerful tool to correlate clinical features and therapeutic outcomes in GLUTEN-RELATED DISORDERS (GRD). The AESKUBLOTS® GRD IgA is a novel membrane-bound multiparametric assay for the overall quantitative determination of IgA antibodies against human tissue transglutaminase (tTG), neo-epitopes of human tTG (tTG-neo), microbial transglutaminase (mTG), neo-epitopes of mTG (mTG-neo), deamidated gliadin-specific peptides (DGP), gliadin, Frazer's fraction, human epidermal transglutaminase (TG3) and total-IgA in plasma or serum. Therefore, the performance of this novel test system in a study cohort of pediatric GRDs was evaluated.

Methods: Antibody titers of AESKUBLOTS® GRD IgA were evaluated in a cohort of 74 pediatric CD, 60 Dermatitis-Herpetiformis patients, and a control group of 51 diseases not related to GRD. Results were correlated to EMA Endpoint titer (EPT), and clinical history of patients.

Results: Highest AUC of antigens was > 0.95 ($p<0.001$) for tTG IgA, followed by tTG-neo IgA and mTG-neo IgA on the AESKUBLOTS® GRD IgA. tTG-neo IgA showed highest correlation with EMA EPT ($r^2>0.75$, $p<0.001$), followed by tTG-IgA and mTG-neo IgA. IgA deficiency was detected in 100% agreement with patient history.

Conclusions: Advantage of multiparametric antibody and autoantibody analysis is time and cost-saving information gain. New antibodies like mTg-neo can be evaluated more quickly and integrated into routine diagnostics. Excellent correlation with well-established markers show its superior performance as screening assay. In conjunction with patient's medical history, professionals can make rapid therapy recommendations or support to avoid unnecessary biopsies, making the AESKUBLOTS® GRD IgA a unique tool.

Keywords: Gluten Related Disorders, Neo-epitope, Multiplex

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POSTER SESSION 13: AUTOIMMUNITY AND BIOMARKERS
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GENERATION OF MABS AGAINST TTG AND MTG NEO EPITOPES

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Background and Aims: Gluten-related diseases occur in 5% of the population. An increase in diagnosis seems to be due to a real increase in the incidence rather than the increased use of food additives, such as microbial transglutaminase (mTG). Gliadins are cross-linked by tissue transglutaminase (tTG) and/or mTG to form complexes, exposing immunogenic neo-epitopes, triggering the production of anti-neo-epitope antibodies. Detection of these antibodies is a powerful tool in early detection of enteric damage in pediatric CD. Anti-neo-epitope transglutaminase antibodies represent a new generation of markers offering several advantages like better diagnostic performance, a higher reflection of intestinal damage, better predictability at an early age, more diverse epitopes, and less false positivity. Recently, we generated monoclonal antibodies specifically recognizing tTG/mTG neo-epitopes.

Methods: In a first-of-its-kind attempt to generate mAbs against tTG/mTG neo epitopes, we injected mice with the tTG-gliadin and mTG-gliadin complexes. The resulting antibodies were tested for specificity using tTG-Neo, tTG, mTG, mTG-Neo, and gliadin ELISAs. Cell lines generating specific mAbs against tTG neo, tTG, mTG, mTG neo, and gliadin were identified and cultured to produce large quantities of the mAbs. These mAbs were purified and stored until further use.

Results: IFA EMA slides using anti-tTG-neo and mTG-neo mAbs revealed novel patterns, previously not observed, and different from the well-known tTG honey-comb pattern, as well as the gliadin pattern.

Conclusions: These new patterns are indicate the recognizing of unique epitopes.The purified mAbs are specific to the neo-epitopes and a unique tool to proof pathogenicity of environmental triggers in autoimmune development in further research projects.

Keywords: microbial Transglutaminase, Neo-epitopes, Autoantibodies Gluten Related Diseases

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POSTER SESSION 13: AUTOIMMUNITY AND BIOMARKERS
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REVIVING ALLERGY MARKERS AND AUTOIMMUNITY

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Background and Aims: Increased IgE levels in patients with systemic lupus erythematosus (SLE) and other autoimmune diseases have been described for the first time almost 40 years ago but largely understudied. Recent studies have shown that increased IgE concentration is not associated with higher prevalence for atopy and allergy in autoimmune diseases patients. Increased IgE levels are associated with anti-IgE isotype antibodies referred as self-damaging autoantibodies with important contribution to disease pathogenesis and progression. This study aims to analyze the casuistry of our clinical laboratory over the past five years, regarding Ig E elevations, correlating with increasing severity of specific type antibodies in SLE, Systemic sclerosis, Myositis, Autoimmune Liver Disease and Inflammatory Bowel Disease.

Methods: Total IgE serum measurement by ImmunoCAP™ Total IgE. ANA screening with IIF in Hep-2 cells (Euroimmun™); ds-DNA by FEIA (Thermofisher ™) with confirmatory IIF with Crithidia luciliae (Euroimmun™). Immunoblotting for SARDS (Euroimmun™). Liver autoimmune diseases study by IIF in Liver mosaic 9 (Euroimmun™) and liver immunoblotting profile by Euroimmun™. Systemic sclerosis and Myositis autoantibody profile by immunoblotting (Euroimmun™).

Results: The authors present 5 years revised casuistic from January 2020 to December 2024 as a reference clinical laboratory center in autoimmune diseases diagnosis focusing on the evidence that IgE plays a significant role in autoimmunity and that IgE autoantibodies are important contributors to disease pathogenesis and progression.

Conclusions: Clinical studies with anti-IgE therapies might bring additional data on IgE autoantibodies pathogenic activity and propose future areas of research in autoimmunity.

Keywords: AUTOANTIBODIES, Autoimmunity diagnosis, Imunoglobuline E

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POSTER SESSION 14: PSORIATIC ARTHRITIS, GOUT & CHECKPOINT INHIBITORS
03-08-2025 1:55 PM - 2:55 PM

HIGH PREVALENCE OF TESTOSTERONE DEFICIENCY IN MEN WITH PSORIATIC ARTHRITIS

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Background and Aims: It is suggested that the presence of a chronic immune-inflammatory rheumatic disease may be a factor increasing the likelihood of developing hypogonadism syndrome in men, and vice versa. **Aim.** To study the incidence of hypogonadism in men with psoriatic arthritis (PsA) and to assess its impact on PsA and comorbidities.

Methods: Materials and methods. A cross-sectional continuous study included 128 men with PsA. The patients underwent determination of their total testosterone levels and subsequent division into subgroups with normal (≥12.0 nmol/l) and reduced levels. An intergroup comparison was conducted.

Results: The incidence of reduced total testosterone levels was 36.7%. Patients with testosterone deficiency were older (47.9±10.3 vs 40.1±12.1; p<0.001), had a higher body mass index (31.2±5.2 kg/m2 vs 27.2±6.1 kg/m2; p<0.001) and were more often obese (48.9% vs 23.4%; p=0.001). They had higher mean glucose levels (5.9±1.39 mmol/l vs 5.34±0.57 mmol/l; p=0.001). Patients with hypogonadism were characterized by higher uric acid levels (402.9±99.3 μmol/L vs 354.0±81.5 μmol/L; p=0.003) and the frequency of hyperuricemia. A lower proportion of HLA-B27 positive patients was noted in the hypogonadism group, as well as a more frequent occurrence of stage III sacroiliitis (p=0.004) and a smaller amplitude of lateral flexion in the spine (10.3±3.3 cm vs. 12.4±4.3 cm; p=0.014).

Conclusions: Hypogonadism was detected in one third of patients with PsA. Decreased testosterone levels were observed in older individuals and were associated with metabolic disorders, as well as with decreased spinal mobility and the presence of III stage of sacroiliitis.

Keywords: psoriatic arthritis, testosterone, hypogonadism

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POSTER SESSION 14: PSORIATIC ARTHRITIS, GOUT & CHECKPOINT INHIBITORS
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RISK OF LYMPHOMA IN PSORIATIC ARTHRITIS PATIENTS: METHOTREXATE VS ANTI-TNF INHIBITORS

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Background and Aims: Background and aim: Psoriatic arthritis (PsA) is a chronic inflammatory disease associated with an increased risk of malignancies, including lymphoma. Immunosuppressive therapies like methotrexate (MTX) and anti-TNF inhibitors are commonly used in PsA management but may further elevate lymphoma risk. Comparative data on lymphoma incidence in PsA patients treated with MTX versus anti-TNF agents remain limited. To assess the risk of lymphoma in PsA patients treated with methotrexate compared to those receiving anti-TNF inhibitors.

Methods: Methods: A retrospective cohort study was conducted in 500 PsA patients treated with either methotrexate (n=250) or anti-TNF inhibitors (n=250). Patients were followed for a mean duration of 6 years, with lymphoma cases confirmed through clinical records and histopathology. Incidence rates were calculated for both groups, and hazard ratios (HRs) were determined using Cox proportional hazard models, adjusting for age, sex, disease severity, and duration.

Results: Results: Lymphoma developed in 6 patients (2.4%) in the methotrexate group and 4 patients (1.6%) in the anti-TNF group. The incidence of lymphoma was higher in the methotrexate group (HR 1.5, 95% CI 0.4–3.6), though the difference was not statistically significant. Adjustments for confounding factors did not substantially alter the results.

Conclusions: Conclusion: Both methotrexate and anti-TNF inhibitors carry a risk of lymphoma in PsA patients, with a slightly higher, though not statistically significant, incidence observed in the methotrexate group. Careful monitoring for lymphoma is recommended in PsA patients undergoing long-term immunosuppressive therapy. Further large-scale studies are warranted to confirm these findings.

Keywords: psoriatic arthritis, lymphoma, methotrexate

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POSTER SESSION 14: PSORIATIC ARTHRITIS, GOUT & CHECKPOINT INHIBITORS
03-08-2025 1:55 PM - 2:55 PM

MUSCULOSKELETAL IMMUNE-RELATED ADVERSE EVENTS IN PATIENTS TREATED WITH CHECK-POINT INHIBITORS

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Background and Aims: Characteristic of musculoskeletal immune-related adverse events (irAE) in patients treated with immune checkpoint inhibitors (ICI) is incomplete. The aim of the study was to characterize musculoskeletal irAE caused by ICI in a case series.

Methods: 24 patients (mean age 56.3±10.6) with female predominance (n=16; 67%) and musculoskeletal manifestations due to ICI therapy (Nivolumab [n=13], Pembrolizumab [n=7], Prolgolimab [n=1], Atezolizumab [n=3] Ipilimumab[n=2]) were observed by rheumatologist.

Results: Musculoskeletal irAE were performed by arthritis (n=21; 88%), rheumatic polymyalgia (n=1; 4%) and periarthritis (n=2; 8%), that manifested 13 [6; 24] weeks after ICI initiation. The knees (n=20; 83%), the ankles (n=14; 58%) and wrist (n=12; 50%) were most often affected joints. Erythrocyte sedimentation rate ≥30 mm/h was found in 13/22 (59%) cases, C-reactive protein >5 mg/l – in 17/21 (81%), positive antinuclear antibody test – in 10/20 (50%), anti–citrullinated protein antibody and rheumatoid factor – in 1/21 (5%). Treatment of MD include non-steroidal anti-inflammatory drugs (n=20), intra-articular corticosteroids (n=3), oral systemic corticosteroids (n=15) with median maximum dose 25 [11; 56] mg daily of prednisone. Arthritis in 12 cases required treatment with >10 mg daily of prednisone. We successfully used methotrexate (n=7) and leflunomide (n=1) as a steroid-sparing agents, while hydroxychloroquine (n=5) and sulfasalazine (n=1) were ineffective. 16 patients required a long-term treatment with corticosteroids and/or disease-modifying anti-rheumatic drugs

Conclusions: inflammatory MD induced by ICI most often represented by arthritis, which requires long-term therapy with glucocorticoids equivalent to 10 mg/day of prednisolone and/or disease-modifying anti-rheumatic drugs.

Keywords: immune-related adverse events, check-point inhibitors, Musculoskeletal disorders

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POSTER SESSION 14: PSORIATIC ARTHRITIS, GOUT & CHECKPOINT INHIBITORS
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LABORATORY FINDINGS IN PATIENTS RECEIVING IMMUNE CHECKPOINT INHIBITORS AND OTHER ANTITUMOR DRUG TREATMENT WITH NEW-ONSET INFLAMMATORY MUSCULOSKELETAL DISORDERS

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Background and Aims: There is a limited data on laboratory findings in patients receiving antitumor drug treatment with new-onset inflammatory musculoskeletal disorders. The aim of the study was to evaluate the ANA, RF, ACPA, ESR and CRP in patients receiving immune checkpoint inhibitors (ICI) and other anticancer drugs with new-onset musculoskeletal disorders

Methods: Tests of ANA (indirect immunofluorescence), RF (nephelometry), ACPA (ELISA), ESR and high-sensitivity CRP were performed in 21 patients receiving ICI (mean age 57±11; M/F – 57% / 43%) and 25 patients receiving other antitumor drugs (mean age 58±13; M/F – 12% / 88%). All patients had rheumatic inflammatory musculoskeletal disorders that debuted during drug antitumor therapy.

Results: ESR≥30 mm/h was in 12 (57%) patients receiving ICI and in 10 (40%) patients receiving other anticancer drugs (p>0.05), median of ESR was 35 [22; 46] mm/h and 24 [13; 49] mm/h respectively (p=0.460). CRP>5 mg/l was in 17 (81%) patients receiving ICI and in 10 (40%) patients receiving other anticancer drugs (p=0.006), median CRP level was 41.5 [6.7; 140] mg/l and 2.2[0.7; 20.5] mg/l respectively (p=0.002). ANA test was positive in 10 (47%) patients receiving ICI and in 13 (52%) patients receiving other anticancer drugs (p>0.05). RF,ACPA were negative in all patients receiving ICI and positive in 3 (13%) and 4 (16%) patients receiving other anticancer drugs respectively.

Conclusions: Laboratory findings in cancer patients receiving drug antitumor therapy with new-onset inflammatory musculoskeletal pathology included an increase in ESR, CRP, ANA.The increase in CRP level in patients receiving ICI was significantly greater than for other patients.

Keywords: immune-related adverse events, check-point inhibitors, Musculoskeletal disorders

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POSTER SESSION 14: PSORIATIC ARTHRITIS, GOUT & CHECKPOINT INHIBITORS
03-08-2025 1:55 PM - 2:55 PM

HEART DAMAGE IN PSORIATIC ARTHRITIS

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Background and Aims: Mortality from cardiovascular disease among patients diagnosed with seronegative spondylitis is constantly increasing. Heart damage in psoriatic arthritis is 18-22% and it is continuously growing.

Methods: The results obtained from the electrocardiogram and echocardiography performed on 75 patients with psoriatic arthritis were collected, established based on the CASPAR criteria, who were being treated in the rheumatology department, during the years 2022-2024.

Results: According to the results of a study, it was determined that most ofen affected valves is the aortic and mitral valves, for each the approximate 37%. Insufficiency of the tricuspid valve had a prevalence of 16.7% and of the pulmonary valve of 8.3%. The data obtained from echocardiography, we determined left ventricular hypertrophy in 37.5%, the left atrial hypertrophy with a prevalence of 29.1%, the right ventricular hypertrophy with 12.5% and right atrial hypertrophy with 4.2%. Patients were examined for rhythm changes by electrocardiogram. The most common ehytm disturbances was atrial fibrillation, approximately 54.5%, followed by ventricular extrasystole 27.2% and supraventricular extrasystole 18.3%. The left bundle branch block of the His bundle is most often attested - 25%, followed by the block of the right branch of the His bundle with a prevalence of 8.3%. Based on the study data, we determined a prevalence of 20.8% among the studied patients who had pericarditis at the time of the study.

Conclusions: Conclusion. The most characteristic heart damage is valve insufficiency in which inflammation plays an important role and inflammatory markers can be additional tools for stratifying cardiovascular risk in these patients.

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03-08-2025 1:55 PM - 2:55 PM

ASSOCIATION OF BODY COMPOSITION WITH SUBCLINICAL MYOCARDIAL IMPAIRMENT IN PSORIATIC ARTHRITIS PATIENTS

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Background and Aims: Subclinical impaired myocardial deformation is common in patients with PsA, even with no clinical evidence for CV disease. Myocardial involvement is associated with disease activity, obesity and other CV risk factors. Biopendance analysis using the phase angle (PhA) is a non-invasive method of assessing body composition that can distinguish fat mass from the fat-free mass.The aim was to assess the association of body composition with subclinical myocardial dysfunction in non-obese PsA patients. Another aim was to determine the relationship between body composition and PsA activity measured by DAPSA.

Methods: 32 PsA patients (13 males) with no clinically evident CV disease or CV risk were included in the study. Upon clinical and laboratory evaluation, DAPSA scoring and biompendance analysis (PhA) all subjects underwent conventional echocardiography and 2-dimensional speckle tracking echocardiography (STE).

Results: Median PhA was 5.25° (4.66° – 5.9°), Global longitudinal strain (GLS) calculated from STE was 20% (19% – 21%) and DAPSA 15.8 (8.6 – 28.8). The median BMI was 25.7 kg/m2 (23.45 - 26.9). STE analysis showed that patients with higher values of PhA had better (higher) GLS. Significant correlation was determined between PhA and DAPSA (Pearson r=-0.4, P=0.026) and PhA and GLS (Pearson r=0.597, P=0.001). In multivariate analysis after adjustment for confounders (age, CRP, LVEF, BMI) only GLS maintained significant association with PhA (P=0.012).

Conclusions: PsA patients with lower PhA values measured by bioimpedance have a higher degree of subclinical myocardial dysfunction measured by GLS. No significant association was found between the PhA and DAPSA score.

Keyword: psoriatic arthritis, myocardial dysfunction, strain, body composition

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POSTER SESSION 14: PSORIATIC ARTHRITIS, GOUT & CHECKPOINT INHIBITORS
03-08-2025 1:55 PM - 2:55 PM

CASE REPORT: UPADACITINIB - A PROMISING THERAPEUTIC APPROACH FOR PSORIATIC ARTHRITIS AND COEXISTING VITILIGO

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Background and Aims: Psoriatic arthritis (PsA) is an autoimmune condition characterized by joint inflammation, often associated with psoriasis. Emerging evidence suggests a potential link between PsA and vitiligo, emphasizing the need for comprehensive management strategies for patients with overlapping autoimmune disorders. This case report evaluates the effectiveness of Upadacitinib in a patient with HLA-B27 positive PsA and coexisting vitiligo.

Methods: A 25-year-old male with a history of vitiligo presented in March 2024 with bilateral joint pain affecting the radiocarpal joints, metacarpophalangeal, and interphalangeal arthritis of the right third finger. He also exhibited a skin rash in the balano-preputial area. MRI findings indicated sacroiliitis, and HLA-B27 testing was positive. With pending Mycoplasma and Ureaplasma results, he was diagnosed with reactive arthritis and treated with maximal doses of NSAIDs (Naproxen, Celecoxib, Ibuprofen) and sulfasalazine (3 g/day). After three months without improvement, treatment was switched to methotrexate (20 mg/week). In August 2024, he returned with new lumbar spine pain and exacerbated arthritis. Laboratory tests revealed elevated acute phase reactants and negative infection markers. Ultrasound confirmed active inflammatory changes in the radiocarpal joint and dactilytis. MRI reiterated active sacroiliitis along with zygapophyseal joint involvement. . The persistent skin rash led to a biopsy, which revealed balanitis psoriasiformis, prompting a revised diagnosis of HLA-B27 positive psoriatic arthritis with axial and peripheral involvement.

Results: Given the lack of response to conventional therapy, targeted treatment with Upadacitinib (15 mg/day) was initiated, showing a favorable response within six weeks.

Conclusions: This case report confirms an efective outcome in managing PsA and Vitiligo with Upadacitinib.

Keywords: Upadacitinib, psoriatic arthritis, vitiligo

PV156 / #483

POSTER SESSION 14: PSORIATIC ARTHRITIS, GOUT & CHECKPOINT INHIBITORS
03-08-2025 1:55 PM - 2:55 PM

ULTRASONOGRAPHIC EVALUATION OF NAIL INVOLVEMENT IN PSORIATIC ARTHRITIS: A SYSTEMATIC REVIEW AND META-ANALYSIS

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Background and Aims: Psoriatic arthritis (PsA) often presents with nail involvement, which may precede joint symptoms. Ultrasonography offers a valuable tool for evaluating subclinical nail pathology, but findings are inconsistent. This systematic review and meta-analysis aimed to assess ultrasonographic nail findings in PsA compared to healthy controls.

Methods: A systematic review following PRISMA guidelines identified studies comparing ultrasonographic findings in PsA patients and healthy controls through PubMed, EMBASE, and Cochrane databases. Meta-analysis was conducted on parameters such as nail plate and bed thickness

Results: Five studies including 272 PsA patients and 161 healthy controls met the inclusion criteria (Table). Meta-analysis showed significantly increased nail plate thickness in PsA patients (pooled mean difference: 0.009 cm, 95% CI: 0.006–0.012 cm; I^2 = 58%) and nail bed thickness (mean difference: 0.096 mm, 95% CI: 0.024–0.169 mm; I^2 = 85%). Increased power Doppler signals indicated active inflammation in PsA patients.

Study Reference	Ultrasonographic Findings
Idolazzi et al. (2018)	Nail plate thickness increased in PsA vs. HC; nail bed thickness increased

Aydin et al. (2012)	Nail plate and matrix thickness increased in PsA
Mahmoud et al. (2024)	Nail bed thickness decreased in PsA compared to HC
Sandobal et al. (2014)	Ventral plate loosening; increased power Doppler signal in PsA
Naredo et al. (2018)	Nail bed thickness increased in PsA; correlation with NAPSI scores

Conclusions: Ultrasonography detects significant structural changes in PsA patients' nails, including increased thickness and vascularization. These findings support its use as a diagnostic and monitoring tool in PsA, particularly for early detection of subclinical disease.

Keywords: Arthritis, Psoriatic, Ultrasonography, Nails/diagnostic imaging

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POSTER SESSION 14: PSORIATIC ARTHRITIS, GOUT & CHECKPOINT INHIBITORS
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BEYOND THE DIAGNOSIS: A PSORIATIC ARTHRITIS CASE WITH AN UNEXPECTED TWIST

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Background and Aims: A patient with known psoriatic arthritis presented with localized pain and swelling in the left sternoclavicular joint, intermittent fever peaking at 38°C, particularly around noon, and a weight loss of 4 kg over the past month. The patient was on methotrexate 15 mg, which had previously provided stable disease control.

Methods: Laboratory tests showed elevated inflammatory markers, with a CRP of 7.34 mg/dL and an ESR of 39 mm/h. Mild macrocytic anemia was also noted (Hb 12.6 g/dL, MCV 97 fL). Initially, clinical suspicion pointed to Tietze syndrome, and an ultrasound of the affected area was performed. The ultrasound revealed a hypoechoic mass over the left sternoclavicular joint, infiltrating the surrounding muscle. Concerns of lymphoma or thymoma led to a CT scan with contrast, which demonstrated peripheral contrast enhancement and a probable direct connection between the mass and the joint, reducing the likelihood of malignancy.

Results: An MRI was subsequently performed, revealing findings suggestive of septic arthritis, with clear communication between the joint and the mass identified on the ultrasound. Methotrexate therapy was discontinued.

Conclusions: The patient was promptly admitted to the infectious diseases ward and started on antibiotic therapy with ceftriaxone and cotrimoxazole. This led to significant clinical improvement and a reduction in swelling. After the infectious episode fully resolved, the patient resumed his original therapy, maintaining good control of his arthritis.

Keywords: psoriatic arthritis, Septic arthritis

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POSTER SESSION 14: PSORIATIC ARTHRITIS, GOUT & CHECKPOINT INHIBITORS
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EFFICACY AND SAFETY OF IXEKIZUMAB IN A PATIENT WITH PSORIATIC ARTHRITIS - A CASE REPORT

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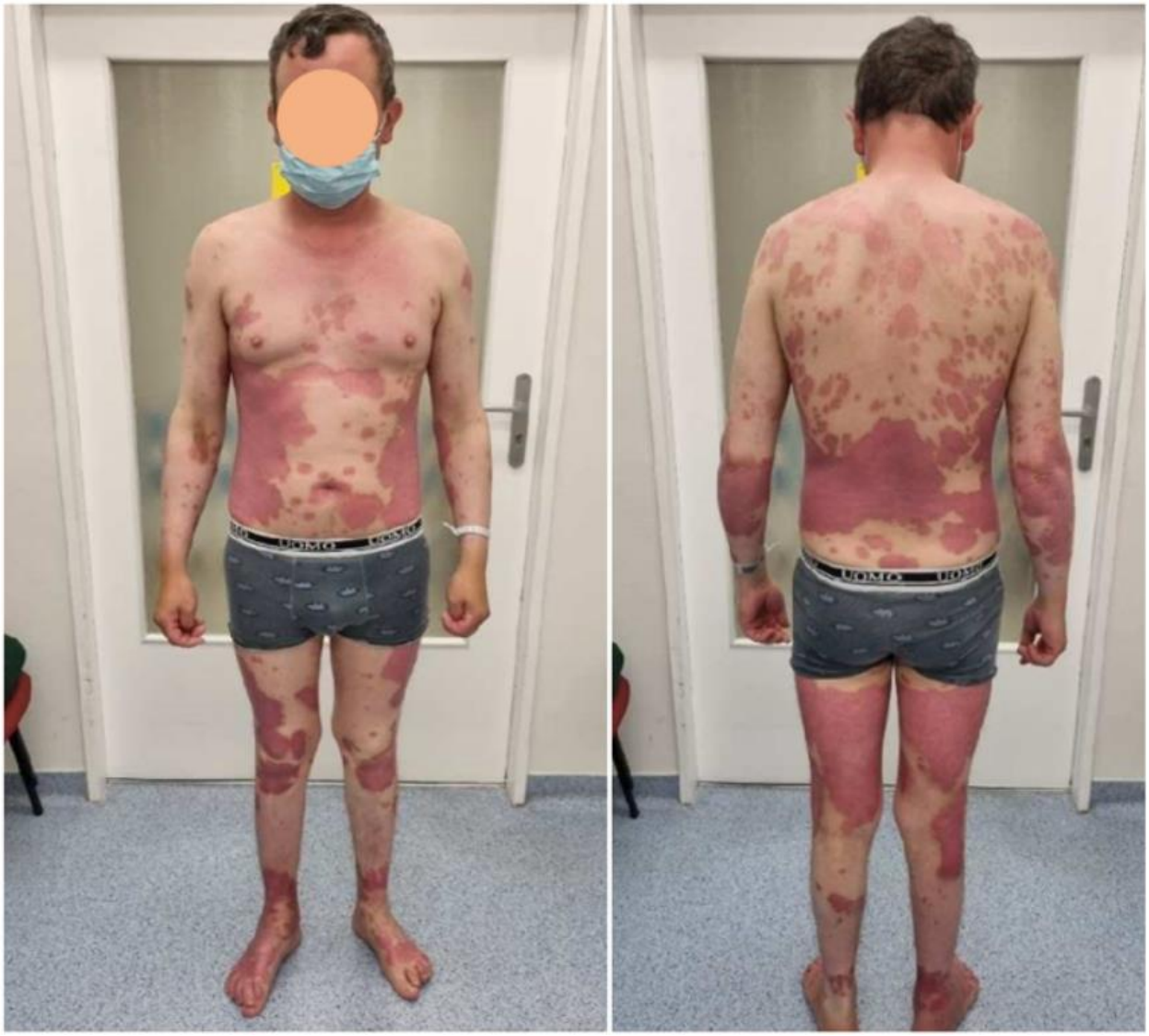
Background and Aims: A 36-year-old man with psoriasis since 2019 with mutilans-type psoriatic arthritis started biological treatment with ixekizumab on November 28, 2022 after obtaining the approval of the Therapeutic Committee of Biological Treatment. The Classification for Psoriatic Arthritis (CASPAR) Criteria were met: - Psoriasis 2 points, - Nail changes - 1 point,. - Sausage finger - 1 point, - Changes in radiological examination - 1 point, - Absent rheumatoid factor - 1 point. Previous treatment: non-steroidal anti-inflammatory drugs, cyclosporine 2.5 mg/kg b.w., methotrexate 15 mg once a week (discontinued due to ALT 103, ASPT 149), methylprednisolone 4 mg daily. A physical examination: psoriasis affected the scalp, face, upper and lower limbs and trunk (>10% of the body surface area) and nails . Foot arthritis mutilans, active dactylitis were observed. X-Ray: changes of the "pencil in a cup" type in metatarsophalangeal joints MTP II-V of the left foot.

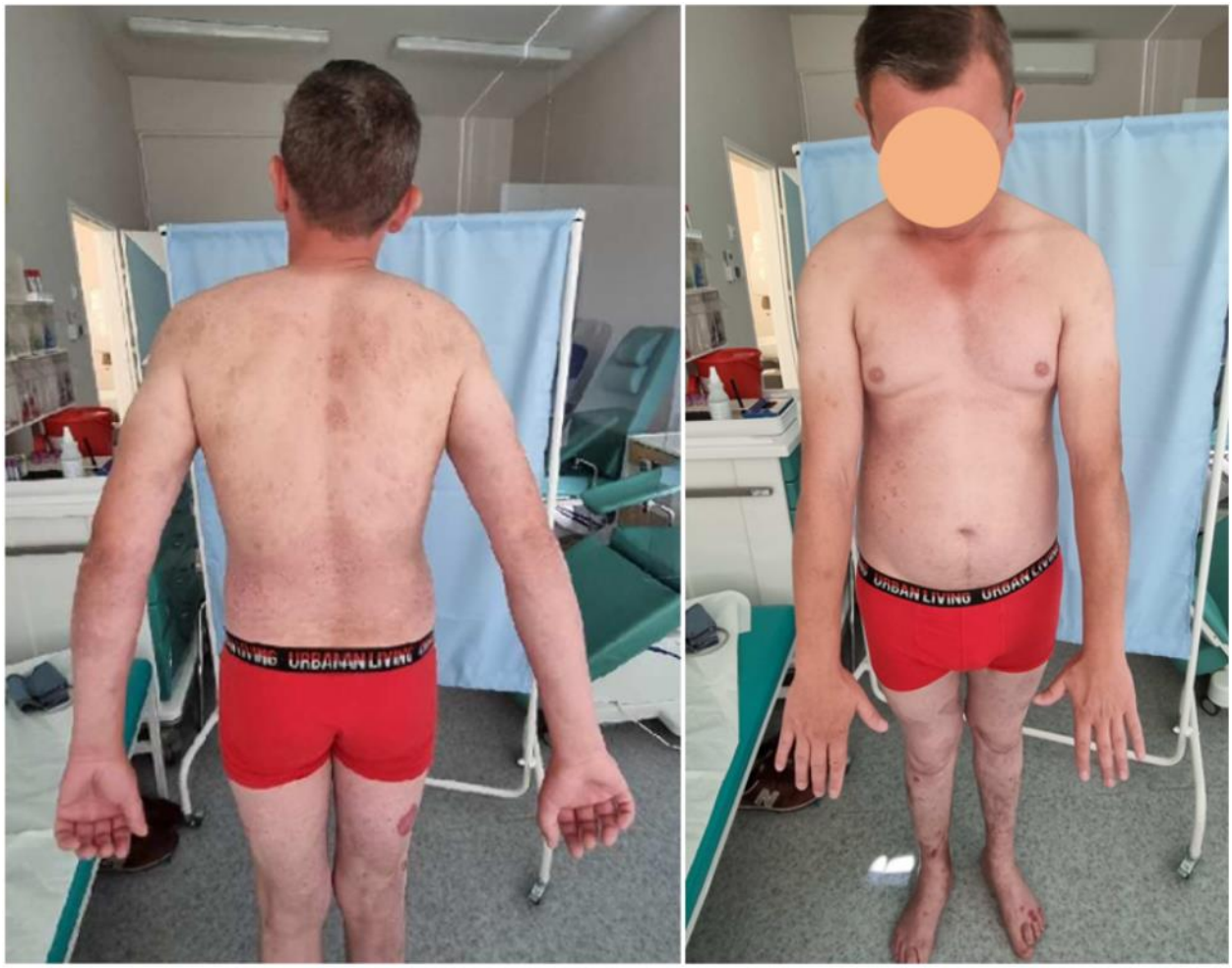
Methods: To evaluate and monitor PsA the PsARC (Psoriatic Arthritis Response Criteria) were used: In addition, X-ray and ultrasound examinations were performed.

Results:

Parameter	Visit_0	Visit_1	Visit_2	Visit_3	Visit_4	Visit_5
Doctor’s opinion (Likert_scale_1-5)	5	4	3	2	1	1
Person’s opinion (Likert_scale_1-5)	5	4	3	2	1	1
Number of tenderness joints	12	8	4	1	1	0

Number of swelling joints	12	8	4	1	1	0
ESR[mm/h]	22	18	19	15	7	8
CRP[mg/l]	10,91	6,35	6,85	2,58	1,66	1,69
ALAT[U/l]	103	72	43	29	19	33
ASPAT[U/l]	149	36	52	43	33	35

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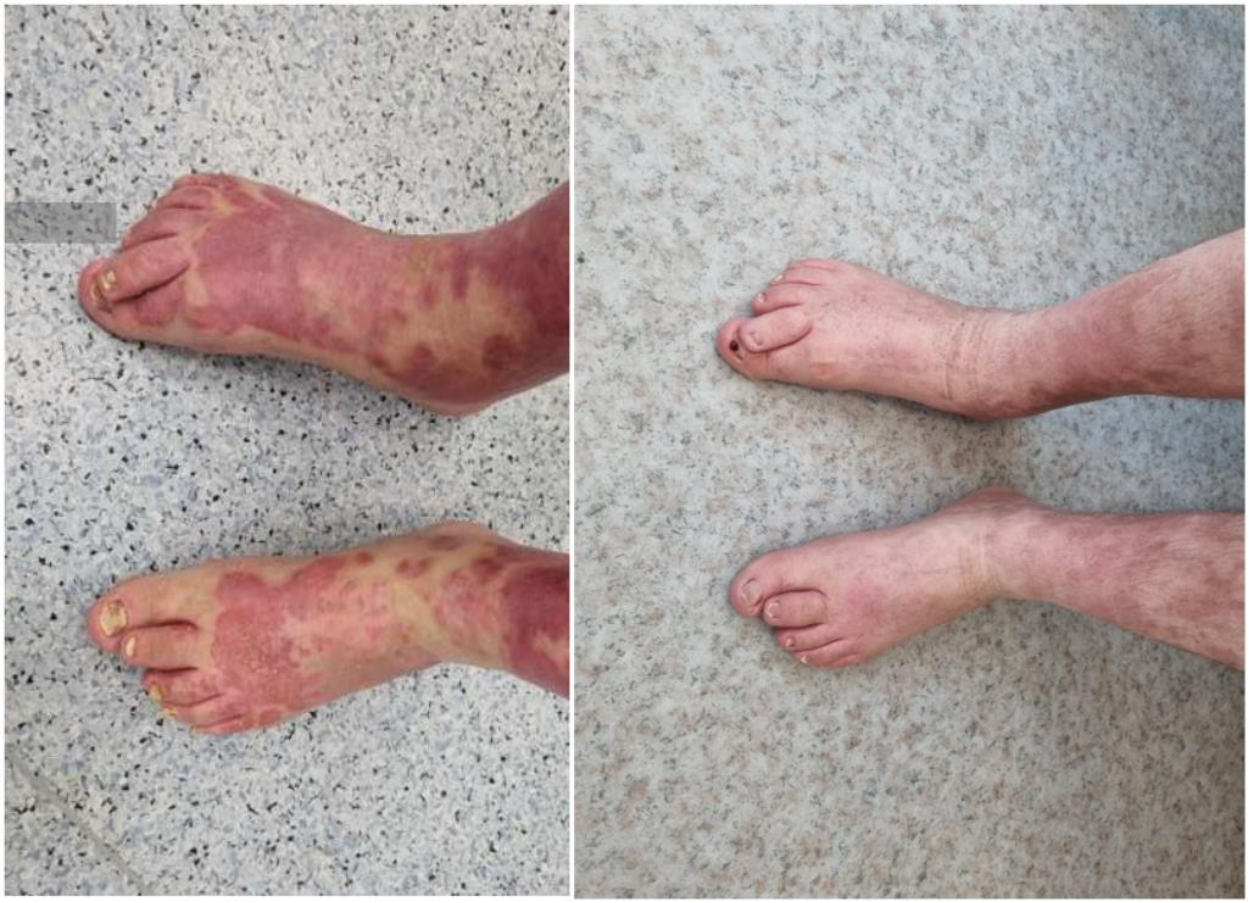

Conclusions: Two years of treatment with ixekizumab monotherapy is effective and safe.

Keywords: dactylitis, psoriatic arthritis, ixekizumab

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POSTER SESSION 15: SYSTEMIC LUPUS ERYTHEMATOSUS – CLINICAL ASPECTS
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NAVIGATING RHUPUS COMPLEXITY

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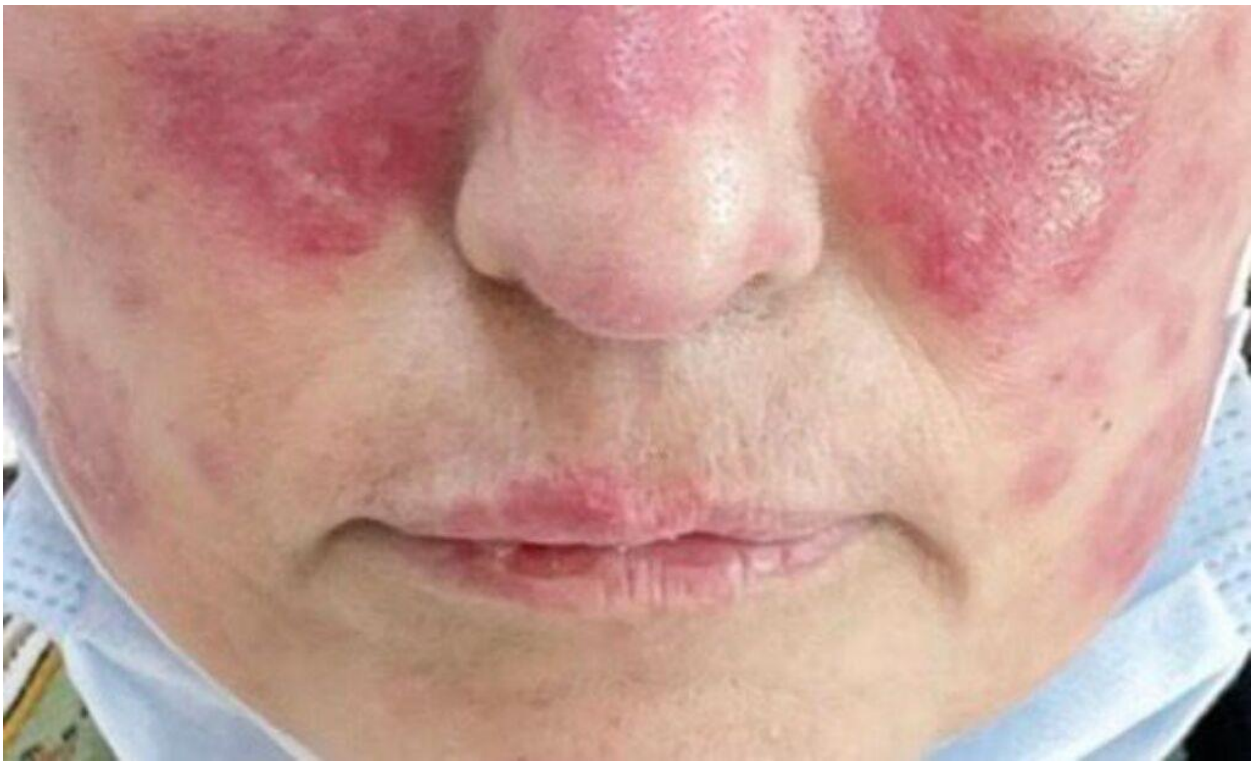
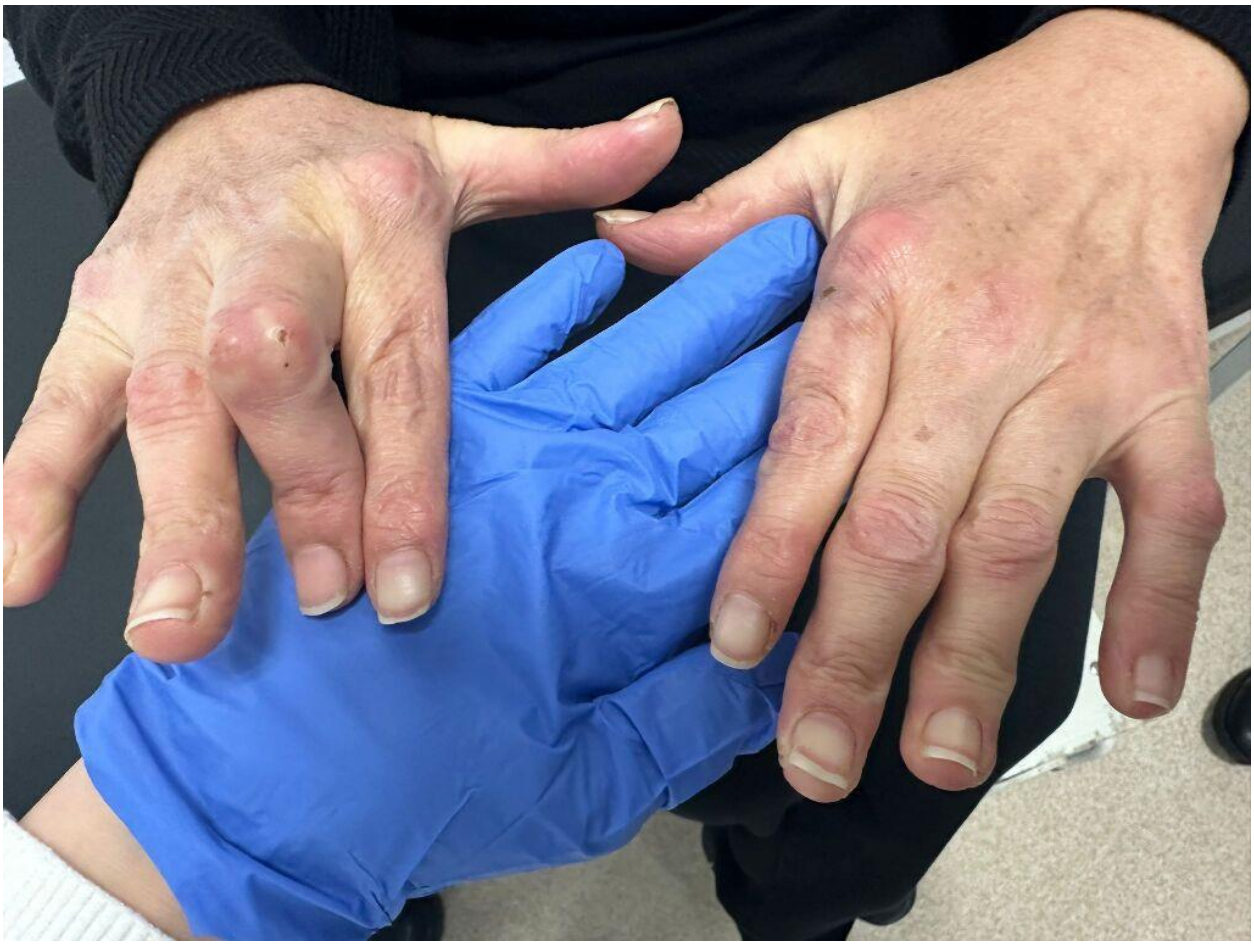
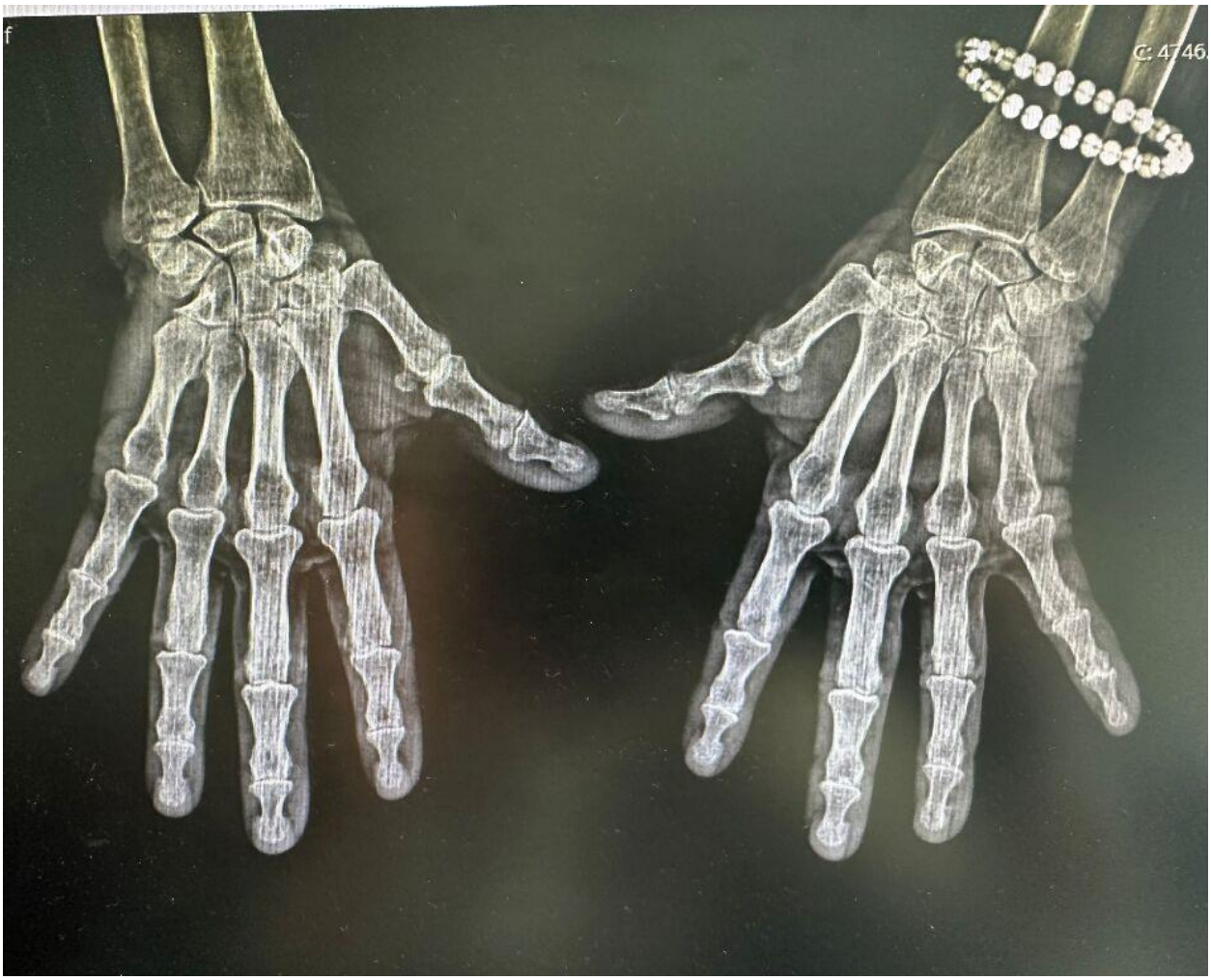
Background and Aims: Autoimmunity is one of the top ten causes of mortality and morbidity in young women. Approximately 20% of patients with a history of autoimmunity may develop additional autoimmune diseases, leading overlapping conditions. The term 'Rhupus,' which can be cited as an example of this situation, was introduced by Peter Schur in 1971 for patients who met the criteria for both RA and SLE. Rhupus is a rare syndrome, and approximately 60 cases have been described.

Methods:







A 42-year-old female patient has been under our rheumatology outpatient clinic's follow-up since 2019, diagnosed with seropositive RA. The diagnosis is supported by clinical evidence of inflammatory arthritis and positivity for rheumatoid factor (RF) and anti-CCP. Until 2022, there was no history of treatment other than methotrexate. In 2022, the patient applied with severe malar rash, oral ulcers and alopecia. ANA was positive at 1/160 dilution. Anti-Sm and anti-ds-DNA positivity was detected. We diagnosed her with SLE in this assessment.

Results: Since she also had seropositive RA, we were following up with the diagnosis of Rhupus syndrome. Hydroxychloroquine and methylprednisolone (24 mg) were added to the patient's treatment. The steroid dose was gradually reduced. Going forward, the plan is to further reduce the steroid dose to 5 mg/day, introduce mycophenolate mofetil to the treatment, and discontinue methotrexate.

Conclusions: This case outlines the presentation of a 42-year-old woman with erosive arthritis , diagnosed as Rhupus syndrome. To the best of our knowledge, this represents the tenth reported case of adult Rhupus syndrome in our country.

Keywords: Rheumatoid Arthritis, lupus, SLE

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POSTER SESSION 15: SYSTEMIC LUPUS ERYTHEMATOSUS – CLINICAL ASPECTS
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MULTIPLE AUTOIMMUNE SYNDROME (MAS) OR TOO MANY FACES OF LUPUS?

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Background and Aims: Multiple autoimmune syndrome (MAS) is a condition requiring a minimum of three distinct autoimmune diseases clustering together. Systemic lupus erythematosus (SLE), ulcerative colitis, autoimmune hepatitis and Hashimoto's disease are uncommonly associated.

Methods: Case presentation

Results: We report the case of a 55-year-old female patient with a history of Hashimoto's disease currently in a state of euthyroidism, who was admitted to our department for polyarthralgia affecting the small joints of her hands and knees, along with fatigue. This symptomatology started eight years ago and was accompanied by nonspecific skin rash on her lower limbs, diarrhea, flatulence and abdominal cramping. A colonoscopy revealed erythema and friability of the intestinal mucosa. A tissue sample analysis led to the diagnosis of ulcerative colitis. The patient was treated with 5-aminosalicylic and ursodeoxycholic acids, which resulted in complete clinical remission of her gastrointestinal (GI) symptoms, though hepatic cytolysis and cholestasis persisted. Further testing showed negative results for viral or autoimmune hepatitis. Liver biopsy could not be performed. At presentation to our clinic, SLE was confirmed based on clinical features and laboratory tests (mild thrombocytopenia, discordant inflammatory markers, hypergammaglobulinemia, hypocomplementemia, positive ANA, anti-dsDNA, anti-nucleosome and histone antibodies). FibroMax confirmed advanced hepatic fibrosis. Azathioprine was started leading to improvement in thrombocytopenia and transaminase levels.

Conclusions: Differentiating between lupus hepatitis and autoimmune hepatitis can be challenging, since they may have similar clinical, laboratory and systemic features. Also, intestinal involvement in SLE can mimic other disorders of the GI tract. It is crucial to be aware that patients may M, necessitating ongoing surveillance.

Keywords: multiple autoimmune syndrome, lupus hepatitis, overlap syndromes

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POSTER SESSION 15: SYSTEMIC LUPUS ERYTHEMATOSUS – CLINICAL ASPECTS
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CALCINOSIS CUTIS – SIGN OF OVERLAP OR PREDICTING SEVERE LUPUS?

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Background and Aims: Aim is to present diagosticaly and therapeutically challenging, rare phenotype of systemic lupus erythematosus (SLE) with progressive generalized form of dystrophic skin calcinosis (calcinosis cutis, CC) and lupus nephritis (LN).

Methods: Patient is middle aged female with 13 years history of SLE. For almost ten years her disease was significant for mild to moderate skin, joints, muscles and bone marrow involvement. On admission in June 2022. she had high febrility, extensive livedo reticularis with palpable, generalized "orange-peel" skin indurations complicated by abscess in right infragluteal region. Skin changes were biopsy confirmed as CC. In February 2023. she had first episode of LN.

Results:

From 2011. to November 2021.	June 2022.
Mild to moderate skin, joints, bone marrow and muscle involvement	Constitutional symptoms, CC, gluteal abscess
ESR, CRP normal	ESR, CRP high
24h proteinuria: normal	24h proteinuria: 0.47g
Moderate lymphopenia, mild thrombocytopenia	Moderate lymphopenia, mild thrombocytopeni
ANA speckled > 1/640 Anti-ds-DNA up to 363 IU/ml Anti-SSA 142 IU/ml c3, c4 normal	ANA homogenous/speckled 1/160 Anti-ds-DNA normal
Normal findings: Antiphospholipid antibodies	Normal findings: Hitotriosidase Quantiferon Pa Jo1, myositis profile, antiphospholipid antibod
Capilaroscopy: nonspecific.	Capilaroscopy: scleroderma-like pattern.

Conclusions: CC is rare cutaneous manifestation of SLE and could be associated with renal disease.

Keyword: calcinosis cutis, overlap syndrome, lupus nephritis

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POSTER SESSION 15: SYSTEMIC LUPUS ERYTHEMATOSUS – CLINICAL ASPECTS
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THE GENETIC SUSCEPTIBILITY, ENVIRONMENTAL TRIGGERS AND DISEASE SEVERITY AT ONSET IN A RUSSIAN COHORT OF PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS

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Background and Aims: Systemic lupus erythematosus(SLE) is a chronic heterogenic autoimmune disease affected by several genetic and environmental factors. The aim of this study was to describe any provoking factors and disease severity in a Caucasian SLE cohort at the Russian rheumatology center

Methods: This observational retrospective-prospective study included 140patients (88% women, median aged 34[26;41]years) with SLE(SLICC 2012). Disease at diagnosis was categorized as Mild, Moderate or Severe, based on SLEDAI-2K: ≤4 – Mild, 5–10 - Moderate, ≥11 - High activity(Severe disease)

Results: SLE was diagnosed at median age 26[19;34] years, and the age of the first SLE manifestations was 23[17;31]years, the period duration between the first symptoms and diagnosis was 12[5;48]months. The median of SLEduration was 3[0,3;12]years, SLEDAI-2K–8[4;11]score, SDI–0[0;1]score.Positive family history of immune-inflammatory rheumatic diseases(IIRD) among the first-line relatives was found in 11% of SLEpts. In the most patients(56%), there were no connection with any provoking factors and SLEonset, in 15% of patients-with ultraviolet radiation/insolation, in 14%-with infection, in 10%-with pregnancy, in a few patients(1-2%)- with combined oral contraceptives use, stress, vaccination and trauma.At the time of SLE diagnosis the most patients had Moderate (SLEDAI-2K=5-10) and Severe/high (SLEDAI-2K≥11) activity: 41% and 35%,respectively. Mild disease (SLEDAI-2K=0-4) was identified in 24% of SLEpatients

Conclusions: In the Russian cohort SLE was diagnosed about a year after the first manifestations. In the majority of cases, there was no specific trigger factor, the most

frequently identified were: positive family history of IIRD, ultraviolet radiation/insolation, infections and pregnancy. In 76% of SLE patients the disease onset was characterized by moderate and high severity.

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POSTER SESSION 15: SYSTEMIC LUPUS ERYTHEMATOSUS – CLINICAL ASPECTS
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CLINICAL MANIFESTATIONS OF SYSTEMIC LUPUS ERYTHEMATOSUS IN A RUSSIAN COHORT: FROM THE FIRST SYMPTOM TO FOLLOW-UP

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Background and Aims: The aim of this study was to analyse the phenotype of systemic lupus erythematosus(SLE) starting from the first manifestation and during follow-up since the diagnosis of SLE in a Caucasian cohort at the Russian rheumatology center.

Methods: This observational retrospective-prospective study included 140 patients (88% women, median aged 34[26;41] years (median [interquartile range 25;75%]), with SLE (SLICC 2012) attending a routine visit at our Clinic between February 2021 and June 2024.

Results: SLE was diagnosed at median age 26[19;34] years, and the age of the first SLE manifestations was 23[17;31]years, the period duration between the first symptoms and SLE diagnosis was 12[5;48]months. The median disease duration at last follow-up was 3[0,3;12]years, SLEDAI-2K–8[4;11]score, Systemic Lupus International Collaborating Clinics damage index (SDI) – 0[0;1]score. The first symptoms of SLE were: inflammatory arthritis-40%, cutaneous lupus-34%, haematological disorders-6%, nephritis-5%, serositis-1%, nervous system involvement-1%, mucosal ulcers-1%. Among 'non-criteria' symptoms the most common were: unexplained fever-6%, interstitial lung disease-3%, lymphadenopathy-2%, Raynaud's phenomenon-1%. During the period of follow-up of patients with diagnosed SLE, the frequency of clinical manifestations changed cumulatively: inflammatory arthritis-92%, haematological disorders-81%, cutaneous lupus-77%, serositis-46%, alopecia-45%, nephritis-44%, mucosal ulcers-35%,nervous system involvement-14%. Among 'non-criteria' symptoms the most common were: unexplained fever-67%, lymphadenopathy-42%, Raynaud's phenomenon-26%, interstitial lung disease-13%, ulcerative necrotising vasculitis-11%, myocarditis-2%.

Conclusions: The first SLE symptoms can be either criterion or “non-criterion”, the most common being arthritis/arthralgia, skin lesions and cytopenias. Nephritis, serositis and unmotivated fever were more common in the follow-up SLE period in our cohort, in contrast to other Caucasians.

Keyword: Systemic lupus erythematosus, first symptoms, clinical manifestations

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POSTER SESSION 15: SYSTEMIC LUPUS ERYTHEMATOSUS – CLINICAL ASPECTS
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A CLINICAL CASE OF SYSTEMIC LUPUS ERYTHEMATOSUS WITH ALPS-LIKE SYNDROME: CHALLENGES IN DISEASE MANAGEMENT, RECURRENT FLARES, AND SUBCUTANEOUS IMMUNOGLOBULIN THERAPY

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Background and Aims: Systemic lupus erythematosus (SLE) is a chronic autoimmune disease with potential multiorgan involvement. This clinical case is about a 24-year-old patient diagnosed in childhood with multiorgan SLE complicated by ALPS-like syndrome and hypogammaglobulinemia. Her clinical course involved renal (lupus nephritis), hematological, pulmonary, and cutaneous manifestations, alongside recurrent infections, including herpes zoster and bacterial sepsis.

Methods: Patient underwent various immunosuppressive therapies, including high-dose corticosteroids, methotrexate, hydroxychloroquine, tacrolimus, thalidomide, eculizumab, bortezomib, belimumab, mycophenolate mofetil and Rituximab (only two doses of 1 gr). Due to persistent disease activity, treatment required frequent adjustments. Intravenous immunoglobulin replacement therapy was initiated for hypogammaglobulinemia, accompanied by prophylactic antiviral and antibacterial measures to mitigate infection risks.

Results: Despite these interventions, disease control remained inadequate, with recurrent flares, particularly affecting the kidneys and lungs. Immunosuppressive regimens were adjusted due to complications like anemia, thrombocytopenia, and persistent infections. Renal involvement fluctuated, with episodes of increased proteinuria, necessitating a progressive increase in mycophenolate mofetil dosage. After initiating intravenous immunoglobulin therapy, the patient's IgG levels stabilized at 780 mg/dL. This allowed for a transition to weekly subcutaneous immunoglobulin therapy at a dose of 40 ml/8g, which the patient is currently receiving.

Conclusions: This case highlights the complexity of treating SLE with ALPS-like syndrome and immune dysfunction. Recurrent disease flares, infection complications, and therapy-related side effects require constant adjustments. Ongoing monitoring, personalized treatment strategies and the use of weekly subcutaneous immunoglobulin therapy are essential for better disease control and to reduce the risk of infections in these complex cases.

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POSTER SESSION 15: SYSTEMIC LUPUS ERYTHEMATOSUS – CLINICAL ASPECTS
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DISCOID LUPUS ERYTHEMATOSUS IN THE CONCHAL BOWL

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Background and Aims: Discoid lupus erythematosus (DLE) is a chronic cutaneous condition characterized by annular plaques, scarring, and follicular plugging, often affecting sun-exposed areas. Auricular involvement, particularly in the pinna and conchal bowls, is common and may occur more frequently in Black patients. This case describes the presentation and treatment of two patients with auricular DLE, emphasizing early recognition and management.

Methods: We reviewed two cases of auricular DLE seen in our Dermatology Clinic. Patient demographics, clinical features, serology, and treatment approaches were documented. Both patients underwent serologic testing and dermoscopic evaluation, but biopsies were avoided due to the difficulty of sampling from the conchal bowl.

Results: A 44-year-old Fitzpatrick II female and a 37-year-old Fitzpatrick V male presented with annular plaques, scarring, and follicular plugging on the face, pinna, and conchal bowls. Both patients were strongly positive for ANA and double-stranded DNA, confirming DLE. Hydroxychloroquine was initiated due to minimal response to topical steroids. The male patient, also diagnosed with SLE, showed lupus nephritis and autoimmune hemolytic anemia, prompting consideration of mycophenolate mofetil and belimumab.

Conclusions: Auricular involvement in DLE is an important diagnostic clue, particularly in Black patients. Early systemic treatment can prevent disfigurement, and emerging biologics offer promising options for refractory cases. Awareness of auricular DLE across skin types is crucial for timely intervention and treatment.

Keyword: Discoid lupus

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POSTER SESSION 15: SYSTEMIC LUPUS ERYTHEMATOSUS – CLINICAL ASPECTS
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DO WE NEED PROMOTE PHYSICAL ACTIVITY IN PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS?

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Background and Aims: Physical exercises can play a crucial role in the treatment of rheumatic diseases, optimizing both physical and mental health, and improving the quality of life. We assessed physical activity and its impact on patients with systemic lupus erythematosus (SLE).

Methods: This cross-sectional clinical study included 99 SLE outpatients. The following questionnaires were utilized: Health Assessment Questionnaire (HAQ-DI), Beck Depression Inventory II (BDI-II), Systemic Lupus Erythematosus Disease index (SLEDAI) and the Short-form Health Survey (SF-36). Physical activity was assessed using the long form of the International Physical Activity Questionnaire (IPAQ – LF) and the Metabolic Equivalent of Task (MET) minutes per week (min/wk). Otherwise, patients underwent a 2-minute walking test to objectively measure physical performance and validate subjective patient assessments.

Results: Only 57.6 % SLE Patients were physically active. Physical inactivity was associated with higher fatigue ($p < 0.04$) and lower «vitality» (SF-36, $p < 0.03$) scores. Otherwise, in SLE patients moderate and severe pain was correlated with physical inactivity ($p = 0.0056$). Although physical inactivity correlated with a higher total disease activity score (SLEDAI ≥ 6 $p = 0.0056$), neither of the single SLEDAI items and organ manifestations including musculoskeletal manifestations was associated with physical inactivity. Furthermore, it was showed a significant association between the IPAQ category low and depressive mood ($p = 0.012$) and physical functioning (SF-36 $p = 0.0079$).

Conclusions: It is important to consider the existing lack of physical activity in SLE patients. In addition, programs for the patient to optimize physical activities in everyday life should be included.

Keywords: phisical activity, systemic lupus erytematodes, fatigue

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MACROPHAGE ACTIVATION SYNDROME - CLINICAL PICTURE OF THE FIRST PRESENTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS, A CASE REPORT

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Background and Aims: Macrophage activation syndrome (MAS) is a rare, very serious complication of rheumatic diseases, with potentially life-threatening consequences. It rarely occurs as the first presentation of SLE.

Methods: We will present the case of a young patient aged 36 who was admitted to the Institute of Rheumatology with complaints in the form of pain in all bones and muscles, polymorphic changes on the skin, malaise, weakness, aphthous changes on the mucous membranes of the mouth and nose, subfebrile temperature in the evening, sore throat, difficult swallowing and findings of leukocytopenia with lymphocytopenia.

Results: Considering the finding of progressive hematological manifestations of the disease in the direction of successive involvement of all three blood lines, as well as the worsening of the patient's general condition, a bone marrow biopsy was performed. The findings confirmed toxically altered bone marrow with signs of macrophage activation syndrome (secondary hemophagocytic lymphohistiocytosis). In support of this diagnosis, in addition to the PH findings of hemophagocytosis, there is a significantly increased ferritin level, elevated triglycerides, a pathological hepatogram and certainly progressive pancytopenia. In addition, the patient had criteria for the diagnosis of systemic lupus erythematosus.

Conclusions: Treatment was started with pulse doses of methylprednisolone, administration of intravenous immunoglobulins (IVIG), granulocyte and macrophage colony stimulating factor (GM-CSF), and then treatment was continued with high doses of Dexasone and Cyclosporin A. The applied therapy resulted in a gradual recovery of the bone marrow, as well as general condition of the patient.

Keywords: SLE, Macrophage activation syndrome, first clinical presentation

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THE INTERTWINED CONNECTIONS BETWEEN SYSTEMIC LUPUS ERYTHEMATOSUS AND INFECTIONS

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Background and Aims: We hereby present the case of a 38-year-old female patient referred to our clinic from the infectious diseases department, where she presented with fever, a vasculitic rash on her lower limbs, and an ulcerous lesion covered by a black crust on her inner right ankle after a tick bite. Given this clinical context, she was tested positive for IgM antibodies against Rickettsia conorii, and after the boutonneuse fever diagnosis, she underwent treatment with Doxycycline for 14 days with the complete resolution of her symptoms.

Methods: One month later she presented in our clinic with complaints and clinical findings consistent with arthralgia and arthritis involving wrists, metacarpophalangeal and proximal interphalangeal joints, shoulders, knees, ankles, myalgia, malar rash developed after UV light exposure, and aphtous ulcers on her buccal mucosa and anterolateral tongue. She also reported fatigue and lower limb bilateral oedema.

Results: Further findings confirmed the presence of increased inflammatory markers, leukopenia, C3, C4 hypocomplementemia, increased muscle enzymes, anti-dsDNA, anti SS-A, anti SS-B, anti-SM antibodies significantly increased, 24h urine protein of 2,7g, hypoalbuminemia, dysmorphic hematuria, absent antiphospholipid antibodies. A thorax-abdominal-pelvic CT scan was negative for neoplasia or serositis and a cardiac ultrasound was also negative for pericarditis or valvular abnormalities. A skin biopsy confirmed the diagnosis of subacute cutaneous lupus erythematosus.

Conclusions: We diagnosed the patient with systemic lupus erythematosus and started hydroxychloroquine 400mg/day and 3g i.v. methylprednisolone with positive response, however, one week after, she was diagnosed with mitral abscess with MRSA for which she is currently being treated in the ICU department.

Keywords: systemic lupus erythematosus, rickettsia conorii, MRSA

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CEPHALGIA IN SYSTEMIC LUPUS ERYTHEMATOSUS

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Background and Aims: Neurological manifestations are a common occurrence in patients with systemic lupus erythematosus. Among these, headache is considered to be the most prevalent, However, the question of whether headache is a manifestation of the underlying disease or a comorbid disease remains unresolved. Background: to investigate the frequency and type of headache in patients with systemic lupus erythematosus

Methods: 75 patients diagnosed with SLE were included in the study. The mean age was 37.0 [32.0;46.0] years, disease duration was 3.0 [1.0;9.0] years, and all patients were interviewed about the presence or absence of headache. The questionnaire included questions about headache including family history, time of onset, frequency, localisation, nature, provoking factors and symptoms accompanying headache (nausea, vomiting, photophobia, phonophobia). After completing the questionnaire, all patients were divided into groups according to the presence of tension-type headache (TTH) or migraine.

Results: A total of 54 patients (72%) out of 75 reported headache that debuted before the first symptoms of SLE. In 24 patients (44.5%), the headache was bilaterally localised, pressing/compressive/pulsating, of moderate intensity, did not increase with normal physical activity, and was not accompanied by nausea and/or vomiting. In 30 patients (55.5%), the headache occurred at a young age, there was a family history, and the headache fulfilled the ID-Migraine criteria.

Conclusions: Cephalgia is a common symptom in patients with SLE, which can result in a reduction in quality of life. Headache that commenced prior to the diagnosis of SLE represents an independent disease entity that requires treatment according to its specific type (migraine or TTH).

Keyword: cephalgia, headache, systemic lupus erythematosus, tension-type headache, migraine

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LUPUS LOW DISEASE ACTIVITY AND REMISSION IN PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS

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Background and Aims: One of the treatment targets for patients with systemic lupus erythematosus (SLE) is to prevent organ damage. Reducing and maintaining lupus low disease activity or remission accomplishes this goal. The aim is to evaluate the achievement of remission and low activity of SLE in patients in a Russian cohort.

Methods: The study included 146 patients (121 women/25 men), the average age was 33.0 [26.0-41.0] y.o., the duration of the disease was 57.0 [18.0; 130.0] months. The SLEDAI-2K, SLE-DAS indices were used to assess activity and LLDAS, DORIS to assess outcomes. Glucocorticoids were given to 144 (99%) patients. Patients were followed up for 36 months with a clinic visit every 6 months.

Results: The activity and damage indices in SLE, glucocorticoid dose (GC), and the number of patients with low disease activity are presented in Table 1. During 36 months of follow-up, there is a significant decrease in the SLE activity indices and a slight increase in the damage index. Despite a significant decrease in the GC dose, it still remained quite high.

Table 1. Characteristics of patients with SLE during the follow-up period

	SLEDAI-2K	SLE-DAS	GCs dose (prednisolone equivalent)	Hydroxychloroquine, n (%)	Immunosuppressants, n (%)	Biologic, n (%)	LLDAS, n (%)	Disease control, n (%)	Clinical remission, n (%)	Complete remission, n (%)	DI SLICC/ACR	Patients with low activity but GC dose is >7,5mg / day
Baseline	8.0 [4.0-12.0]	9.38 [4.41-19.26]	15.0 [10.0-25.0]	140 (96)	77 (53)	67 (46)					1.0 [0.0-2.0]	
6 months, n=106	4.0 [2.0-8.0]	2.08 [1.12-7.64]	10.0 [7.5-15.0]	106 (100)	56 (51)	59 (56)	12 (11)	0 (0)	10 (9)	5 (5)	1.0 [0.0-2.0]	33 (31)
12 months, n=97	2.0 [1.0-6.0]	2.08 [1.12-6.47]	10.0 [7.5-10.0]	91 (94)	46 (47)	40 (41)	21 (22)	5 (5)	8 (8)	4 (4)	1.0 [0.0-2.0]	29 (30)
18 months, n=62	2.0 [0.0-6.0]	1.33 [0.37-4.48]	7.5 [5.0-10.0]	58 (94)	28 (45)	31 (50)	12 (19)	1 (2)	7 (11)	7 (11)	1.0 [0.0-3.0]	15 (24)
24 months, n=64	2.0 [0.0-5.0]	1.22 [0.37-5.00]	7.5 [5.0-10.0]	61 (95)	28 (44)	20 (31)	15 (23)	10 (16)	3 (5)	8 (13)	1.0 [0.5-3.0]	11 (17)
30 months, n=31	2.0 [2.0-6.0]	2.08 [1.12-4.78]	7.5 [5.0-10.0]	27 (87)	17 (55)	14 (45)	3 (10)	5 (16)	4 (13)	3 (10)	1.0 [1.0-3.0]	7 (23)
36 months, n=43	2.0 [1.0-6.0]	2.08 [1.12-3.95]	7.5 [5.0-10.0]	40 (93)	20 (47)	14 (33)	6 (14)	4 (9)	7 (16)	4 (9)	1.0 [1.0-3.0]	9 (21)
Wilcoxon signed-rank test	p=0.0003	p=0.0003	p<0.0001								p=0.001	

Conclusions: No one in the patient group was satisfied with remission due to the persistence of high doses of GC. In the Russian cohort, an average of 24% of patients

receive high maintenance doses of GC despite achieving low disease activity. This may be explained by the high adherence of healthcare providers to GC therapy due to fear of disease flares and lower adherence to cytostatic therapy.

Keywords: systemic lupus erythematosus, remission, low disease activity

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REFRACTORY ANTI-MDA5 + DERMATOMYOSITIS SUCCESSFULLY TREATED WITH ANIFROLUMAB: A CASE REPORT

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Background and Aims: Anti-MDA5-positive dermatomyositis (DM) is associated with several clinical features, including interstitial lung disease (ILD), articular and cutaneous manifestation. Type I interferon (IFN) pathways have shown to be highly upregulated in DM, therefore an anti-IFN approach is reasonable also in this disease.

Methods: A 55-year-old woman was diagnosed in 2020 with anti-MDA5-positive DM, having low grade fever, weight loss, erythematous-desquamative lesions on hands, cuticle dystrophy and severe skin ulcerations (figure 1). She was treated during the follow-up with cyclosporine (gastrointestinal intolerance), steroid pulses, hydroxychloroquine and mycophenolate, without improvement. Intravenous immunoglobulins and prostanoids led to ulcer improvement but were stopped due to gastrointestinal intolerance. Rituximab was stopped at the first infusion for the same reason. From 2022 the patient developed ILD, shown by HRCT and spirometry with DLCO. Significant cutaneous improvement with healing of ulcers was achieved with tofacitinib; the drug was stopped due to hypotension and gastrointestinal intolerance. Given the subsequent loss of appetite and weight, development of polyarthritis (DAS28-CRP 4,79), skin ulcers, alopecia, Gottron's signs (CDASI 5), worsening of ILD and the refractoriness of the disease, anifrolumab was started in July 2024 (300 mg IV every four
weeks).

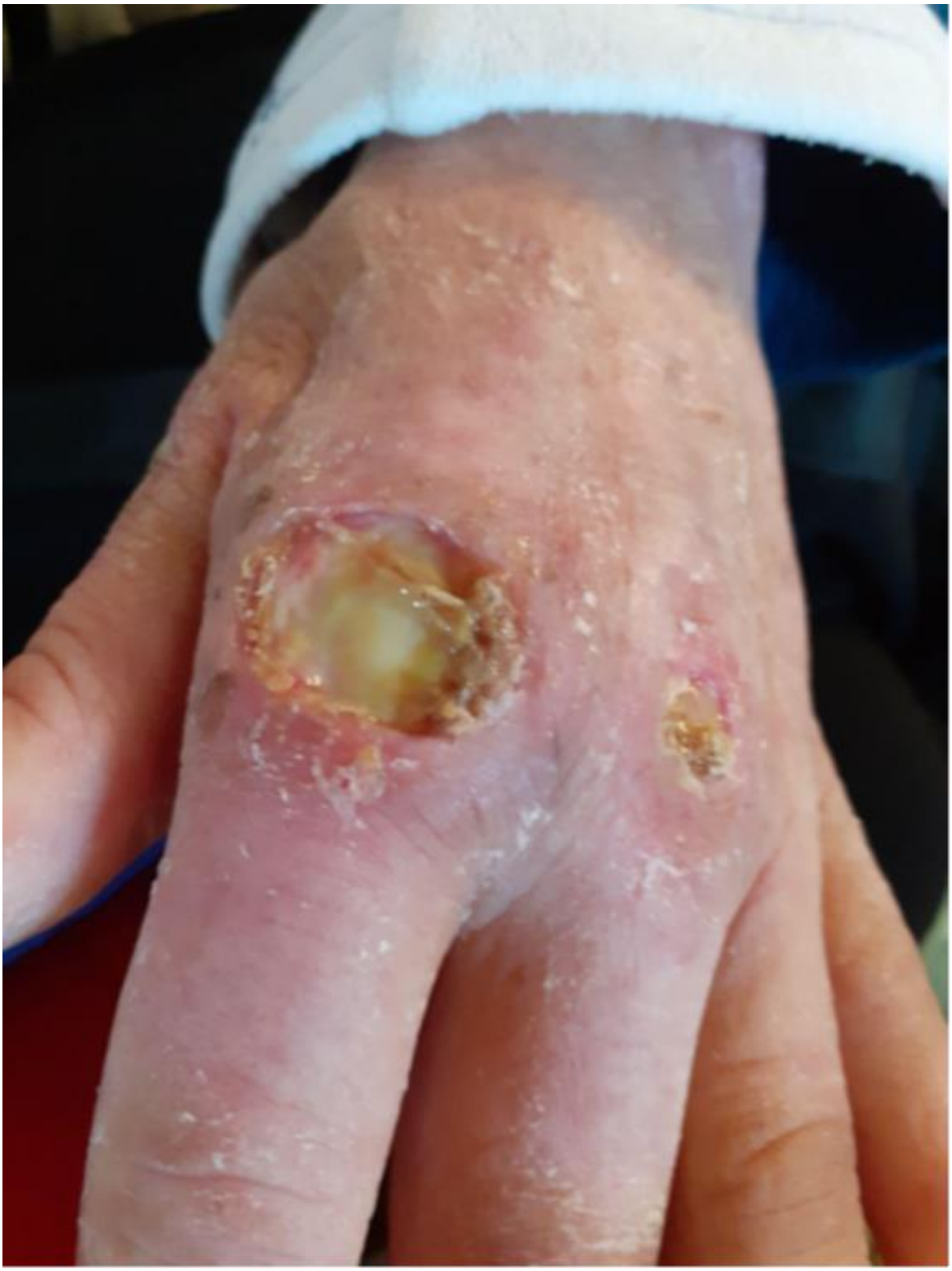

Figure 1: severe skin ulcer

Results: After three anifrolumab infusions, the patient reported improved appetite with 3 kg weight gain, improvement of arthritis (DAS28-CRP 3,36) and persistent absence of

cutaneous ulcers (figure 2) (CDASI 1), without respiratory symptoms. No adverse effects were
observed.

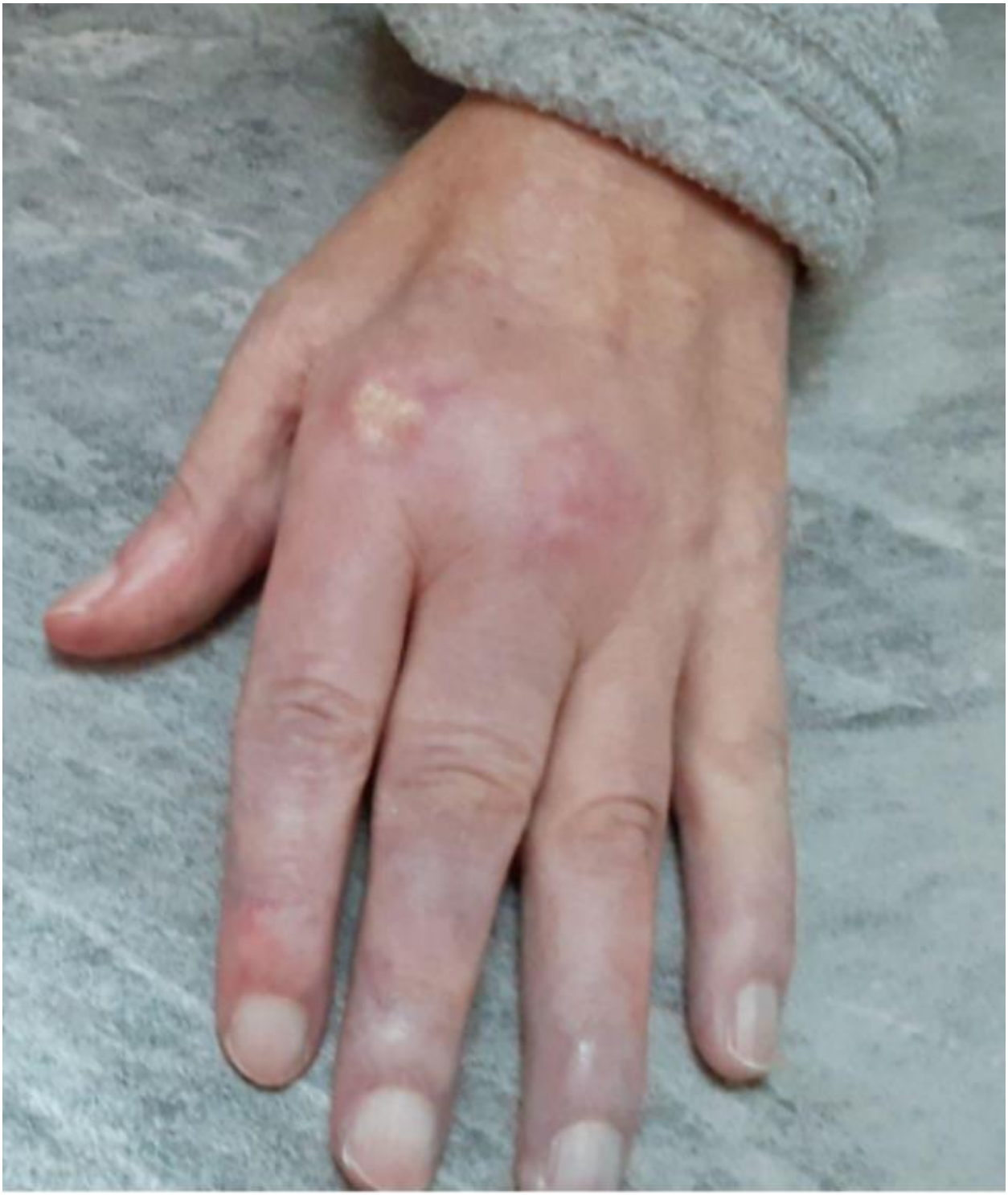

Figure 2: healing of skin ulcers

Conclusions: This case suggests the potential efficacy of anifrolumab in refractory anti-MDA5-positive DM.

Keywords: interferon, ulcers, anti-MDA5

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TWO DIFFERENT CASES OF ANTI-HMGCR POSITIVE IMMUNE-MEDIATED NECROTIZING MYOPATHY

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Background and Aims: Immune-mediated necrotizing myopathy (IMNM) can occur as a result of statin exposure, evidenced by anti-HMGCR autoantibodies. It is a rare side effect with an incidence of 2 or 3 of every 100,000 patients taking statins, frequently delayed by months or years after statin commencement. Patients are usually presented with subacute onset with progressive symmetric proximal muscle weakness and elevated creatinine kinase level that persists even when the statin is discontinued, with dysphagia occurring in approximately one-third of patients. It does not resolve after the discontinuation of statins.

Methods: We present two anti-HMGCR positive immune-mediated necrotizing myopathy cases with different clinical presentations.

Results: The first patient was exposed to atorvastatin for five months, after which gradual muscle weakness developed with elevated creatine kinase levels in laboratory findings, myopathic pattern verified by EMNG, and histology confirmed inflammatory changes. She was treated with glucocorticoids with good clinical response. The other patient was on atorvastatin therapy for two years, and three months before hospital admission, rapidly progressive distal and proximal muscle weakness with dysphagia developed. The disease was very resistant to treatment with glucocorticoids and IVIG and was finally treated with cyclophosphamide with a good response.

Conclusions: The report aims to point out the heterogeneity of the clinical presentation of IMNM, and the importance of taking it into account in the differential diagnosis of myopathies in patients on statin therapy.

Keywords: Immune-mediated necrotizing myopathy, atorvastatin, anti-HMGCR

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THE IMPACT OF THE OCTOBER 7TH ATTACK AND WAR ON THE PHYSICAL AND MENTAL HEALTH OF FIBROMYALGIA PATIENTS: A CROSS-SECTIONAL ANALYSIS

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Background and Aims: Fibromyalgia symptoms can be exacerbated by acute or chronic stress. While military conflicts have been associated with increased rates of PTSD and fibromyalgia in military personnel, civilian populations exposed to such conflicts are also at higher risk of chronic pain and somatic symptoms. This study aims to assess the physical and mental health of fibromyalgia patients in Israel following the October 7th attacks and to identify key risk and protective factors.

Methods: An online survey conducted in January 2024 gathered data from fibromyalgia patients across Israel, a population exposed to intense conflict-related stressors, including missile attacks, mass evacuations, and widespread military mobilization. The survey addressed demographics, health behaviors, access to medical services, anxiety, depression, coping mechanisms, social support, and fibromyalgia-specific indices like the Widespread Pain Index (WPI) and Symptom Severity Scale (SSS).

Results: Among 246 respondents, nearly 70% ceased complementary treatments and psychotherapy, and 83.3% reduced or stopped physical exercise. These patients reported heightened pain, anxiety, depression, and perceived deterioration. Strong correlations were found between higher anxiety/depression levels and increased pain and somatic symptoms ($p<0.01$). Social support was associated with reduced anxiety and depression but not pain. Positive life orientation was linked to lower pain, somatic symptoms, and mental distress ($p<0.05$). Avoidant coping strategies worsened outcomes, while problem-focused coping and acceptance had protective effects.

Conclusions: The war has negatively affected the mental and physical health of fibromyalgia patients. Cessation of physical activity and therapy may contribute, while positive coping and life orientation can offer protection.

Keywords: fibromyalgia, chronic pain, stress

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DIAGNOSTIC AND CLINICAL SIGNIFICANCE OF ANTI-TIF1 GAMMA ANTIBODY IN PATIENTS WITH DERMATOMYOSITIS

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Background and Aims: Dermatomyositis (DM) is a subtype of idiopathic inflammatory myopathy with primary involvement of the skin and muscles, with possible involvement of other organs, such as lungs and esophagus. Antitranscription intermediary factor 1γ (anti-TIF1γ) is one of the myositis-specific antibodies in DM, and it is associated with the occurrence of malignant disease.

Methods: The retrospective study included 8 patients with dermatomyositis in whom anti-TIF1γ antibody was detected in the period from February 2022 to June 2024.

Results: The median age of patients was 66.6 years, while the ratio of women to men was 7:1. All patients presented with dominantly skin symptoms (heliotrope rash, V neck sign, Gottron's papules, Holster's sign, scalp involvement). Among the other clinical manifestations, muscle weakness was present in all patients, dysphagia in two patients, while interstitial lung disease wasn't noted in any patient. First treatment choice were glucocorticoids (100% of patients), and other drugs which are used were methotrexate (37.5%), azathioprine (25%), intravenous immunoglobulins (25%), cyclosporine (12.5%) and hydroxychloroquine (12.5%); multiple therapy at the same time showed better effect. In some patients skin changes were very refractory to the therapy. Considering the association of anti-TIF1γ DM with paraneoplastic syndrome, all patients were extensively evaluated for potential malignant disease which was diagnosed in four of them (2 lung cancer, 1 ovarian cancer, 1 breast cancer).

Conclusions: The anti-TIF1γ antibody showed as a specific marker of skin manifestations of the disease, highly resistant to therapy, while the previously known importance of the diagnostic evaluation for malignant disease in these patients was confirmed.

Keywords: anti-TIF1γ antibody, dermatomyositis, malignant disease

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ADMINISTRATION OF PCSK9 INHIBITORS IN PATIENTS WITH IMNM: DATA FROM A CASE SERIES

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Background and Aims: Immune-mediated necrotizing myopathies (IMNMs) are a subset of inflammatory myopathies associated with the presence of anti-SRP or anti-HMGCR antibodies. HMG-CoA reductase (HMGCR) is the pharmacological target of statins, reinforcing the hypothesis that IMNMs may be induced by statin administration. The administration of PCSK9 inhibitors could be an alternative lipid-lowering therapy in these patients. Our study aimed to record the response of patients with IMNM who were administered a PCSK9 inhibitor as lipid-lowering therapy.

Methods: Parameters such as muscle strength, CPK levels, LDL levels, and immunosuppressive therapy were evaluated before and after the administration of a PCSK9 inhibitor.

Results: Five female patients with IMNM were identified, who were administered a PCSK9 inhibitor. All patients except one had a history of statin exposure. The patient who had not been exposed to statins was positive for the anti-SRP antibody, while the others were positive for the anti-HMGCR antibody. The average follow-up duration was 18.2 months (range 9-26 months). The average CPK level before PCSK9 inhibitor treatment was 10,286 ± 749.43 IU/L, decreased by 893.2 ± 688.73 IU/L at the most recent measurement. All patients received steroid therapy (with gradual reduction in dosage). Four out of five received second-line immunosuppressive therapy. No patient showed a clinical relapse of the disease after starting the PCSK9 inhibitor, or needed an escalation of its treatment.

Conclusions: The administration of PCSK9 inhibitors might be an effective and safe option for the treatment of dyslipidemia in patients with immune-mediated necrotizing myopathy (IMNM).

Keywords: IMNM, HMGCR, PCSK9 INHIBITORS

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DIAGNOSIS AND CLINICAL ASSOCIATIONS OF ANTI-PMSCL ANTIBODY IN A RETROSPECTIVE COHORT

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Background and Aims: Anti-PmScl antibody has been described in SAD such as IIM, SSc and overlap syndromes. To analyse the final diagnosis and characteristics among patients with positive anti-PmScl antibodies at the General Hospital of Granollers.

Methods: Retrospective observational study including patients with positive PmScl antibodies from January 2020 to July 2024 at the General Hospital of Granollers with reference population of 305,000 inhabitants.

Results: 28 patients were identified, 67% women and 33% men. 25% were PmScl-75 positive, 63% PmScl-100 and 12% were positive for both.

	Weak Positive n=16	**Moderate/Strong Positive** n=12
IIM/SSc	4/16 (25%) - DMM 3 - UCTD 1	8/12 (58%) - DMM 3 - SAS 2 - SSc 3
Other Autoimmune or Inflammatory Conditions	SAD 4/16 (25%) - RA 1 - SLE 2 - SS 1	SAD 0

	ILD 3/16 (18%) - IPAF 2 - UIP 1	ILD 2/12 (16%) - IPAF 1 - UIP 1
No Disease	5/16 (31%)	2 (16%)

2/3 of patients had positive ANA (50% nucleolar pattern) ILD is the most frequent manifestation (37%). Other clinical manifestations: muscle weakness (18.5%), mechanic's hands (14.8%), arthritis (11%), 3 patients cancer

Conclusions: - 42% of patients were diagnosed with IIM or SSc, 30% with a SAD or ILD, and 25% had no association with any SAD. - It is more likely that a diagnosis of IIM or SSc will be made in patients with moderate or high PmScl titers. Weak titers are less likely to be associated with IIM or SSc. - A small subgroup of patients (2) are clinically diagnosed with SAS

Keywords: anti-PMscl, dermatomiositis, antisynthethase syndrome

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BEYOND MUSCLE WEAKNESS: A CASE OF ANTI-KU ANTIBODIES AND NEUROLOGICAL INVOLVEMENT

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Background and Aims: Ku proteins are involved in the repair of DNA and the regulation of phosphorylation in various nuclear proteins, such as enzymes and transcription factors. The occurrence of anti-Ku antibodies is relatively low and they were described in ovelap syndromes (SLE/myositis, scleroderma/myositis) but does not correlate with any particular autoimmune disorder.

Methods: We present the case of a patient admitted in our clinic in 2023, highlighting the investigation process.

Results: A 60-year-old female with atrial fibrillation and type II diabetes presented for rheumatological evaluation, reporting arthralgias in small joints and no significant biological changes. One year later, she returned with dysesthesias in her lips and tongue, and hypoesthesia in the lower left side of her face. Paraclinical tests showed a creatine kinase (CK) level seven times above normal, with strongly positive anti-Ku antibodies and positive anti-PM/Scl p75 antibodies, while there was no clinical evidence of muscular involvement and the muscle biopsy was normal. In the following months, the facial dysesthesias progressively extended to her eyebrows, the angle of the mandible, and the preauricular area. The patient continued to show no muscular symptoms despite persistently elevated CK levels, and electromyography revealed myopathic changes, predominantly distal in the lower limbs and proximal in the upper limbs.

Conclusions: Considering that the patient shows no other causes for the elevation of CK, a suspicion of myositis is raised, although there is currently no evident muscular involvement. The literature suggests possible neurological involvement of the trigeminal or facial nerves, in the context of positive anti-Ku antibodies.

Keywords: Myositis, anti-Ku antibodies, neurological involvement

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ANTI-RNA SYNTHETASE AUTOANTIBODIES (ARS) ARE EQUALLY PRESENT IN ANTI-RO52 POSITIVE AND ANTI-RO52 NEGATIVE PATIENTS WITH INFLAMMATORY MYOSITIS

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Background and Aims: Previous studies demonstrated a striking co-occurrence of anti-Ro52 autoantibodies and anti- RNA synthetase autoantibodies (ARS) in patients with idiopathic inflammatory myositis (IIM). This has led investigators to suggest that these autoantibodies are driven by the same antigenic stimulus. The aim was to assess such co-occurrence in a single-center study of IIM patients from single-center.

Methods: Forty-four patients with IIM were tested for ARS and anti-Ro52 antibodies by line immunoblotting (EUROIMMUN) testing amongst others anti-ARS antibodies against anti-Jo1, anti-PL7, anti-PL12, anti-EJ, anti-OJ, anti-Ha, anti-Ks and anti-Zo, respectively.

Results: Overall, anti-Ro52 antibodies were present in 18 (41%) IIM patients including 6 anti-Jo1 positive and anti-ARS antibodies were detectable in 21 (48%). Amongst the 18 anti-Ro52 positive, 9 (50%) were anti-ARS positive and 9 (50%) were anti-ARS negative. Amongst, 26 anti-Ro52 negative, 12 (46%) were anti-ARS positive (but none of those was anti-Jo1 positive) and 14 (54%) were negative.

Conclusions: Our results do not support the simultaneously co-occurrence of anti-Ro52 and anti-ARS antibodies in IIM, other than that of anti-Jo1, raising concerns regarding the pathophysiological origin of the two autoantibody specificities.

Keywords: autoantibody, anti-Ro52, ant-synthetase syndrome

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SEASONALITY OF INFLAMMATORY MYOPATHIES: ANALYSIS OF CLINICAL CASES IN A TROPICAL COUNTRY

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Background and Aims: Inflammatory myopathies are rare autoimmune diseases characterized by muscle involvement, but they also affect other organs. Subgroups of myopathies have been defined based on specific autoantibodies and clinical phenotypes, with several studies showing a tendency toward a temporal pattern. Information from countries near and south of the equator is scarce.

Methods: Four cases of myopathies of different etiology were included according to classification criteria, evaluating association between an environmental determinant and seasonality.

Results: We present four cases of inflammatory myopathy treated in the rheumatology department of a high-complexity institution between February and April. An 83-year-old man and a 47-year-old woman with anti-HMG CoA antibodies and necrotizing myopathy; a 46-year-old female patient with antisynthetase syndrome, anti-Mi-2B, and anti-Ro 52 antibodies; and a 45-year-old woman with an overlap of myopathy, systemic sclerosis, systemic lupus erythematosus, and anti-PL-12 antibodies, the latter with a fatal outcome. The narrow time frame in which these cases occurred is noteworthy, a rare phenomenon in our local setting given the low incidence of these diseases and the presence of higher precipitation.

Conclusions: In Colombia and neighboring countries, there is no available information regarding the impact of geographic latitude and seasonality, which may reflect a common environmental factor in inflammatory myopathies. These data suggest the influence of local environmental factors, which are unique to our geographical location, in patients with a genetic predisposition and their relationship with disease progression, prognosis, and response to treatment. **Acknowledgements.** Colombian Association of Rheumatology.

Keyword: antisynthetase syndrome, immune-mediated necrotizing myopathy, seasonality, inflammatory myopathies

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ANTI-TRANSCRIPTIONAL INTERMEDIARY FACTOR 1Γ (TIF-1Γ) ANTIBODY POSITIVE CASES OF INFLAMMATORY MYOSITIS(IIM): A SINGLE CENTRE EXPERIENCE.

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Background and Aims: Anti-TIF1γ is a myositis-specific autoantibody associated with cancer-related myositis. This study aims to assess its prevalence among idiopathic inflammatory myopathy (IIM) patients and characterize their clinical features.

Methods: A retrospective review of IIM cases in our department was conducted, focusing on anti-TIF1γ+ cases. Testing included a line immunoassay (Euroimmun) for 20 antigen reactivities and ANA detection via indirect immunofluorescence.

Results: Among 121 IIM cases, 16 (13.2%) were anti-TIF1γ+. Of these, 50% had malignancies identified within five years of IIM diagnosis. The mean age of the anti-TIF1γ+ group was 65.56 years(SD=12.12), with malignancy cases averaging 68.23(SD=8)years. Cancers included breast, lung, ovarian, oropharyngeal, and renal cell. The mean high-risk factor score for the anti-TIF1γ+ group was 3.87(SD=0.8), with malignancy cases scoring 4.12(SD=0.56). Skin involvement occurred in 93.7%, with a mean CDASI activity of 29.43(SD=17.9), damage=1.37(SD=3.56) and muscle involvement was universal (MMT8=111.37/150 SD=22.25), CPK=1586 SD=1,413). Dysphagia was noted in 50% of cases, equally distributed across malignancy subgroups, and one patient had ILD. All patients were ANA+, and 50% had other myositis-related autoantibodies.

Conclusions: Anti-TIF1γ is linked to high malignancy risk, elevated CDASI score, moderate MMT8 score, and a high dysphagia rate in our patient group.

Keywords: Myositis, AUTOANTIBODIES, MALIGNANCY

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ONSET OF IDIOPATHIC POLYMYOSITIS REFRACTORY TO TREATMENT OF IN YOUNG FEMALE: A CASE REPORT

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Background and Aims: Idiopathic inflammatory myopathies (IIM) are a group rare autoimmune diseases characterized by progressive proximal muscle weakness. The definition of polymyositis is controversial, in some cases IIM have unique clinical and laboratory signs. The aim of our work is to describe the clinical manifestation IIM in our patient as well as the therapeutic aproaches.

Methods: We report the case of a 35-year-old female with rapid generalized symmetrical weakness in the upper and lower extremities and fewer, which led to her hospitalization in our Rheumatology Center for deep diagnostic.

Results: Laboratory tests didn't show any significant changes - normal level of CK- 101 U/L, slowly increase ALT - 66 U/L, normal ESR and CRP. Autoantibodies: ANA. Jo-1, IgG<0.2 U/ml; PM-Scl, IgG<0.2 U/ml; Mi-2, IgG<0.2 U/ml; SS-A/Ro, IgG<0.2 U/ml. No evidence of infections. CT scan without significant findings. Patient underwent neurological examination with electromyography, that showed increased activity, positive sharp waves, and fibrillation, which corresponded with a primary myogenic pattern. We decided to use IV methylprednisolone 500 mg #3 with next oral administration 48 mg/daily and cyclophosphamide 500 mg IV every 2 weeks #6) without achieving clinical improvement. We provide treatment escalation with rituximab and after two doses of rituximab 500 mg get a major improvement patient condition, increased muscle strength and quality of life.

Conclusions: We suggest that our patient suffers from an unknown subset of IIM, that had an atypical clinical manifestations and resistant disease that needs further monitoring of the patient's condition will allow making a conclusion about the longterm effectiveness of immunosuppressive therapy.

Keywords: Idiopathic inflammatory myopathies, POLYMYOSITIS, resistant disease