

## 001

### SYSTEMIC INFLAMMATION, LYMPHOPROLIFERATION AND VASCULOPATHY IN A PATIENT WITH ARHGAP10 MUTATION

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## 002

### BIOLOGICS IN THE TREATMENT OF PEDIATRIC BEHÇET'S DISEASE: RESULTS OF AN INTERNATIONAL COLLABORATIVE STUDY BY THE PRES VASCULITIS WORKING PARTY

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## 004

### COFILIN-1 IS A REDOX SENSOR REGULATING THE NLRP3 INFLAMMASOME

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## 005

### SGT1 CONTROLS NLRC4 INFLAMMASOME ACTIVATION IN AUTO-INFLAMMATORY DISEASES

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## 006

### GSDMD AND GSDME AMPLIFY NLRP3 ACTIVATION IN AUTOINFLAMMATION

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#### 007

#### **PROTEIN ARRAY PROFILING IDENTIFIES DISTINCT AUTO ANTIBODY SIGNATURES IN SYSTEMIC AUTO INFLAMMATORY DISEASES**

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#### 008

#### **VALIDATION OF HUMAN PHENOTYPE ONTOLOGY (HPO) TERMS AND DEVELOPMENT OF AN AI-BASED DIAGNOSTIC TOOL FOR SAIDS USING THE EUROFEVER REGISTRY: THE ODINO PROJECT**

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#### 009

#### **ASSESSING THE IMPACT OF FAMILIAL MEDITERRANEAN FEVER (FMF) ON PHYSICAL ACTIVITY IN CHILDREN USING THE PHYSICAL ACTIVITY QUESTIONNAIRE FOR CHILDREN (PAQ-C): A COMPARATIVE PRELIMINARY STUDY WITH HEALTHY CONTROLS**

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#### 010

#### **INCORPORATION OF RECENT SELECTION SIGNALS IMPROVES VARIANT IMPACT PREDICTION IN IMMUNE-MEDIATED GENES**

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#### 011

#### **IMPROVED MACHINE LEARNING MODELS FOR PREDICTING COLCHICINE RESISTANCE IN FAMILIAL MEDITERRANEAN FEVER**

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## 012

### DIAGNOSIS OF CRYOPYRIN-ASSOCIATED PERIODIC SYNDROME (CAPS) IN ADULTHOOD: LESSONS FROM A FRENCH COHORT

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## 013

### NEUTROPHIL EXTRAVASATION AND BBB DISRUPTION IN MURINE NOMID: INSIGHTS INTO NEUROINFLAMMATION

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## 014

### IRAK2 DEFICIENCY CAUSES A NEW IMMUNE DYSREGULATION DISORDER

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## 015

### CHANGING THE LANDSCAPE OF ACQUIRED SAIDS - REPORT FROM THE UK REFERENCE GENETIC LABORATORY

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## 016

### RARE TNFAIP3 HYPMORPHIC VARIANTS ARE A MASSIVELY UNDERESTIMATED DRIVER OF HUMAN AUTOINFLAMMATORY DISEASE GLOBALLY

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## 017

## JAK INHIBITORS ARE EFFECTIVE IN PEDIATRIC REFRACTORY NLRC4 GAIN OF FUNCTION MUTATION

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## O18

### DOMINANT NEGATIVE ADA2 MUTATIONS CAUSE ADA2 DEFICIENCY IN HETEROZYGOUS CARRIERS

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## O19

### PULMONARY ARTERIAL HYPERTENSION WITH STILL'S DISEASE: A NEW PULMONARY MANIFESTATION ASSOCIATED WITH HLA-DRB1\*15

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## O20

### FUNCTIONAL ANALYSIS OF C- AND N-TERMINAL CDC42 VARIANTS REVEALS DISTINCT PATHWAYS OF AUTOINFLAMMATION RESPONSIBLE FOR DIFFERENT CDC42-ASSOCIATED AUTOINFLAMMATORY DISEASES

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**O21**

**CURRENT TREATMENT OF MACROPHAGE ACTIVATION SYNDROME WORLDWIDE: THE METAPHOR PROJECT, A PRES/PRINTO REAL-LIFE INTERNATIONAL SURVEY**

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**O22**

**MANAGING PATIENTS WITH CRYOPYRIN ASSOCIATED PERIODIC SYNDROME (CAPS). HOW DOES INITIATING TREATMENT WITH IL-1 MEDICATIONS AFFECT PATIENT'S SYSTEMIC INFLAMMATION, SYMPTOM REPORTING AND QUALITY OF LIFE. EXPERIENCE OF A SPECIALISED UK CENTRE.**

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**O23**

**PROGRESS REPORT ON IL6 INHIBITION IN ROSAH AUTOINFLAMMATORY DISEASE**

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**O24**

**PROSPECTIVE FOLLOW UP OF 37 PREGNANCIES IN WOMEN RECEIVING IL1 INHIBITORS FOR SYSTEMIC AUTOINFLAMMATORY DISEASES: AN MULTICENTRIC FRENCH STUDY FROM THE GR2 COHORT.**

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## 025

### THE LONG JOURNEY OF CONGENITAL SYPHILIS DIAGNOSIS: THROUGH MALIGNANCY AND AUTOINFLAMMATORY DISEASE SUSPICION

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## 026

### CLINICAL PRESENTATION AND COURSE OF PULMONARY INVOLVEMENT IN CHRONIC NONBACTERIAL OSTEOMYELITIS

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## 027

### RETROPERITONEAL FIBROSIS (RPF) IN A CASE OF H SYNDROME – A DIAGNOSTIC CHALLENGE AND LITERATURE REVIEW

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## 028

### UNMASKING DIAGNOSTIC CHALLENGES: H SYNDROME MISTAKEN FOR CAPS

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## 029

### BEYOND NOONAN SYNDROME: QUESTIONING THE ROLE OF PTPN11 MUTATION IN PEDIATRIC AUTOINFLAMMATORY DISEASE: A CASE REPORT

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## 030

### OGFRL1 GENE MUTATIONS MAY LINK CHERUBISM TO CHRONIC RECURRENT MULTIFOCAL OSTEOMYELITIS (CRMO)

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## 031

### EFFICACY AND TOLERABILITY OF BISPHOSPHONATES IN THE MANAGEMENT OF CHRONIC RECURRENT MULTIFOCAL OSTEOMYELITIS (CRMO) IN CHILDREN: A 30-PATIENT RETROSPECTIVE COHORT STUDY

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## 032

### TRANSCRIPTOMIC INSIGHTS INTO PFAPA SYNDROME: RNA-SEQUENCING ANALYSIS OF FLARE RELATIVE TO NON-FLARE STATES

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## 033

### STRUCTURE AND FUNCTION OF PYRIN INFLAMMASOME: MECHANISTIC LINK BETWEEN FMF AND NOCARH SYNDROME

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### 034

#### **MEVALONATE KINASE DEFICIENCY – AN AUTOINFLAMMATORY DISEASE OF DYSREGULATED NK CELLS**

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### 035

#### **INFLAMMATORY AND DYSIMMUNE MANIFESTATIONS IN T/NK-CELL TYPE CHRONIC ACTIVE EBV INFECTION: A DESCRIPTION OF 14 CASES OF A RARE AND HETEROGENEOUS CLONAL LYMPHOID HEMATOLOGICAL DISORDER**

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### 036

#### **EMERGING TREATMENT STRATEGIES FOR VEXAS SYNDROME: A SYSTEMATIC REVIEW AND META-ANALYSIS**

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### 037

#### **ROLE OF IL-18 AS A BIOMARKER IN MONITORING PEDIATRIC PATIENTS WITH STILL'S DISEASE**

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**038****EVALUATING SIGLEC-1 EXPRESSION ON MONOCYTES AS A DIAGNOSTIC BIOMARKER FOR TYPE I IFN-RELATED PEDIATRIC AUTOINFLAMMATORY DISEASES**

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**039****DEEP PHENOTYPING IDENTIFIES INFLAMMATORY PATHWAYS ASSOCIATED WITH DISEASE ACTIVITY OF VEXAS SYNDROME**

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**040****IMPROVEMENT OF REFRACTORY STILL'S/SYSTEMIC JUVENILE IDIOPATHIC ARTHRITIS LUNG DISEASE IN 6/7 CHILDREN TREATED WITH A NOVEL, BI-SPECIFIC IL-1BETA/IL-18 NEUTRALIZING ANTIBODY**

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**041****PHOSPHOMEVALONATE KINASE DEFICIENCY: UNCOVERING NEW DIMENSIONS OF THE DISEASE PHENOTYPE**

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**042****CHARACTERISTICS AND PROBLEMS OF JAPANESE PATIENTS WITH UBA1 VARIANT-NEGATIVE VEXAS SYNDROME-LIKE AUTOINFLAMMATORY DISEASE**

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**043****TARGETING THE DYSREGULATED TYPE I IFN RESPONSE IN ADENOSINE DEAMINASE 2 DEFICIENCY EFFECTIVELY MITIGATES INFLAMMATION VIA PATHWAY INHIBITION AND GENE THERAPY**

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#### 044

#### FIRST GLOBAL SERIES OF VEXAS SYNDROME IN WOMEN: A COMPARATIVE ANALYSIS OF 14 FEMALE AND 274 MALE CASES

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#### PT01

#### VEXAS SYNDROME IN FRANCE: A MULTICENTER CASE-SERIES OF 318 CASES FROM THE FRENCH VEXAS STUDY GROUP (FRENVEX).

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#### PT02

#### PROGRESSIVE GLOMERULONEPHRITIS IN PEDIATRIC SAVI PROVIDES INSIGHTS INTO PATHOGENESIS AND THE ROLE OF TYPE I IFN IN RENAL OUTCOMES

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#### PT03

#### PHOSPHOMEVALONATE KINASE DEFICIENCY EXPANDS THE GENETIC SPECTRUM OF SYSTEMIC AUTOINFLAMMATORY DISEASES

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#### PT04

#### PREGNANCY OUTCOMES IN WOMEN WITH FAMILIAL MEDITERRANEAN FEVER TREATED WITH ANAKINRA: A RETROSPECTIVE STUDY

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#### PT05

#### IMMUNOLOGICAL INSIGHTS INTO H SYNDROME: A FRENCH NATIONAL COHORT STUDY OF 33 PATIENTS HIGHLIGHTING AUTO-INFLAMMATORY MANIFESTATIONS

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#### PT06

#### EFFECTS OF CANAKINUMAB DOSE ADJUSTMENTS ON DISEASE CONTROL OF AUTOINFLAMMATORY PERIODIC FEVER SYNDROMES – INTERIM RESULTS OF THE RELIANCE NON-INTERVENTIONAL STUDY

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## PT07

### **IMPACT OF AUTOINFLAMMATORY DISEASES: INSIGHTS FROM AN INTERIM ANALYSIS OF THE PRO-AID STUDY**

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## PT08

### **PREDICTION OF THE COLCHICINE RESPONSE ACCORDING TO FAMILIAL MEDITERRANEAN FEVER (FMF) 50 SCORE IN PEDIATRIC PATIENTS: ACUTE PHASE REACTANTS OR EXISTING SCORING SYSTEMS?**

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## PT09

### **TREATMENT OUTCOMES IN VEXAS SYNDROME: A RETROSPECTIVE STUDY FROM THE UK VEXAS INTEREST GROUP (VEXNET-UK)**

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#### **PT10**

#### **INFECTION BURDEN IN PATIENTS WITH GENETIC INTERFERONOPATHIES: A MONOCENTRIC RETROSPECTIVE COHORT STUDY**

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#### **PT11**

#### **DISCONTINUATION OF COLCHICINE TREATMENT IN PATIENTS WITH FAMILIAL MEDITERRANEAN FEVER**

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#### **PT12**

#### **HUMAN ADA2 DEFICIENCY IS CHARACTERIZED BY THE ABSENCE OF AN INTRACELLULAR HYPOGLYCOSYLATED FORM OF ADENOSINE DEAMINASE 2**

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#### **PT13**

#### **PYRIN INFLAMMASOME ACTIVATION LEADS TO IL-18 SECRETION AND PERPETUATES IFN-GAMMA SECRETION IN A NOVEL CULTURE-BASED MEVALONATE KINASE DEFICIENCY MODEL**

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**PT14****AUTOINFLAMMATORY PATIENTS WITH GOLGI-TRAPPED CDC42 EXHIBIT INTRACELLULAR TRAFFICKING DEFECTS LEADING TO STING HYPERACTIVATION AND ER STRESS**

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**PT15****GAIN-OF-FUNCTION HUMAN UNC93B1 VARIANTS AS A NOVEL CAUSE OF TYPE I INTERFERONOPATHY VIA ENHANCED TLR7 AND TLR8 SIGNALLING**

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**PT16****PERSISTENT IFN SIGNATURE IN PATIENTS WITH PAPA SYNDROME AND ITS REGULATION BY JAK INHIBITION**

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#### **PT17**

#### **SOMATIC GAIN-OF-FUNCTION MUTATION IN TLR7 CAUSES EARLY-ONSET SYSTEMIC LUPUS ERYTHEMATOSUS**

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#### **PT18**

#### **ASSESSMENT OF ADA2 ACTIVITY LEVELS: REPORT FROM THE ITALIAN STUDY GROUP ON DADA2**

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#### **PT19**

#### **NOVEL CDC42 MUTATION REVEALS A MECHANISM OF PYRIN INFLAMMASOME ACTIVATION.**

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#### **PT21**

#### **EVIDENCE FOR DYSREGULATED ERYTHROPOEISIS IN MICE AND HUMANS WITH MEVALONATE KINASE DEFICIENCY**

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#### **PT22**

#### **COMPARISON OF IMMUNOLOGICAL BIOMARKERS AND LUNG HISTOLOGY IN PATIENTS WITH ELEVATED IL18 - PULMONARY ALVEOLAR PROTEINOSIS AND RECURRENT MACROPHAGE ACTIVATION SYNDROME (IL-18PAP-MAS) AND OTHER INFLAMMATORY LUNG DISEASES**

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#### **PT23**

#### **INVESTIGATING NK CELL DEFICIENCY AND DYSFUNCTION IN FAMILIAL MEDITERRANEAN FEVER WITHIN THE IMMUNAID COHORT: A MULTI-OMICS PERSPECTIVE**

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#### **PT24**

#### **GENERATION OF PATIENT-DERIVED IPSCS FOR HYPERIMMUNOGLOBULIN D SYNDROME**

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#### **PT25**

#### **COMPOUND HETEROZYGOUS VARIANTS IN PIGO LEADING TO A NOVEL COMPLEMENT-MEDIATED AUTOINFLAMMATORY DISEASE**

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#### **PT27**

#### **THE GENETIC LANDSCAPE OF PRIMARY IMMUNE REGULATORY DISORDERS IN POLAND**

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#### **PT28**

#### **ANALYSIS OF CLINICAL MANIFESTATIONS ACROSS THE SPECTRUM OF UBA1 MUTATION BURDEN**

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#### PT29

#### A NOVEL REXO2 VARIANT IN A PATIENT WITH LIVEDO RETICULARIS, PALMOPLANTAR ERYTHEMA AND DENTAL DISEASE

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#### PT30

#### MEVALONATE PATHWAY IN AUTOINFLAMMATION: VISUALIZING THE BIOCHEMICAL IMPAIRMENTS OF MEVALONATE KINASE DEFICIENCY

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#### PT31

#### PERFORMANCE OF TARGETED GENE PANEL FOR ROUTINE DIAGNOSIS OF AUTOINFLAMMATORY DISEASES AT THE NATIONAL AMYLOIDOSIS CENTRE

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#### PT32

#### STILL'S DISEASE ASSOCIATED LUNG DISEASE: DATA FROM THE EUROPEAN REGISTRY

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### PT33

#### **EXTRA-OCULAR INVOLVEMENT IN CHILDREN WITH A PHENOTYPE SUGGESTIVE OF OCULAR SARCOIDOSIS**

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### PT34

#### **ADULT-ONSET STILL'S DISEASE: A SINGLE-CENTER REVIEW OF CLINICAL FEATURES, TREATMENT, AND OUTCOMES**

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### PT35

#### **ANAKINRA DERIVED AMYLOIDOSIS DETECTED IN TWO PATIENTS, REPORT FROM THE UK NATIONAL AMYLOIDOSIS CENTRE**

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### PT36

#### **UNRAVELING THE GENETIC AND TRANSCRIPTOMIC DRIVERS OF MONOGENIC AUTOINFLAMMATORY DISEASES IN CHILE: BRIDGING GAPS IN DIAGNOSIS AND TARGETED THERAPY**

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### PT37

#### **SITRAME SYNDROME: INSIGHTS FROM 46 PATIENTS: THE LARGEST COHORT STUDY OF A NOVEL SYSTEMIC AUTOINFLAMMATORY DISEASE**

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### PT38

**THE PREVALENCE AND SPECTRUM OF DAMAGE IN PATIENTS WITH UNDIFFERENTIATED SYSTEMIC AUTOINFLAMMATORY DISEASE**

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**PT39**

**RELEVANCE OF PATTERN RECOGNITION RECEPTOR SIGNALING IN CONTEXT OF MULTI-MEDIATOR INFLAMMATION – TOWARDS UNDERSTANDING A ROLE OF TLR4-DEPENDENT DAMAGE ASSOCIATED MOLECULAR PATTERN SIGNALING IN AUTOINFLAMMATION**

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**PT40**

**PHENOTYPIC AND FUNCTIONAL CHARACTERIZATION OF INNATE LYMPHOID CELLS IN SYSTEMIC JUVENILE IDIOPATHIC ARTHRITIS PATIENTS**

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**PT41**

**TACKLING THE DIAGNOSIS OF HA20 IN CHILDREN: CHALLENGES OF A HIGHLY VARIABLE CLINICAL AND GENETIC SPECTRUM**

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**PT42**

**WHAT CAN WE LEARN FROM THE DRAWING OF CHILDREN WITH AUTOINFLAMMATORY DISEASES?**

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**PT43**

**USING A T-CELL DIRECTED APPROACH IN THE TREATMENT OF DADA2-RELATED NEUTROPENIA RESULTS IN RECOVERY OF MYELOID CELL DEVELOPMENT PRE-TRANSPLANT AND SUCCESSFUL ENGRAFTMENT POST-TRANSPLANT**

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**PT44**

**CLINICAL FEATURES AND EFFICACY OF DIFFERENT MODALITIES OF TREATMENT IN A PATIENT WITH NEMO DELETED EXON 5 AUTOINFLAMMATORY SYNDROME (NDAS)**

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**PT45**

**RENAL INVOLVEMENT IN AUTOINFLAMMATORY DISEASES: DATA FROM THE EUROFEVER REGISTRY (RIAID PROJECT)**

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**PT46**

**SURFING ON VUS: EXPERIENCE IN NORTH-EAST ITALY AND DESCRIPTION OF A COHORT OF ADULT AUTOINFLAMMATORY PATIENTS THROUGH A VALIDATED NEXT-GENERATION SEQUENCING PANEL OF GENES**

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**PT47**

**PREGNANCY OUTCOMES AFTER MATERNAL AND PATERNAL ANTI-IL-1 TREATMENT EXPOSURE IN CRYOPYRIN ASSOCIATED PERIODIC SYNDROMES (CAPS)**

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#### PT48

#### INTERLEUKIN-18 AND INTERLEUKIN-1B BLOCKADE TO CONTROL INFLAMMATION IN PAMI SYNDROME BEFORE AND AFTER HSCT.

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#### PT49

#### CLINICAL, GENETIC, AND IMAGING FEATURES OF AICARDI-GOUTIÈRES SYNDROME IN A LOCAL COHORT IN QATAR

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#### PT50

#### EFFECTS OF CANAKINUMAB TREATMENT ON COMMON LONG-TERM COMPLICATIONS IN AUTOINFLAMMATORY PERIODIC FEVER SYNDROMES – 60-MONTH DATA FROM THE RELIANCE NON-INTERVENTIONAL STUDY

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## PT51

### AUTOINFLAMMATORY DISEASES IN THE NETHERLANDS: CLINICAL AND GENETIC INSIGHTS FROM THE EUROFEVER REGISTRY

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## PT52

### EULAR/PRES ENDORSED RECOMMENDATIONS FOR THE MANAGEMENT OF FAMILIAL MEDITERRANEAN FEVER (FMF): 2024 UPDATE

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## PT53

### A PATIENT WITH A NOVEL DOCK11 MUTATION MANAGED WITH COLCHICINE: A ROLE FOR PYRIN IN DOCK11-ASSOCIATED DISEASE?

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#### PT54

#### Biomarker Evaluation of Disease Activity and Cardiovascular Risk in Familial Mediterranean Fever

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#### PT55

#### Clinical Practice Strategies for the Use of BDMARDs in Colchicine Resistant Familial Mediterranean Fever Across the Countries; A CLIPS Network Interim Analysis

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#### PT56

#### Compound Heterozygosity for MEFV I692DEL and V726A Pathogenic Variants is Associated with a Severe Phenotype of Pyrin-Associated Autoinflammatory Disease with Elevated Interleukin-18

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#### PT57

#### Clinical Features of Patients with Familial Mediterranean Fever over 50 Years of Age:

## A SINGLE-CENTER EXPERIENCE

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## PT58

### CANAKINUMAB TREATMENT IN PATIENTS WITH FAMILIAL MEDITERRANEAN FEVER: A TERTIARY CENTER EXPERIENCE

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## PT59

### CLINICAL PRACTISE STRATEGIES FOR THE DEFINITION OF COLCHICINE RESISTANCE IN FAMILIAL MEDITERRANEAN FEVER ACROSS THE COUNTRIES; A CLIPS NETWORK ANALYSIS

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## PT60

### EPIDEMIOLOGICAL AND ECONOMICAL FACTORS INFLUENCING THE COLCHICINE RESISTANCE DEFINITIONS FOR FAMILIAL MEDITERRANEAN FEVER ACROSS THE COUNTRIES; A CLIPS NETWORK ANALYSIS

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#### PT61

#### TRISOMY 8 MOSAICISM WITH MULTIPLE AUTOINFLAMMATORY MANIFESTATIONS INCLUDING CHRONIC NON-BACTERIAL OSTEOMYELITIS

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#### PT62

#### IL1 BLOCKADE IN COLCHICINE RESISTANT FAMILIAL MEDITERRANEAN FEVER - REAL WORLD DATA

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#### PT63

#### CLINICAL CHARACTERISTICS AND TREATMENT STRATEGIES FOR A20 HAPLOINSUFFICIENCY IN JAPAN: A NATIONAL EPIDEMIOLOGICAL SURVEY

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#### PT64

#### AA AMYLOIDOSIS COMPLICATING SYSTEMIC AUTOINFLAMMATORY DISEASES: DATA FROM THE UK NATIONAL AMYLOIDOSIS CENTRE

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#### PT65

## **UNRAVELING GENETIC COMPLEXITY: DIFFERENT DISEASES IN SIBLINGS WITH SHARED CLINICAL PRESENTATION**

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### **PT66**

#### **NEUROLOGICAL MANIFESTATIONS IN CRYOPYRIN-ASSOCIATED PERIODIC SYNDROMES (CAPS): A RETROSPECTIVE MONOCENTRIC STUDY**

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### **PT67**

#### **TRANSLATIONAL AUTOINFLAMMATORY RESEARCH NETWORK (TARN): A GLOBAL NETWORK APPROACH TO ENHANCING CLINICAL TRIAL READINESS FOR RARE AUTOINFLAMMATORY DISEASES**

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### **PT68**

#### **THREE-YEAR FOLLOW-UP OF CANAKINUMAB DOSE EXTENSION IN CHILDREN WITH COLCHICINE-RESISTANT FMF: PERA-RG EXPERIENCE**

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### **PT69**

#### **BEYOND MEFV: HOW ADDITIONAL AID-ASSOCIATED MUTATIONS SHAPE FAMILIAL MEDITERRANEAN FEVER IN CHILDREN**

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### **PT70**

**GERANYGERANIOL SUPPLEMENTATION LEADS TO AN IMPROVEMENT IN INFLAMMATORY PARAMETERS AND REVERSAL OF THE DISEASE SPECIFIC PROTEIN AND METABOLIC SIGNATURE IN PATIENTS WITH HYPERIGD SYNDROME**

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**PT71**

**PUTTING THE PREDICT-CRFMF SCORE TO THE TEST: PROSPECTIVE PERFORMANCE EVALUATION**

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**PT73**

**DISEASE PRESENTATION, RESPONSE TO TREATMENT AND OUTCOME OF PEDIATRIC AND ADULT PATIENTS WITH DADA2 (DEFICIENCY OF ADENOSINE DEAMINASE 2): RESULTS FROM THE EUROFEVER REGISTRY**

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**PT74**

**CLINICAL OUTCOMES OF BARICITINIB TREATMENT IN AICARDI-GOUTIÈRES SYNDROME: A RETROSPECTIVE COHORT STUDY AT GREAT ORMOND STREET HOSPITAL**

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**PT75**

**PAEDIATRIC AUTOIMMUNE AND AUTOINFLAMMATORY DISEASE-RELATED CATATONIA IS ASSOCIATED WITH ELEVATED CSF INTERFERON-A TITRES AND EFFICIENTLY TREATED WITH IMMUNOADSORPTION IN SEVERE CASES**

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**PT76**

**LOSS OF PSMD7 CAUSES DYSREGULATED PROTEIN DEGRADATION, ENHANCED INFLAMMASOME ACTIVATION, AND INTERFERON RESPONSES**

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## **PT77**

### **NEONATAL-ONSET VASCULITIS DRIVEN BY PATHOGENIC VARIANTS IN THE SRC FAMILY KINASE HAEMATOPOIETIC CELL KINASE (HCK): A REPORT OF TWO FAMILIES AND A NOVEL MUTATION**

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## **PT78**

### **PRE-CLINICAL CHARACTERIZATION AND CLINICAL EVALUATION OF MAS825, AN ANTI-IL-1 BETA / ANTI-IL-18 BISPECIFIC ANTIBODY FOR THE TREATMENT OF INFLAMMASOMOPATHIES**

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## **PT79**

### **PRECLINICAL EVALUATION OF LENTIVIRAL GENE THERAPY FOR THE TREATMENT OF DADA2: ENGRAFTMENT AND BIODISTRIBUTION STUDIES IN HUMANISED NBSGW MICE**

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## **PO001**

### **A NOVEL IKBKB VARIANT INCREASES PROTEIN STABILITY AND DRIVES PERSISTENT AUTOINFLAMMATION**

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## **PO002**

### **PATHOPHYSIOLOGICAL MECHANISMS REGULATING THE PENETRANCE OF MEFV GENE VARIANTS**

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## **PO003**

### **UNRAVELING THE CELLULAR MECHANISMS UNDERLYING INFLAMMASOPATHIES USING GENETIC MOUSE MODELS**

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**PO005****ELEVATED SERUM GASDERMIN D LEVELS IN PATIENTS WITH FAMILIAL MEDITERRANEAN FEVER**

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**PO006****CELL MIGRATION DEFECT IN HYPERIMMUNOGLOBULIN D SYNDROME PATIENT CELLS**

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**PO007****HUMAN PRIMARY MONOCYTES CELL DEATH AND IL-1B PRODUCTION IS DIFFERENTLY REGULATED IN FMF PATIENTS COMPARED TO HEALTHY CONTROLS**

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**PO008****COMPREHENSIVE ANALYSIS OF IMMUNE DYSREGULATION INDUCED BY A NOVEL GAIN-OF-FUNCTION UNC93B1 HOMOZYGOUS MUTATION IN LUPUS**

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**PO009****CYP3A4 REGULATION BY MIR-505-5P: A NOVEL INSIGHT INTO COLCHICINE RESISTANCE IN FMF PATIENTS**

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**PO010****DISTINCT SERUM IMMUNOREACTIVITY PATTERNS IN MULTIPLE SCLEROSIS AND BEHÇET'S DISEASE: A COMPARATIVE ANALYSIS**

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**PO011****INVESTIGATION OF THE MECHANISMS UNDERLYING THE ALTERED EXPRESSION OF MIR-197-3P IN PATIENTS WITH FAMILIAL MEDITERRANEAN FEVER**

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## PO012

### ASSESSMENT OF C26:0 LYSOPHOSPHATIDYLCHOLINE AND CHITOTRIOSIDASE LEVELS IN PATIENTS WITH DEFICIENCY OF ADENOSINE DEAMINASE 2

F. Poswar<sup>1,2,\*</sup>, L. G. da Silva<sup>3</sup>, L. A. da Silva<sup>4,5</sup>, G. Iop<sup>4,5</sup>, M. Botton<sup>2,6</sup>, M. Riegel-Giugliani<sup>4,7</sup>, R. Giugliani<sup>1,2,4,7,8</sup>

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## PO015

### PSTPIP1 P.E250K VARIANT ATTENUATES PROTEIN EXPRESSION AND PODOSOME FORMATION IN PATIENT-DERIVED MACROPHAGES

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## PO016

### THE ROLE OF FATTY ACIDS IN FAMILIAL MEDITERRANEAN FEVER

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## PO017

### INVESTIGATION OF INNATE LYMPHOID CELLS IN CHILD PATIENTS DIAGNOSED WITH FAMILIAL MEDITERRANEAN FEVER WITH SPONDYLARTHRITIS

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## PO019

### ASEPTIC ABSCESS SYNDROME: LINKS TO MONOGENIC AUTOINFLAMMATORY DISEASES

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## PO020

### THE IMPACT OF NEXT GENERATION SEQUENCING: TEN YEARS' EXPERIENCE OF THE GREAT ORMOND STREET HOSPITAL AUTOINFLAMMATION CENTRE OF EXCELLENCE (GOSH-ACE)

F. Price-Kuehne<sup>1,\*</sup>, A. Burleigh<sup>1</sup>, E. Omoyinmi<sup>1</sup>, Y. Hong<sup>1</sup>, K. McLellan<sup>1</sup>, K. Nazmutdinova<sup>1</sup>, D. Eleftheriou<sup>1,2</sup>, P. Brogan<sup>1,2</sup>

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#### **PO021**

#### **TWO NOVEL GAIN-OF-FUNCTION VARIANTS IN ELF4 IN PATIENTS WITH SYSTEMIC UNDEFINED AUTOINFLAMMATORY DISEASE**

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#### **PO022**

#### **A RARE AUTOINFLAMMATORY SYNDROME ASSOCIATED WITH A C2ORF69 FRAMESHIFT MUTATION: A CASE REPORT**

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#### **PO023**

#### **A NOVEL STAT4 VARIANT AS THE POTENTIAL CAUSE OF A LONG-LASTING CASE OF DISABLING PANSCLEROTIC MORPHEA**

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#### **PO024**

#### **A NOVEL NONSENSE MUTATION IN LPIN2 ASSOCIATED WITH MAJEDD SYNDROME: CASE REPORT AND INSIGHTS INTO GENE EXPRESSION**

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#### **PO025**

#### **WHOLE EXOME SEQUENCING IN PAEDIATRIC-ONSET COGAN'S SYNDROME**

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#### **PO026**

#### **LATE ONSET OF AN AUTOINFLAMMATORY DISEASE: IDENTIFICATION AND FUNCTIONAL CHARACTERIZATION OF A MOSAIC VARIATION OF NLRC4**

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#### PO027

#### EVALUATING GENETIC VARIANTS AND THEIR CLINICAL CORRELATIONS IN UNDIFFERENTIATED SYSTEMIC AUTOINFLAMMATORY DISEASES (USAIDS)

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#### PO028

#### CLINICAL UTILITY OF EXOME SEQUENCING IN ADULTS WITH AUTOINFLAMMATORY DISORDERS: A PROSPECTIVE STUDY ON 138 PATIENTS

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#### PO029

#### MEFV MUTATIONAL SPECTRUM AND CLINICAL MANIFESTATIONS IN GEORGIAN FMF PATIENTS

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#### PO031

#### HARDY-WEINBERG DISEQUILIBRIUM OF MEFV DISEASE ASSOCIATED GENOTYPES IN A LARGE ISRAELI COHORT OF INDIVIDUALS TESTED FOR PRENATAL CARRIER STATE OF GENETIC DISEASES

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#### PO033

#### DEFINITION OF DISEASE PHENOTYPES IN PEDIATRIC SAPHO SYNDROME: A NATIONAL MULTICENTRIC STUDY

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#### **PO034**

#### **MACROPHAGIC ACTIVATION SYNDROME IN STILL'S DISEASE: A MULTICENTER OBSERVATIONAL COHORT STUDY**

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#### **PO035**

#### **USE AND SAFETY OF DIFFERENT BISPHOSPHONATES IN CHRONIC RECURRENT MULTIFOCAL OSTEOMYELITIS AND A COMPARISON OF THE SIDE EFFECT PROFILE BETWEEN CHRONIC RECURRENT MULTIFOCAL OSTEOMYELITIS AND OTHER UNDERLYING DIAGNOSES**

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#### **PO036**

#### **ARE BLOOD MONOCYTES USEFUL IN DIFFERENTIATING PFAPA FROM FMF? INSIGHTS INTO THEIR LIMITS AND COMPLEMENTARY MARKERS**

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#### **PO037**

#### **WORLDWIDE EVALUATION OF CLINICAL PRACTICE STRATEGIES (CLIPS) FOR LUNG INVOLVEMENT IN STILL'S DISEASE WITHIN THE JIR-CLIPS NETWORK: A COST ACTION**

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#### **PO038**

## **SYNDROME OF UNDIFFERENTIATED RECURRENT FEVER (SURF): CLINICAL AND GENETIC INSIGHTS FROM A MONOCENTRIC HOMOGENEOUS COHORT OF 101 PATIENTS**

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### **PO039**

## **DECODING THE SYNDROME OF UNDIFFERENTIATED RECURRENT FEVER: CLINICAL INSIGHTS, BIOMARKERS, AND TREATMENT OUTCOMES FROM A NATIONAL UK AUTOINFLAMMATORY CENTRE**

C. Papadopoulou<sup>1,\*</sup>, E. Omoyinmi<sup>1</sup>, D. Rowczenio<sup>1</sup>, B. Sen<sup>1</sup>, M. Wood<sup>1</sup>, H. Lachmann<sup>1</sup>

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### **PO040**

## **THE ASSESSMENT OF IL-15 IN PATIENTS WITH STILL'S DISEASE, IN VITRO AND EX-VIVO FINDINGS**

I. Di Cola<sup>1,\*</sup>, M. Vomero<sup>2</sup>, C. Castellini<sup>1</sup>, O. Berardicurti<sup>2</sup>, M. Breccia<sup>1</sup>, L. Navarini<sup>2</sup>, P. Cipriani<sup>1</sup>, R. Giacomelli<sup>2</sup>, P. Ruscitti<sup>1</sup>

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### **PO041**

## **PREDICTIVE FACTORS FOR RELAPSE IN ADULT-ONSET STILL'S DISEASE: A RETROSPECTIVE COHORT STUDY**

I. Y. Çakır<sup>1</sup>, Y. Mahmutoğlu<sup>2</sup>, R. C. Kardaş<sup>1</sup>, E. Uslu<sup>2</sup>, S. Sezer<sup>2</sup>, M. E. Yayla<sup>2</sup>, H. Küçük<sup>1</sup>, A. Ateş<sup>2</sup>, A. Tufan<sup>1,\*</sup>, M. Turgay<sup>2</sup>, A. Erden<sup>1</sup> on behalf of --- Select One ---

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### **PO042**

## **EVALUATION OF THE PATIENTS WITH CHRONIC NON-BACTERIAL OSTEOMYELITIS BASED ON MAGNETIC RESONANCE IMAGING**

A. Paç Kısaarslan<sup>1,\*</sup>, S. Özdemir Çiçek<sup>1</sup>, Z. F. Karaman<sup>2</sup>, E. Kayhan<sup>1</sup>, A. Yekedüz Bülbül<sup>1</sup>, G. Ozan Altaş<sup>1</sup>, P. Garipçin<sup>1</sup>, H. Taştanoglu<sup>1</sup>, E. Esen<sup>1</sup>, C. Arslanoğlu<sup>1</sup>, M. H. Poyrazoğlu<sup>1</sup>

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### **PO043**

## **STILL'S DISEASE OVER 2 DECADES: LEARNING FROM THE PAST**

A. Kozáková<sup>1,\*</sup>, K. Šingelová<sup>1</sup>, N. Husáková<sup>1</sup>, P. Šeferna<sup>1</sup>, Š. Fingerhutová<sup>1</sup>, P. Doležalová<sup>1</sup>

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### **PO046**

## **ADULT PATIENTS WITH UNCLASSIFIED SYSTEMIC AUTOINFLAMMATORY DISEASE - A SINGLE CENTER CASE SERIES**

J. An<sup>1,\*</sup>

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**PO047****CUTANEOUS MANIFESTATIONS IN A CHILEAN COHORT WITH SYSTEMIC AUTOINFLAMMATORY DISEASES**C. Downey<sup>1,2</sup>, G. Moreno<sup>3</sup>, J. De la Cruz<sup>3</sup>, A. Borzutzky<sup>4</sup>, M. C. Poli<sup>1,3,5,\*</sup><sup>1</sup>Departamento de Pediatría, Clínica Alemana de Santiago, <sup>2</sup>Unidad de Dermatología, Hospital Luis Calvo Mackenna,<sup>3</sup>Programa de Inmunogenética e Inmunología Traslacional, Instituto de Ciencias e Innovación en Medicina, Facultad de Medicina, Clínica Alemana Universidad del Desarrollo, <sup>4</sup>Departamento de Pediatría, Facultad de Medicina, Pontificia Universidad Católica de Chile, <sup>5</sup>Unidad de Inmunología y Reumatología, Hospital de Niños Dr. Roberto del Río, Santiago, Chile**PO048****WORLDWIDE ASSESSMENT OF CLINICAL PRACTICE STRATEGIES (CLIPS) IN STILL'S DISEASE TREATMENT****THROUGH THE JIR-CLIPS NETWORK: A COST ACTION**S. Ferreira Azevedo<sup>1</sup>, R. Naddei<sup>2</sup>, F. Aguiar<sup>3,4</sup>, M. Jouret<sup>5</sup>, C. Girard-Guyonvarc'h<sup>6</sup>, Y. Vyzhga<sup>7</sup>, F. Ramos<sup>8</sup>, C. Costa Lana<sup>9</sup>, R. Guedri<sup>10</sup>, A. Lefevre-Utile<sup>11,12</sup>, D. Hadef<sup>13</sup>, J. M. Mosquera Angarita<sup>14</sup>, S. Ozen<sup>15</sup>, S. Sahin<sup>16</sup>, S. Hashad<sup>17</sup>, K. Daghor-Abbacij<sup>18</sup>, D. Foell<sup>19</sup>, S. Georgin-Lavialle<sup>20,\*</sup> on behalf of CEREMAIA & ERN RITA, K. Theodoropoulou<sup>21</sup> on behalf of for the JIR-CLIPS Network<sup>1</sup>Rheumatology, Unidade Local de Saude da regiao de Aveiro, Aveiro, Portugal, <sup>2</sup>Department of translational Medical Sciences, University of Napoles Frederico II, Napoles, Italy, <sup>3</sup>Adult and Pediatric Rheumatology, Unidade Local de Saude de São João, <sup>4</sup>., Faculdade de Medicina da Universidade do Porto, Porto, Portugal, <sup>5</sup>Pediatric Nephrology, Rheumatology, Dermatology Unit, Hospices Civils de Lyon, Lyon, France, <sup>6</sup> Department of Medicine, Division of Rheumatology, University of Geneva, Geneva, Switzerland, <sup>7</sup>., National Pirogov Memorial Medical University, Vinnytsya, Ukraine, <sup>8</sup>Pediatric Rheumatology, Unidade Local de Saude de Santa Maria, Faculdade de Medicina da Universidade de Lisboa, Lisbon, Portugal, <sup>9</sup>Rheumatology, Medical School of Medicine, Federal University of Minas Gerais State (UFMG), Belo Horizonte, Brazil, <sup>10</sup>Pediatric rheumatology unit, Béchir Hamza Children's Hospital- Faculty of Medicine of Tunis, Tunis, Tunisia, <sup>11</sup>Service de pédiatrie, Département femme-mère-enfant (DFME), <sup>12</sup>., Faculté de biologie et de médecine de l'Université de Lausanne, Lausanne, Switzerland, <sup>13</sup>Faculty of Medicine, , Batna 2 University, Batna, Algeria, <sup>14</sup>, PediaRheumatology Uni, Sant Joan de Déu Children's Hospital, Barcelona, Spain, <sup>15</sup>Pediatrics, Hacettepe University, Ankara, <sup>16</sup>Pediatric Rheumatology unit, University Cerrahpasa, Istambul, Türkiye, <sup>17</sup>Pediatric Rheumatology unit, Tripoli Children Hospital, University of Tripoli, Tripoli, Libya, <sup>18</sup>Department of Internal medicine, Bab El Oued University Hospital center, University of Algiers, Algeria Rheumatology and Immunology, Algeria, Algeria, <sup>19</sup>Pediatric Rheumatology and Immunology, University Children's Hospital Münster, Münster, Germany, <sup>20</sup>Internal Medicine Department, DMU3ID, , Sorbonne University, Tenon Hospital, AP-HP, Paris, France, <sup>21</sup> Unit of Pediatric Immunology, Allergology and Rheumatology, Lausanne University Hospital, Lausanne, Switzerland**PO049****CHRONIC RECURRENT MULTIFOCAL OSTEOMYELITIS AND LUNG INVOLVEMENT: REPORT OF TWO CASES**C. Conti<sup>1,\*</sup>, R. Papa<sup>1</sup>, B. Cafferata<sup>2</sup>, V. G. Vellone<sup>2</sup>, S. Volpi<sup>1</sup>, M. Gattorno<sup>1</sup>, R. Caorsi<sup>1</sup><sup>1</sup>Rheumatology and autoinflammatory diseases, <sup>2</sup>Pathology Unit, Istituto Giannina Gaslini, Genoa, Italy**PO050****MACROPHAGE ACTIVATION SYNDROME IN A PATIENT WITH CLERICUZIO-TYPE POIKILODERMA NEUTROPENIA SYNDROME**A. Yekedüz Bülbül<sup>1</sup>, C. Arslanoğlu<sup>2</sup>, P. Garipçin<sup>1</sup>, A. Özcan<sup>3</sup>, E. Ünal<sup>4</sup>, A. Paç Kısaarslan<sup>1,\*</sup>, M. H. Poyrazoğlu<sup>1</sup><sup>1</sup>Pediatric Rheumatology, Erciyes University Faculty of Medicine, Kayseri, <sup>2</sup>Pediatric Rheumatology, Kocaeli City Hospital, Kocaeli, <sup>3</sup>Pediatric Oncology and Hematology, Erciyes University Faculty of Medicine, Kayseri, <sup>4</sup>Pediatric Oncology and Hematology, Medical Point Hospital, Gaziantep, Türkiye

**PO051****CLINICAL AND RADIOLOGICAL FEATURES OF MANDIBULAR CHRONIC NONBACTERIAL OSTEOMYELITIS (CNO): A RETROSPECTIVE CASE SERIES**

S. Sutnga<sup>1,\*</sup>, A. Khan<sup>1,2</sup>, H. Panwala<sup>3</sup>, S. Garg<sup>3</sup>, B. Jankharia<sup>4</sup>, R. Khubchandani<sup>1</sup>

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**PO052****DISPENSED PRESCRIPTIONS OF ADHD MEDICATIONS TO CHILDREN WITH PFAPA**

K. Rydenman<sup>1,2,\*</sup>, A. Fasth<sup>1,3</sup>, S. Berg<sup>1,3</sup>, P. Wekell<sup>1,2,3</sup>

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**PO053****MONITORING COLCHICINE EFFECTIVITY IN CHILDREN WITH PFAPA BY USING AUTOINFLAMMATORY DISEASES ACTIVITY INDEX (AIDAI) SCORES**

L. Koru<sup>1,\*</sup>, F. Haslak<sup>1</sup>

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**PO054****ATTITUDES TOWARD GENETIC TESTING IN PFAPA SYNDROME: UNVEILING CLINICAL TRENDS FROM THE JIR-CLIPS SURVEY**

H. E. Sönmez<sup>1,\*</sup>, C. Vinit<sup>2,3</sup>, N. Toplak<sup>4</sup>, O. Boyarchuk<sup>5</sup>, M. Gattorno<sup>6</sup>, G. Ozomay Baykal<sup>7</sup>, K. Pateras<sup>8</sup>, E. D. Batu<sup>9</sup>, F. Hofer<sup>10</sup>, C. Jurcut<sup>11</sup>, M. Rodrigues<sup>12</sup>, O. Gilliaux<sup>13</sup>, K. Laskari<sup>14</sup>, M. Santos Faria<sup>15</sup>, R. Craveiro da Costa<sup>16</sup>, R. Craveiro da Costa<sup>16</sup>, M. Hofer<sup>17</sup> on behalf of JIR-CliPS

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**PO056****CARD8-FS VARIANT IN SLOVAK COHORT OF PFAPA PATIENTS**

L. Kapustova<sup>1,\*</sup>, A. Bobcakova<sup>2</sup>, D. Kapustova<sup>1</sup>, B. Šlenker<sup>1</sup>, A. Markocsy<sup>1</sup>, O. Petrovicova<sup>2</sup>, E. Jurkova Malicherova<sup>1</sup>, M. Jesenak<sup>2</sup>

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#### PO057

#### EVALUATION OF SECOND-LINE TREATMENTS IN CHRONIC RECURRENT MULTIFOCAL OSTEOMYELITIS

S. Pacaud<sup>1,\*</sup>, A. Carbasse<sup>1</sup>, O. Prodhomme<sup>1</sup>, M. DELPONT<sup>1</sup>, T. A. TRAN<sup>2</sup>, E. Jeziorski<sup>1</sup>

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#### PO058

#### USE AND SAFETY OF DIFFERENT BISPHOSPHONATES IN CHRONIC RECURRENT MULTIFOCAL OSTEOMYELITIS AND A COMPARISON OF THE SAFETY PROFILE: GOSH EXPERIENCE

O. Kul Cinar<sup>1,2,\*</sup>, M. Yildiz<sup>1,3</sup>, N. Maduaka<sup>1</sup>, Q. Wu<sup>1</sup>, C. A. Pilkington<sup>1,2</sup>, S. Compeyrot-Lacassagne<sup>1,2</sup>

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#### PO059

#### YAO SYNDROME IN CHILDREN: A PEDIATRIC CASE SERIES FROM AUTOINFLAMMATION REFERENCE CENTER TÜBINGEN

F. Kara Eroglu<sup>1,\*</sup>, O. Satirer<sup>1</sup>, O. Buzoianu<sup>1</sup>, C. Reiser<sup>2</sup>, J. Kuemmerle-Deschner<sup>1</sup>

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#### PO060

#### CHRONIC MACROPHAGE ACTIVATION SYNDROME IN STILLS DISEASE: A CASE REPORT OF SUCCESSFUL TREATMENT WITH JAK AND INTERLEUKIN-1 INHIBITORS

R. Bin Salman<sup>1,\*</sup>, S. Habiballah<sup>2</sup>, A. Aldahlawi<sup>1</sup>, A. Alrasheed<sup>3</sup>, M. Nashawi<sup>4</sup>

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#### PO061

#### DIAGNOSIS, TREATMENT AND MONITORING OF PEDIATRIC BEHCET'S DISEASE (BD) AND BD-RELATED PHENOTYPES ON IDENTIFIED MONOGENIC MIMICS: A SYSTEMATIC REVIEW PROTOCOL

M. Romano<sup>1,\*</sup>, N. Zitoun<sup>2</sup>, E. Sag<sup>3</sup>, D. Poddighe<sup>4</sup>, D. Piskin<sup>5</sup>, I. Tugal-Tutkun<sup>6</sup>, I. Aksentijevich<sup>7</sup>, I. Kone-Paut<sup>8</sup>, E. Demirkaya<sup>2</sup> on behalf of Pediatric Behcet Disease Task Force

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#### PO062

#### NEUROLOGIC PRESENTATIONS IN ELDERLY PATIENTS WITH YAO SYNDROME

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**PO063****DE NOVO TGFBR1 MUTATION ASSOCIATED WITH ATYPICAL AUTOINFLAMMATORY AND PERIOSTEAL INVOLVEMENT: DIAGNOSTIC CHALLENGES AND THERAPEUTIC APPROACHES**M. Rossano<sup>1,\*</sup>, L. Baselli<sup>1</sup>, F. Di Stasio<sup>1</sup>, F. A. Vianello<sup>1</sup>, S. Torreggiani<sup>2</sup>, F. Minoia<sup>1</sup>, G. Filocamo<sup>1</sup><sup>1</sup>Pediatric Rheumatology, Fondazione IRCCS Cà Granda Ospedale Maggiore Policlinico, Milan, Italy, <sup>2</sup>Inflammatory Disease Section, National Human Genome Research Institute, National Institutes of Health, Bethesda, MD, United States**PO064****A NOVEL CASE OF P2XR7 ASSOCIATED AUTOINFLAMMATORY DISEASE SUCCESSFULLY TREATED WITH ANTI-IL1 THERAPY**S. D. Arık<sup>1,\*</sup>, B. Menentoğlu<sup>1</sup>, Ö. Akgün<sup>1</sup>, N. Aktay Ayaz<sup>1</sup> on behalf of İstanbul<sup>1</sup>Pediatric Rheumatology, İstanbul University Faculty of Medicine , İstanbul, Türkiye**PO065****A PATIENT WITH VERY EARLY ONSET INFLAMMATORY BOWEL DISEASE (VEO-IBD), HEPATITIS, SPECIFIC ANTIBODY DEFICIENCY AND A VARIANT OF UNCERTAIN SIGNIFICANCE IN PIK3CD**S. M. S. Lee<sup>1,\*</sup>, W. L. J. Tan<sup>1</sup>, X. R. Lim<sup>1</sup><sup>1</sup>Rheumatology, Allergy & Immunology, Tan Tock Seng Hospital, Singapore, Singapore**PO066****TRNT1 RELATED AUTOINFLAMMATORY SYNDROME IN A PATIENT WITH PRIMARY CILIARY DYSKINESIA**S. Di Gennaro<sup>1,\*</sup>, R. Naddei<sup>1</sup>, F. Della Casa<sup>2</sup>, A. Petraroli<sup>2</sup>, M. Alessio<sup>1</sup><sup>1</sup>Department of Translational Medical Science, Paediatric Rheumatology, <sup>2</sup>Department of Translational Medical Science, Division of Autoimmune & Allergic Diseases, University Federico II, Naples, Italy**PO067****NEMO-NDAS: CLINICAL DIVERSITY AND THERAPEUTIC CHALLENGES IN PEDIATRIC CASES**F. Faron<sup>1,\*</sup>, F. delion<sup>2</sup>, P. Mertz<sup>3</sup>, G. Boursier<sup>4</sup>, A. T. Nguyen<sup>1</sup>, V. Hentgen<sup>1</sup><sup>1</sup>pediatric (ceremaia), CH Versailles, Versailles, <sup>2</sup>pediatric, chu point à pitre, point à pitre, <sup>3</sup>rheumatology, aphp, paris ,<sup>4</sup>genetic, chu Montpellier, Montpellier, France**PO068****HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS DURING A RELAPSE OF A VEXAS SYNDROME**B. Farrulo<sup>1,\*</sup>, A. Boissais<sup>1</sup>, A. Bigot<sup>1</sup>, B. Thoreau<sup>1</sup>, F. Maillot<sup>1</sup>, A. Audemard-Verger<sup>1</sup><sup>1</sup> Internal medicine, University Hospital Center of Tours, Tours, France**PO071****A RETROSPECTIVE ANALYSIS OF THREE PATIENTS WITH A VEXAS-LIKE SYNDROME LACKING DETECTABLE UBA1 MUTATIONS**K. Tsujimoto<sup>1,\*</sup>, A. Maeda<sup>2</sup>, Y. Kirino<sup>2</sup>, A. Kumanogoh<sup>1</sup><sup>1</sup>Respiratory Medicine and Clinical Immunology, The University of Osaka, Osaka, <sup>2</sup>Stem Cell and Immune Regulation, Yokohama City University Graduate School of Medicine, Yokohama, Japan**PO072****CHRONIC RECURRENT WHEALS AND APHTHOUS ULCERS ASSOCIATED WITH AN MEFV K695R MUTATION**L. S. Herzog<sup>1,2,\*</sup>, K. Krause<sup>2,3</sup>, P. Kolkhir<sup>2,3</sup>, H. Bonnekoh<sup>2,3</sup><sup>1</sup>Institute of Allergology, Charité – Universitätsmedizin Berlin, Corporate Member of Freie Universität Berlin and Humboldt-Universität zu Berlin, <sup>2</sup>Immunology and Allergology, Fraunhofer Institute for Translational Medicine and

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**PO073**

**CHALLENGING DIAGNOSTIC AND THERAPEUTIC JOURNEY IN VEXAS SYNDROME: A CASE REPORT**

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**PO074**

**AN UNUSUAL CASE OF WRISTS AND ANKLES "BOGGY SYNOVITIS": AUTOINFLAMMATION LINKING AUTOIMMUNITY?**

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**PO075**

**CASE REPORT: IDENTIFICATION OF POST-ZYGOTIC MOSAICISM WITH A PATHOGENIC TNFRSF1A VARIANT IN A PATIENT WITH ELEVATED INFLAMMATORY MARKERS**

B. Bangol<sup>1,\*</sup>, B. Vignolo<sup>1</sup>, J. Seytter<sup>2</sup>, O. Wachter<sup>2</sup>, K. Hirv<sup>3</sup>

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**PO076**

**AN UNUSUAL KIDNEY PRESENTATION IN GENETICALLY-CONFIRMED FAMILIAL MEDITERRANEAN FEVER**

G. Frisoni<sup>1</sup>, A. Omenetti<sup>2,\*</sup>, L. Caponi<sup>1</sup>, F. Pugliese<sup>1</sup>, S. Gatti<sup>1</sup>, L. Santoro<sup>1</sup>, I. D'Alba<sup>3</sup>, B. Lattanzi<sup>2</sup>, M. E. Lionetti<sup>1</sup>

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**PO077**

**EVALUATION OF A DOMINANTLY INHERITED MEFV VARIANT IN A FAMILY WITH FMF-LIKE PHENOTYPE**

C. Esperanze<sup>1,\*</sup>, J. J. Chae<sup>1</sup>, W. Lee<sup>2</sup>, G. Wood<sup>1</sup>, N. Deutch<sup>1</sup>, B. Matthiasardottir<sup>1</sup>, D. L. Kastner<sup>1</sup>, I. Aksentijevich<sup>1</sup>

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**PO078**

**FATAL RHEUMATOID VASCULITIS ASSOCIATED WITH ANTIPHOSPHOLIPID ANTIBODY POSITIVITY LEADING TO CRITICAL LIMB ISCHEMIA**

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**PO079**

**EFFICACY OF HIGH DOSES INTRAVENOUS ANAKINRA IN TWO PAEDIATRIC CASES OF TAFRO SYNDROME**

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**PO080****A RARE CONDITION THAT CAN BE MISTAKEN FOR VASCULITIS: PROLIDASE ENZYME DEFICIENCY**R. Duran<sup>1</sup>, D. Yıldırım<sup>1</sup>, A. Tufan<sup>1,\*</sup><sup>1</sup>Rheumatology, Gazi University, Ankara, Türkiye**PO081****A RARE PRESENTATION OF OPTIC DISC EDEMA DIAGNOSIS OF CRYOPYRIN-ASSOCIATED PERIODIC SYNDROMES (CAPS): A CASE REPORT**S. Atamyildiz Ucar<sup>1</sup>, Z. gokdemir<sup>1</sup>, B. Sozeri<sup>1,\*</sup><sup>1</sup>Peditric Rheumatology, Umraniye Trainig and Research Hospital, Istanbul, Türkiye**PO082****RECURRENT MAS-HLH IN A 68-YEAR-OLD WOMAN WITH ADULT-ONSET STILL'S DISEASE AND STXBP2 MUTATION**S. Uziel<sup>1,2</sup>, N. Fanadka<sup>3</sup>, Y. Kitay Cohen<sup>1,3</sup>, M. Yeruchimovich<sup>3</sup>, A. Zoref-Lorenz<sup>4</sup>, S. Kivity<sup>1,5,\*</sup><sup>1</sup>School of Medicine, Tel Aviv University, Tel Aviv, <sup>2</sup>Pediatric Rheumatology Institute, <sup>3</sup>Department of Internal Medicine C,<sup>4</sup>Hematology Institute, <sup>5</sup>Rheumatology Institute, Meir Medical Center, Kfar Saba, Israel**PO083****AN ATYPICAL PRESENTATION OF ANTSYNTETASE SYNDROME**R. Lethem<sup>1,\*</sup>, S. Lacassagne<sup>2</sup><sup>1</sup>Paediatric Rheumatology, <sup>2</sup>Pediatric Rheumatology, Great Ormond Street Hospital for Children NHS Foundation Trust, London, United Kingdom**PO084****CASE REPORT OF A FEMALE PATIENT WITH OVER 25 YEARS OF RECURRENT FEVER - STILL MORE QUESTIONS THAN ANSWERS**E. Więsik-Szewczyk<sup>1,\*</sup>, K. Jahnz-Rozyk<sup>1</sup><sup>1</sup>Department of Internal Medicine, Pneumonology, Allergology, Clinical Immunology and Rare Diseases, Military Institute of Medicine National Research Institute, Warsaw, Poland**PO085****A PATIENT WITH A VARIANT OF UNKNOWN SIGNIFICANCE (VUS) ON AUTOINFLAMMATORY PHOSPHOLIPASE CG2 (PLCG2) GENE: NEW VARIANT WITH A RARE CLINICAL PRESENTATION?**I. D'Alba<sup>1</sup>, B. Lattanzi<sup>2</sup>, A. Omenetti<sup>2,\*</sup>, L. Pacillo<sup>3</sup>, M. Chiriaco<sup>4</sup>, L. Caponi<sup>5</sup>, L. Antonini<sup>1</sup>, B. Bruschi<sup>1</sup>, S. Gobbi<sup>1</sup>, V. Petroni<sup>1</sup>, P. Coccia<sup>1</sup><sup>1</sup>Department of Pediatric Onco Hematology, <sup>2</sup>Department of Mother and Child Health, Pediatric Unit, AOU Marche, Salesi Children's Hospital, Ancona, <sup>3</sup>Research Unit of Clinical Immunology and Vaccinology, IRCCS Bambino Gesù Children's Hospital, <sup>4</sup>Department of Systems Medicine, University of Rome Tor Vergata, Rome, <sup>5</sup>Pediatric Clinic, Polytechnic University of Marche, Ancona, Italy**PO086****DEFICIENCY OF ADENOSINE DEAMINASE 2: A TALE OF TWO PATIENTS, ONE MUTATION**A. Dudaklı<sup>1</sup>, S. D. Arik<sup>1,\*</sup>, B. Menentoğlu<sup>1</sup>, G. Kavrul Kayaalp<sup>1</sup>, A. Doğru<sup>1</sup>, B. Başer Taşkın<sup>1</sup>, V. Guliyeva<sup>2</sup>, Ö. Akgün<sup>1</sup>, N. Aktay Ayaz<sup>1</sup><sup>1</sup>Pediatric Rheumatology, İstanbul University İstanbul Faculty of Medicine, İstanbul, Türkiye, <sup>2</sup>Pediatric Rheumatology, Liv Bona Dea Hospital Bakü, Bakü, Azerbaijan

**PO087****DIFFICULT-TO-TREAT RARE DISEASES: A CHALLENGING CASE OF PASH SYNDROME REFRACTORY TO CONVENTIONAL THERAPY**S. Bindoli<sup>1,\*</sup>, P. Sfriso<sup>1</sup><sup>1</sup>Department of Medicine, University of Padova, Rheumatology Unit, Padova, Italy**PO089****PYODERMA GANGRENOsum (PG) PRECEDING TAKAYASU ARTERITIS (TA) IN A PEDIATRIC PATIENT: AN ONGOING UNSOLVED SAGA - A CASE REPORT AND LITERATURE REVIEW**S. Sutnga<sup>1,\*</sup>, A. Khan<sup>1,2</sup>, R. Khubchandani<sup>1</sup><sup>1</sup>Pediatric Rheumatology, SRCC Children's Hospital, <sup>2</sup>Pediatric Rheumatology, MRR Children's Hospital, Mumbai, India**PO090****PERINATAL ENCEPHALOPATHY IN INFANT WITH TREX1 VARIANT**I. Romankevych<sup>1,\*</sup>, K. Plona<sup>2</sup>, R. Hopkin<sup>2</sup>, A. Grom<sup>1</sup>, H. Brunner<sup>1</sup><sup>1</sup>Rheumatology, <sup>2</sup>Human Genetics , Cincinnati Children's Hospital Medical Center, Cincinnati, United States**PO091****MEVALONATE KINASE DEFICIENCY: AN UNDERDIAGNOSED CAUSE OF INFLAMMATION-RELATED ISCHEMIC STROKE – CASE REPORT AND NOVEL GENE MUTATION**L.-N. Hamidi<sup>1,\*</sup>, J. C. Drda<sup>2</sup>, M. Belhocine<sup>3</sup>, H. Elfassy<sup>4</sup>, S. Ducharme-Benard<sup>3</sup>, M. Chayer-Lanthier<sup>1</sup>, S. Lanthier<sup>1</sup><sup>1</sup>Department of neurology, Hôpital du Sacré-Coeur de Montréal, Montréal, Canada, <sup>2</sup>Department of microbiology, University of Pittsburgh, Pittsburgh, United States, <sup>3</sup>Department of internal medicine, <sup>4</sup>Department of immunology, Hôpital du Sacré-Coeur de Montréal, Montréal, Canada**PO092****NEUROLOGICAL PHENOTYPES OF SOCS1 HAPLOINSUFFICIENCY: INSIGHTS FROM FUNCTIONAL AND HISTOLOGICAL INVESTIGATIONS**S. Palmeri<sup>1,2,\*</sup>, I. Prigione<sup>2</sup>, F. Schena<sup>2</sup>, A. Bertoni<sup>2</sup>, F. Penco<sup>2</sup>, P. Bocca<sup>2</sup>, S. Massucco<sup>1</sup>, C. Venturi<sup>3</sup>, A. Schenone<sup>4</sup>, M. Lanciotti<sup>5</sup>, M. Miano<sup>5</sup>, C. Matucci-Cerinic<sup>1,2</sup>, G. Viglizzo<sup>6</sup>, R. Papa<sup>2</sup>, R. Caorsi<sup>1,2</sup>, M. Gattorno<sup>2</sup>, S. Volpi<sup>1,2</sup><sup>1</sup>Department of Neuroscience, Rehabilitation, Ophthalmology, Genetics, Maternal and Child Health (DINOGLMI), University of Genoa, <sup>2</sup>Rheumatology and Autoinflammatory Diseases Unit, IRCCS Istituto Giannina Gaslini, <sup>3</sup>Pathology Unit,<sup>4</sup>Neurology Unit, IRCCS Ospedale Policlinico San Martino, <sup>5</sup>Hematology Unit, Department of Pediatric Hematology/Oncology, <sup>6</sup>Dermatology Unit, IRCCS Istituto Giannina Gaslini, Genoa, Italy**PO093****STEROID-SENSITIVE NEUROINFLAMMATION IN 2 SIBLINGS WITH AUTOIMMUNE LYMPHOPROLIFERATIVE SYNDROME**M. Auletta<sup>1</sup>, C. Sottani<sup>1</sup>, G. Prencipe<sup>2</sup>, L. L. Slcignano<sup>3</sup>, E. Verrecchia<sup>3</sup>, R. Iorio<sup>1</sup>, G. Silvestri<sup>1</sup>, E. Tabolacci<sup>4</sup>, E. Sangiorgi<sup>4</sup>, D. Rigante<sup>5</sup>, M. Genuardi<sup>4</sup>, R. Manna<sup>3,\*</sup><sup>1</sup>Neurosciences, UCSC, <sup>2</sup>Rheumatology, OPBG, <sup>3</sup>Internal Medicine, <sup>4</sup>Genomic Medicine, <sup>5</sup>Pediatrics, UCSC, Rome, Italy**PO094****(MONO-) GENETIC MIMICS OF BEHÇET'S DISEASE**P. Kurt<sup>1,\*</sup>, B. Bader-Meunier<sup>2</sup>, G. Baulier<sup>3</sup>, M.-C. Besse<sup>4</sup>, A. Chausset<sup>5</sup>, S. Georgin-Lavialle<sup>6</sup>, A. L. Jurquet<sup>7</sup>, I. Koné-Paut<sup>8,9</sup>, C. Labrèze<sup>10</sup>, E. Merlin<sup>5</sup>, F. Morice-Picard<sup>11</sup>, P. Pillet<sup>11</sup>, H. Reumaux<sup>12</sup>, E. Riviere<sup>13</sup>, D. Saadoun<sup>9,14</sup>, F. A. Aeschlimann<sup>\*1</sup>, G. Boursier<sup>\*15</sup>

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#### **PO096**

#### **CENTRAL NERVOUS SYSTEM (CNS) VASCULITIS IN ACTIVATED PHOSPHOINOSITIDE 3-KINASE DELTA SYNDROME 1 (APDS1) TREATED WITH HEMATOPOIETIC STEM CELL TRANSPLANTATION (HSCT): A 39-MONTH FOLLOW-UP AND LITERATURE REVIEW**

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#### **PO097**

#### **AUTOINFLAMMATORY MANIFESTATIONS IN A PATIENT WITH TYPE II D2-HYDROXYGLUTARIC ACIDURIA**

R. Mackeh<sup>1</sup>, N. Al Anni<sup>2</sup>, B. Al Adba<sup>2</sup>, M. J. A. Osman<sup>1</sup>, S. Bout-Tabaku<sup>2,\*</sup>, B. Lo<sup>1,3</sup>

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#### **PO098**

#### **A CROSS-SECTIONAL OVERVIEW OF BEHÇET'S DISEASE MANAGEMENT: A TUNISIAN EXPERIENCE**

W. Helali<sup>1,2</sup>, R. Bourguiba<sup>1,2,\*</sup>, H. Bettaieb<sup>1,2</sup>, T. Jomni<sup>1,2</sup>, M. Boudokhane<sup>1,2</sup>, S. Bellakhal<sup>1,2</sup>

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#### **PO100**

#### **THE PATHOLOGY OF THE SKIN, LYMPH NODES, LIVER, AND BONE MARROW AND RELATED CLINIC-BIOLOGICAL FEATURES IN PATIENTS WITH SEVERE SYSTEMIC JUVENILE IDIOPATHIC ARTHRITIS : A CASE SERIES OF 11 PATIENTS**

E. Sun<sup>1,\*</sup>, V. Guillet<sup>2</sup>, B. Bader-Meunier<sup>1</sup>, P. Quartier-Dit-Maire<sup>1</sup>, M.-L. Frémond<sup>1</sup>, P. Schneider<sup>3</sup>, A. François<sup>4</sup>, V. Hentgen<sup>5</sup>, C. Glaser<sup>6</sup>, D. Moshous<sup>1</sup>, I. Melki<sup>7</sup>, A. Welfringer<sup>8</sup>, J. Bruneau<sup>2</sup>, P. Drabent<sup>2</sup>, B. Fournier<sup>1</sup>

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#### **PO101**

#### **CLINICAL OUTCOME AND QUALITY OF LIFE IN PATIENTS WITH ARPC1B DEFICIENCY MANAGED CONSERVATIVELY OR WITH ALLOGENEIC HEMATOPOIETIC STEM CELL TRANSPLANTATION - ON BEHALF OF THE ESID/EBMT INBORN ERRORS WORKING PARTY**

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## PO102

### AN UPDATE ON THE CLINGEN MONOGENIC AUTOINFLAMMATORY DISEASES EXPERT CURATION PANELS: A FRAMEWORK FOR INTERPRETING GENETIC FINDINGS IN AUTOINFLAMMATORY DISEASES

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## PO103

### PREGNANCY OUTCOMES IN AUTOINFLAMMATORY DISEASES: A PROSPECTIVE STUDY OF 117 CASES, INCLUDING 79 WITH FAMILIAL MEDITERRANEAN FEVER.

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**PO104****TRACKING COLCHICINE COMPLIANCE IN CHILDREN WITH FMF: CAN HAIR COLCHICINE DOSING PROVIDE USEFUL INFORMATION?**V. Hentgen<sup>1,2,\*</sup>, C. VINIT<sup>3,4</sup>, I. Elhani<sup>5,6</sup>, M. LABOURET<sup>4</sup>, P. Dusser<sup>7</sup>, E. Abe<sup>8</sup>, I. A. Larabi<sup>8,9</sup>, J.-C. 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Lachmann<sup>1</sup><sup>1</sup>National Amyloidosis Centre, Royal Free London NHS Foundation Trust, <sup>2</sup>Paediatric Rheumatology, Great Ormond Street Hospital For Children NHS Foundation Trust, London, United Kingdom**PO106****LONG-TERM SAFETY AND EFFICACY OF COLCHICINE AND ANTI-IL-1 BLOCKERS IN FMF: RESULTS FROM THE EUROFEVER MULTICENTER OBSERVATIONAL STUDY**S. La Bella<sup>1</sup>, M. Bustaffa<sup>1</sup>, Y. Bayindir<sup>2</sup>, G. Amaryan<sup>3</sup>, R. Gallizzi<sup>4</sup>, E. Papadopoulou-Alataki<sup>5</sup>, G. Fabio<sup>6</sup>, N. Assalia<sup>7</sup>, G. Amarylio<sup>7</sup>, M. Jesenak<sup>8</sup>, L. Breda<sup>9</sup>, J. Anton<sup>10</sup>, E. Legger<sup>11</sup>, M. Alessio<sup>12</sup>, G. Simonini<sup>13</sup>, D. Rigante<sup>14</sup>, L. Obici<sup>15</sup>, J. Kuemmerle-Deschner<sup>16</sup>, O. Kasapcopur<sup>17</sup>, A. Insalaco<sup>18</sup>, M. Glerup<sup>19</sup>, J. Frenkel<sup>20</sup>, J. Brunner<sup>21</sup>, G. Horneff<sup>22</sup>, J. Sánchez-Manubens<sup>23</sup>, S. Bakkaloglu<sup>24</sup>, L. Cantarini<sup>25</sup>, A. Spagnolo<sup>4</sup>, S. Alataki<sup>5</sup>, M. Carrabba<sup>6</sup>, A. Porreca<sup>26</sup>, R. Caorsi<sup>1</sup>, N. Ruperto<sup>27</sup>, M. Gattorno<sup>1,\*</sup>, S. 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#### **PO107**

#### **COMBINATION OF BIOLOGICS AND JAK INHIBITORS IN THE TREATMENT OF REFRACTORY SYSTEMIC AUTOINFLAMMATORY DISEASES**

A. Tufan<sup>1,2,\*</sup>, D. Stone<sup>2</sup>, P. Hoffmann<sup>2</sup>, T. Romeo<sup>2</sup>, L. Wilson<sup>2</sup>, N. Deuitch<sup>2</sup>, C. Kozycki<sup>2</sup>, K. Manthiram<sup>2</sup>, D. Schwartz<sup>2</sup>, A. Ombrello<sup>2</sup>, D. Kastner<sup>2</sup> on behalf of MD

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#### **PO108**

#### **AA AMYLOIDOSIS RELATED TO MONOGENIC AUTOINFLAMMATORY DISEASES IN FRANCE: A COHORT STUDY OF 77 CASES**

M. El Moussaoui<sup>1,2,\*</sup>, R. Bourguiba<sup>2</sup>, L. Savey<sup>2</sup>, J.-J. Boffa<sup>2</sup>, G. Grateau<sup>2</sup>, D. Buob<sup>3</sup>, S. Georgin-Lavialle<sup>2</sup>

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#### **PO110**

#### **"BALANCING IL-1 BLOCKADE IN DIRA: A TALE OF REMISSION, FLARE, AND THERAPEUTIC PRECISION"**

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#### **PO111**

#### **EVALUATION OF PULMONARY INVOLVEMENT IN COLCHICINE RESISTANT FMF PATIENTS**

E. Hepkaya<sup>1</sup>, N. Z. Özaslan<sup>2</sup>, T. Özer<sup>3</sup>, B. Öksel<sup>2</sup>, Y. Anık<sup>3</sup>, N. Şahin<sup>2,\*</sup>, H. E. Sönmez<sup>2</sup>

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#### **PO112**

#### **GLOBAL MORTALITY OF FRENCH PATIENTS WITH SYSTEMIC AUTOINFLAMMATORY DISEASES**

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#### **PO113**

#### **BURDEN OF FATIGUE IN CRYOPYRIN-ASSOCIATED PERIODIC SYNDROMES (CAPS)**

Ö. Satirer<sup>1,\*</sup>, Y. Satirer<sup>2</sup>, A.-K. Gellner<sup>3</sup>, S. M. Benseler<sup>4</sup>, J. Kümmerle-Deschner<sup>1</sup>

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#### **PO114**

## **TYPE I INTERFERON SCORE AS A BIOMARKER OF DISEASE ACTIVITY IN ADA2 DEFICIENCY**

S. Federici<sup>1,\*</sup>, M. Wouters<sup>2</sup>, V. Matteo<sup>3</sup>, A. Hombrouck<sup>2</sup>, G. Prencipe<sup>3</sup>, L. Moens<sup>2</sup>, F. De Benedetti<sup>1,3</sup>, I. Meyts<sup>2,4</sup>, A. Insalaco<sup>1</sup>

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## **PO115**

### **CLINICAL PHENOTYPE AND LABORATORY MARKERS IN PATIENTS AFFECTED BY A20 HAPLOINSUFFICIENCY (HA20): A CASE SERIES FROM TWO ITALIAN CENTERS**

L. De Nardi<sup>1,\*</sup>, S. Federici<sup>1</sup>, E. De Martino<sup>2</sup>, M. F. Natale<sup>1</sup>, C. Celani<sup>1</sup>, V. Matteo<sup>1</sup>, G. Prencipe<sup>1</sup>, A. Tesser<sup>2</sup>, A. Pin<sup>2</sup>, F. De Benedetti<sup>1</sup>, A. Tommasini<sup>2</sup>, A. Insalaco<sup>1</sup>

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## **PO116**

### **GENETICS, CLINICAL CHARACTERISTICS, AND MANAGEMENT OF FAMILIAL MEDITERRANEAN FEVER IN DIVERSE POPULATIONS: A COMPARATIVE STUDY OF BARI AND ISTANBUL**

L. Yagci<sup>1,\*</sup>, B. Kilic<sup>1</sup>, A. Ozturk<sup>1</sup>, N. Jaber<sup>2</sup>, M. Khalil<sup>2</sup>, A. Stella<sup>3</sup>, A. Di Ciaula<sup>2</sup>, S. Ugurlu<sup>1</sup>, P. Portincasa<sup>2</sup>

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## **PO117**

### **DIAGNOSTIC DELAY IN SYSTEMIC AUTOINFLAMMATORY DISEASE: PRELIMINARY RESULTS FROM THE EUROFEVER REGISTRY**

D. Piskin<sup>1</sup>, M. Romano<sup>2</sup>, N. Zitoun<sup>2</sup>, R. Caorsi<sup>3</sup>, Y.-H. Choi<sup>1</sup>, M. Gattorno<sup>3</sup>, E. Demirkaya<sup>2,\*</sup> on behalf of EUROFEVER Registry Collaborators

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## **PO118**

### **ATTACK TRIGGERS IN CHILDHOOD FAMILIAL MEDITERRANEAN FEVER**

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## **PO119**

### **IFIH1 GENE MUTATION AS A CAUSE OF SEVERE HYPERINFLAMMATION – CASE REPORT**

M. D. Genšor, Mph.<sup>1,\*</sup>, Z. Bystricanova<sup>2</sup>, E. Fronkova<sup>3</sup>, M. Jesenák<sup>1,4,5</sup>

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**PO120****THE ROLE OF DEMOGRAPHICS IN THE NATURE OF FAMILIAL MEDITERRANEAN FEVER AND ITS OUTCOMES: A COMPARATIVE INTERNATIONAL STUDY**B. Han Egeli<sup>1,2</sup>, K. Marzan<sup>1,3</sup>, T. Stein<sup>4</sup>, M. Heshin-Bekenstein<sup>5,6,\*</sup><sup>1</sup>Pediatric Rheumatology, Children's Hospital of Los Angeles, <sup>2</sup>Pediatric Rheumatology, <sup>3</sup>School of Medicine, University of Southern California, Los Angeles, United States, <sup>4</sup>Pediatrics, <sup>5</sup>Pediatric Rheumatology Service, Dana Children's Hospital of Tel Aviv Medical Center, <sup>6</sup>School of Medicine, Faculty of Medical and Health Sciences, Tel Aviv University, Tel Aviv, Israel**PO122****NEUROLOGICAL MANIFESTATIONS AND VASCULOPATHY IN PLCG2-ASSOCIATED ANTIBODY DEFICIENCY AND IMMUNE DYSREGULATION (APLAID): REPORT OF TWO CASES**K. Asna Ashari<sup>1,2</sup>, R. MacRae<sup>3</sup>, M. Ashoor<sup>1</sup>, D. Hoytema<sup>1</sup>, C. Platt<sup>1</sup>, P. See<sup>4</sup>, A. Kielian<sup>3</sup>, A. Danehy<sup>5</sup>, R. Kumar<sup>6</sup>, M. Gorman<sup>3</sup>, F. Dedeoglu<sup>1,\*</sup>, M. Andzelm<sup>3</sup><sup>1</sup>Immunology, Boston Children's Hospital, Boston, United States, <sup>2</sup>Rheumatology, Tehran University of Medical Sciences, Tehran , Iran, Islamic Republic Of, <sup>3</sup>neurology, <sup>4</sup>Neurosurgery, Cerebrovascular Surgery and Interventions Center, <sup>5</sup>radiology, <sup>6</sup>Dana Farber, Boston Children's Hospital, Boston, United States**PO123****GENOTYPE-PHENOTYPE CORRELATIONS IN CRYOPYRIN-ASSOCIATED PERIODIC SYNDROME AMONG TURKISH PATIENTS IN GERMANY AND TURKEY: BEYOND BORDERS**Ö. Satirer<sup>1</sup>, Y. Bayindir<sup>2,\*</sup>, V. Cam<sup>2</sup>, S. Türkmen<sup>3</sup>, B. Sözeri<sup>3</sup>, J. Kümmel-Deschner<sup>1</sup>, S. Özén<sup>2</sup><sup>1</sup>Pediatric Rheumatology, Tübingen University, Tübingen, Germany, <sup>2</sup>Pediatric Rheumatology, Hacettepe University, Ankara, <sup>3</sup>Pediatric Rheumatology, Umraniye Training and Research Hospital, Istanbul, Türkiye**PO124****EVERYDAY LIFE OF PATIENTS WITH AUTOINFLAMMATORY PERIODIC FEVER SYNDROMES DURING LONG-TERM TREATMENT WITH CANAKINUMAB – 5-YEAR DATA FROM THE RELIANCE NON-INTERVENTIONAL STUDY**J. Rech<sup>1,\*</sup>, J. Henes<sup>2</sup>, N. Blank<sup>3</sup>, A. Pankow<sup>4</sup>, T. Kallinich<sup>5</sup>, K.-G. Birgit<sup>6</sup>, P. T. Oommen<sup>7</sup>, T. Krickau<sup>8</sup>, C. Schuetz<sup>9</sup>, A. Janda<sup>10</sup>, I. Foeldvari<sup>11</sup>, G. Horneff<sup>12</sup>, F. Weller-Heinemann<sup>13</sup>, M. Hufnagel<sup>14</sup>, T. Kümpfel<sup>15</sup>, F. Meier<sup>16</sup>, F. Dressler<sup>17</sup>, D. Windschall<sup>18</sup>, I. Andreica<sup>19</sup>, M. Borte<sup>20</sup>, M. Krusche<sup>21</sup>, M. Fiene<sup>22</sup>, J. B. Kuemerle-Deschner<sup>23</sup><sup>1</sup>Department of Rheumatology and Immunology, University Hospital Erlangen, Erlangen, <sup>2</sup>Center of Interdisciplinary Rheumatology, Immunology and autoimmune diseases (INDIRA) and department of Internal Medicine I, University Hospital Tuebingen, Tübingen, <sup>3</sup>Division of Rheumatology, Internal Medicine V, University Hospital Heidelberg, Heidelberg,<sup>4</sup>Department of Rheumatology and Clinical Immunology, <sup>5</sup>Department of Pediatric Respiratory Medicine, Immunology and Critical Care Medicine, Charité-Universitätsmedizin Berlin, Berlin, <sup>6</sup>Department of Internal Medicine, Division ofNephrology, University Hospital of Giessen and Marburg, Marburg, <sup>7</sup>Department of Pediatric Oncology, Hematology and Clinical Immunology, Division of Pediatric Rheumatology, Medical Faculty, Heinrich-Heine-University Duesseldorf, Düsseldorf, <sup>8</sup>Pediatrics, Erlangen, Germany; <sup>9</sup>Centre for rare diseases Erlangen (ZSEER), Erlangen, Germany; <sup>10</sup>DZI (Deutsches Zentrum für Immuntherapie), Germany, <sup>11</sup>Friedrich-Alexander University Erlangen-Nuernberg (FAU), Erlangen,<sup>12</sup>Department of Pediatrics, Medizinische Fakultät Carl Gustav Carus, <sup>13</sup>Technische Universität Dresden, Dresden, <sup>14</sup>Department of Pediatrics and Adolescent Medicine, Ulm, University Medical Center Ulm, Ulm, <sup>15</sup>Pediatric and Adolescence Rheumatology, Hamburg Centre for Pediatric and Adolescence Rheumatology,, Hamburg, <sup>16</sup>Department of Pediatrics, Asklepios Kinderklinik Sankt Augustin, , Sankt Augustin, <sup>17</sup>Division of Pediatric Rheumatology, Prof. Hess Children's Hospital Bremen, Bremen, <sup>18</sup>Division of Pediatric Infectious Diseases and Rheumatology, University Medical Center, Medical Faculty, University of Freiburg, Freiburg, <sup>19</sup>Institute of Clinical Neuroimmunology, University Hospital, Ludwig-Maximilians Universität München, München, <sup>20</sup>ITMP, Fraunhofer Institute for Translational Medicine and

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#### **PO125**

#### **CLINICAL FEATURES AND OUTCOMES OF A SMALL VEXAS SYNDROME PATIENTS' COHORT**

C. Salvado<sup>1,2</sup>, M. Henriques<sup>1,3</sup>, A. Lamas<sup>1,4,5</sup>, A. R. Gonçalves<sup>6</sup>, M. Oliveira<sup>6</sup>, L. Regadas<sup>4,5,7</sup>, R. Faria<sup>1,4,5,\*</sup>

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#### **PO126**

#### **FAMILIAL MEDITERRANEAN FEVER AND PAPASH: IL-1 BLOCKERS RECOVER COMPLEX HIDRADENITIS**

M. Carrabba<sup>1,\*</sup>, C. Moltrasio<sup>2</sup>, M. A. Zarantonello<sup>3</sup>, S. Serafino<sup>1</sup>, C. A. Maronese<sup>2</sup>, A. V. Marzano<sup>4</sup>, G. Fabio<sup>1</sup>

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#### **PO127**

#### **EVOLVING PHENOTYPIC AND GENOTYPIC SPECTRUM OF HUMAN ISG15 AND USP18 DEFICIENCIES**

A. Alsaleem<sup>1,\*</sup>, W. Abdulghaffar<sup>1</sup>, L. Akbar<sup>2</sup>, M. Alhassan<sup>2</sup>, A. Almutairi<sup>3</sup>, F. Alkhars<sup>4</sup>, S. Almayouf<sup>1</sup>

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#### **PO128**

#### **IMPORTANCE OF POTENTIAL PROTHROMBOTIC STATE MARKERS IN BEHÇET'S DISEASE AND CORRELATION WITH VASCULAR ULTRASOUND**

H. Karadeniz<sup>1,2</sup>, T. Mammadov<sup>3</sup>, M. Cerit<sup>3</sup>, R. Kardas<sup>1</sup>, A. Avanoglu Guler<sup>1</sup>, D. Yıldırım<sup>1</sup>, I. Vasi<sup>1</sup>, H. Pasaoglu<sup>4</sup>, A. Erden<sup>1</sup>, H. Kucuk<sup>1</sup>, B. Goker<sup>1</sup>, M. Ozturk<sup>1</sup>, A. Tufan<sup>1,5,\*</sup>

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#### **PO129**

#### **NEUTRALIZING ANTIBODIES AND ANTI-TUMOR NECROSIS FACTOR (TNF) MONOCLONAL ANTIBODY MEDICATIONS**

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#### **PO130**

## **CARDIOVASCULAR COMORBIDITIES IN PATIENTS WITH FAMILIAL MEDITERRANEAN FEVER**

S. Karup<sup>1</sup>, S. N. Baspinar<sup>2</sup>, B. Ayci<sup>1</sup>, F. N. Azman<sup>3</sup>, A. Akyel<sup>3</sup>, Y. Guler<sup>3</sup>, S. Ugurlu<sup>4,\*</sup>

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### **PO132**

#### **AN UNEXPECTEDLY HIGH PREVALENCE OF FAMILIAL MEDITERRANEAN FEVER IN SLOVAKIA – RESULT FROM NATIONAL AWARENESS CAMPAIGN**

M. Jesenak<sup>1,2,3,\*</sup>, E. Malicherova Jurkova<sup>1</sup>, A. Bobcakova<sup>1,2,3</sup>, K. Hrubiskova<sup>4</sup>, O. Petrovicova<sup>1</sup>, L. Kapustova<sup>1</sup>, T. Dallos<sup>5</sup>, A. Markocsy<sup>1,3</sup>

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### **PO133**

#### **CUTANEOUS MANIFESTATIONS OF AUTOINFLAMMATORY DISEASES IN PATIENTS WITH GENETIC TESTING**

M. Beketova<sup>1,\*</sup>, S. Salugina<sup>1</sup>, E. Fedorov<sup>1</sup>, E. Zakharova<sup>2</sup>, A. Lila<sup>1,3</sup>

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### **PO134**

#### **DADA2: THE FIRST REPORT OF A MULTICENTER EGYPTIAN EXPERIENCE**

Y. El Chazli<sup>1,\*</sup>, P. Lee<sup>2</sup>, Q. Zhou<sup>3</sup>, J. I. Arostegui<sup>4</sup>, J. Wang<sup>3</sup>, E. Fahmi<sup>5</sup>, A. Beltagy<sup>6</sup>, A. Sobh<sup>7</sup>, N. Othman<sup>8</sup>, N. Radwan<sup>9</sup>, N. Ragab<sup>10</sup>, N. Abdelrahman<sup>1</sup>, H. Gadelrab<sup>10</sup>, W. Shoman<sup>10</sup>

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### **PO135**

#### **PERIODONTITIS IN DADA2 PATIENTS WITH SEVERE NEUTROPENIA**

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### **PO136**

#### **DETERMINATION OF THE RELATIONSHIP BETWEEN SERUM PROTEIN 14-3-3 ETA LEVELS AND CLINICAL FEATURES OF THE DISEASE IN PATIENTS WITH FAMILIAL MEDITERRANEAN FEVER**

A. Avci<sup>1</sup>, D. Yıldırım<sup>2,3</sup>, A. Erden<sup>2</sup>, H. Kucuk<sup>2</sup>, A. Tufan<sup>2,\*</sup>, M. A. Ozturk<sup>2</sup>

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**PO138****ST2 AS AN INFLAMMATORY AND CARDIOVASCULAR MARKER IN FMF-ASSOCIATED AMYLOIDOSIS**

S. Karup<sup>1</sup>, O. C. Kilinc<sup>1</sup>, F. N. Azman<sup>2</sup>, T. Ayalti<sup>1</sup>, O. Kizilkaya<sup>2,\*</sup>, O. Er<sup>1</sup>, Z. S. Furkan<sup>2</sup>, I. M. Bolayirli<sup>3</sup>, S. Ugurlu<sup>4</sup>

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**PO140****REAL-LIFE DATA ON TAPERING OF ANTI-IL-1 THERAPY IN PATIENTS WITH MEVALONATE KINASE DEFICIENCY (MKD)**

B. Sozeri<sup>1,\*</sup>, E. tunc<sup>1</sup> on behalf of İSTANBUL

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**PO141****INFLAMMATORY LINEAR VERRUCOUS EPIDERMAL NEVUS (ILVEN) - A NEW SKIN MANIFESTATION IN A CHILD WITH BLAU SYNDROME**

P. Pimpale Chavan<sup>1,2,\*</sup>, S. Khubchandani<sup>3</sup>, R. Khubchandani<sup>2</sup>

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<sup>3</sup>Department of Histopathology, Jaslok Hospital and Research Center, Mumbai, India

**PO142****A TIGHT ROPE (D) WALK IN A CHILD WITH ACUTE ABDOMEN**

A. Jamal<sup>1</sup>, D. Suri<sup>1,\*</sup> on behalf of Dr Gayathri CV, Dr Dev Desai, Dr Abarna Thangaraj, Dr Vignesh Pandiarajan, Dr Ankur Jindal, Dr Rakesh Kumar Pilania, Dr Amit Rawat, Dr Manpreet Dhaliwal, Dr Saniya Sharma, Prof Surjit Singh

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**PO143****VEXAS SYNDROME AND INFECTIONS: ANALYSIS OF A MULTICENTRIC COHORT AND BRIEF LITERATURE REVIEW**

S. Bindoli<sup>1,\*</sup>, I. Guidea<sup>1</sup>, R. Bixio<sup>2</sup>, L. Iorio<sup>1</sup>, R. Padoan<sup>1</sup>, P. Sfriso<sup>1</sup>

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**PO144****VALIDATION OF THE COLCHICINE RESISTANCE PREDICTION CRITERIA FROM THE TURPAID COHORT TO THE JIR COHORT: A MULTICENTER DESCRIPTIVE ANALYSIS**

P. Mertz<sup>1,\*</sup>, I. Koné-Paut<sup>2</sup>, D. Dan<sup>3</sup>, S. Roque<sup>4</sup>, K. Theodoropoulou<sup>5</sup>, J. Pachlopnik<sup>6</sup>, B. Bader-Meunier<sup>7</sup>, A. Belot<sup>8</sup>, C. Dumaine<sup>9</sup>, C. Kevorkian-Verguet<sup>10</sup>, P. Pillot<sup>11</sup>, S. Georgin-Lavialle<sup>12</sup>, V. Hentgen<sup>1</sup>

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<sup>11</sup>Pediatrics Department, CHU Bordeaux, Bordeaux, <sup>12</sup>Internal Medicine, Tenon Hospital, Paris, France

**PO145****EXPANDING THE GENETIC AND CLINICAL SPECTRUM OF NFKB1 VARIANTS IN CHINESE PATIENTS WITH PRIMARY IMMUNODEFICIENCY**

J. Zhang<sup>1,\*</sup>, H. Cai<sup>2</sup>, T. Zhu<sup>2</sup>, M. Shen<sup>1</sup>

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#### PO146

#### A CASE OF CANDLE SYNDROME PRESENTING AS SERONEGATIVE POLYARTHRITIS

S. Cooray<sup>1,\*</sup>, A. Kelly<sup>2</sup>, M. Gattens<sup>3</sup>, R. Armstrong<sup>3</sup>, W. Kelsall<sup>4</sup>, G. Ambegaonkar<sup>5</sup>, L. Allen<sup>6</sup>, M. Cook<sup>7</sup>, H. Baxendale<sup>7</sup>, P. Bale<sup>1</sup>, K. Armon<sup>1</sup>

<sup>1</sup>Department of Paediatric Rheumatology, <sup>2</sup>Department of Paediatric Oncology and Haematology, <sup>3</sup>Department of Clinical Genetics, <sup>4</sup>Department of Cardiology, <sup>5</sup>Department of Paediatric Neurology, <sup>6</sup>Department of Ophthalmology, <sup>7</sup>Department of Immunology, Cambridge University Hospitals NHS Foundation Trust, Cambridge, United Kingdom

#### PO147

#### FAMILIAL MEDITERRANEAN FEVER: DISEASE SEVERITY AND AMYLOIDOSIS IN AN EGYPTIAN COHORT

W. Ibrahim Shoman<sup>1,\*</sup>, Y. El Chazli<sup>2</sup>, F. Moursi<sup>3</sup>, N. Abdelsalam<sup>3</sup>, S. Elshafey<sup>3</sup>, N. Ragab<sup>1</sup>, H. Gadelrab<sup>1</sup>, A. Bakry<sup>1</sup>, M. Allam<sup>4</sup>, G. Elderiny<sup>1</sup>

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#### PO149

#### A CASE WITH DIAGNOSIS OF MACROPHAGE ACTIVATION SYNDROME AND FAMILIAL MEDITERRANEAN FEVER

E. Kayhan<sup>1,\*</sup>, A. Yekedüz Bülbül<sup>1</sup>, S. Özdemir Çiçek<sup>1</sup>, A. Paç Kısaarslan<sup>1</sup>, M. H. Poyrazoğlu<sup>1</sup>

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#### PO150

#### A PARTICULAR PHENOTYPE IN TWO PATIENTS WITH SIDEROBLASTIC ANEMIA, B-CELL IMMUNODEFICIENCY, PERIODIC FEVERS AND DEVELOPMENTAL DELAY

M. T. Bataneant<sup>1,2,\*</sup>, A. Chirita-Emandi<sup>3,4</sup>, E. Boeriu<sup>1,2</sup>, M. Baica<sup>5</sup>, B. Caprioru<sup>2</sup>, D. Matea<sup>2</sup>, P. Urtila<sup>1,2</sup>

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#### PO151

#### INFLAMMATORY BOWEL DISEASE IN NEONATAL-ONSET MULTISYSTEM INFLAMMATORY DISEASE: A GENETIC RISK ASSESSMENT

S. Alehashemi<sup>1,\*</sup>, A. de Jesus<sup>1</sup>, S. Srinivasan<sup>2</sup>, Z. Fazal<sup>2</sup>, J. Yan<sup>2</sup>, A. Oler<sup>2</sup>, J. Berghout<sup>2</sup>, I. Fuss<sup>3</sup>, R. Goldbach-Mansky<sup>1</sup>

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#### PO152

#### DIAGNOSTIC CHALLENGE: TACKLING VEXAS SYNDROME IN LOW RESOURCE COUNTRIES

R. Bourguiba<sup>1,\*</sup>, S. Bellakhal<sup>1</sup>, S. Ndongo<sup>2</sup>, A. Allaoui<sup>3</sup>, H. Jilani<sup>4</sup>, F. Said<sup>5</sup>, I. Naceur<sup>5</sup>, T. El Murr<sup>6</sup>, I. Ksontini<sup>7</sup>, G. Khellaf<sup>8</sup>, R. Ben Amor<sup>9</sup>, K. Abbaci<sup>10</sup>, L. Ben Jemaa<sup>11</sup>, W. Ammour<sup>12</sup>, C. Jurcut<sup>13</sup>

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Slim, University Tunis el Manar, Faculté de médecine de Tunis, La Marsa, <sup>5</sup>Internal medicine, Hopital la Rabta, Tunis, Tunisia, <sup>6</sup>Internal medicine, Hopital Hotel Dieu de France, Beyrouth, Lebanon, <sup>7</sup>Internal medicine, Internal Medecine office, La Marsa, Tunisia, <sup>8</sup>Nephrology, Babeloued university hospital center,mohamed lamine Debaghine hospita, Alger, Algeria, <sup>9</sup>Hematology, Hematology Office, Tunis, Tunisia, <sup>10</sup>Internal medicine, Benyoucef Benkhedda faculty of medicine , Alger, Algeria, <sup>11</sup>Genetics, Hopital Mongi slim , La Marsa, Tunisia, <sup>12</sup>Internal medicine, Ibn Sina Hospital, Rabat, Morocco, <sup>13</sup>Internal medicine, Dr. Carol Davila Central University Emergency Military Hospital, Bucharest, Romania

#### PO153

#### ADULT PATIENTS WITH FAMILIAL MEDITERRANEAN FEVER - A SINGLE CENTER CASE SERIES

J. An<sup>1,\*</sup>, M. Bassuony<sup>2</sup>, H. Goldhar<sup>3</sup>, P. Lahiry<sup>4</sup>, A. Makhzoum<sup>5</sup>, J. Malette<sup>6</sup>, J. Shu<sup>7</sup>, A. Kovacs-Litman<sup>8</sup> on behalf of ON<sup>1</sup>Rheumatology, Hospital for Sick Children , Toronto, <sup>2</sup>Internal Medicine, Thunder Bay Regional Health Sciences Centre, Thunder Bay, <sup>3</sup>Rheumatology, William Osler Health Centre, Brampton, <sup>4</sup>Rheumatology, 1440 Bathurst, Toronto, <sup>5</sup>Rheumatology, 1235 Trafalgar Rd, Oakville , <sup>6</sup>Rheumatology, Windsor Regional Hospital, Windsor, <sup>7</sup>Rheumatology, 3333 Brimley Road, Scarborough, <sup>8</sup>Rheumatology, Sunnybrook Health Sciences Centre, Toronto, Canada

#### PO154

#### PATH TO DIAGNOSIS IN FAMILIAL MEDITERRANEAN FEVER (FMF)

S. Karup<sup>1</sup>, D. Polat<sup>2</sup>, E. Dincsoy<sup>2</sup>, F. N. Azman<sup>2</sup>, M. E. Cokrak<sup>2</sup>, E. Berkiten<sup>2</sup>, S. Ugurlu<sup>3,\*</sup>  
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#### PO155

#### TNF-RECEPTOR ASSOCIATED PERIODIC SYNDROME: AN ANALYSIS OF SLOVAKIAN COHORT OF TRAPS PATIENTS

B. Šlenker<sup>1,2,\*</sup>, M. Jeseňák<sup>1,2,3</sup>, L. Kapustová<sup>1,2</sup>, D. Kapustová<sup>1,2</sup>, P. Bánovčin<sup>1</sup>, K. Hrubišková<sup>4</sup>, V. Vargová<sup>5</sup>, A. Bobčáková<sup>2,3</sup>, D. Genšor<sup>1</sup>, E. Jurková Malicherová<sup>1,2</sup>, A. Markocsy<sup>1,2</sup>, O. Petrovičová<sup>1,2</sup>

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#### PO157

#### FAMILIAL MEDITERRANEAN FEVER AMONG ADULT PATIENTS: A MULTICENTRIC STUDY IN TUNISIA

R. Bourguiba<sup>1,\*</sup>, W. skouri<sup>2</sup>, I. Naceur<sup>3</sup>, M. S. Hamdi<sup>4</sup>, B. Arfaoui<sup>5</sup>, I. Chaabene<sup>6</sup>, M. Somai<sup>7</sup>, Z. Tayeb<sup>8</sup>, N. Adaily<sup>9</sup>, F. Chelbi<sup>10</sup>, M. Trabelsi<sup>11</sup>, H. Jilani<sup>12</sup>, L. Ben Jemaa<sup>12</sup>, T. Larbi<sup>13</sup>, A. Kefi<sup>4</sup>

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#### PO158

#### NEMO-NDAS: DIVERSE CLINICAL PRESENTATIONS AND PENETRANCE IN THREE PEDIATRIC PATIENTS

A. A. Alrasheed<sup>1,2,3,\*</sup>, F. Alroqi<sup>4,5,6</sup>, J. Alqanatish<sup>1,2</sup>, W. sewairi<sup>1,2</sup>, M. Al Marri<sup>7</sup>, K. Almutairy<sup>7</sup>, L. alahmadi<sup>8</sup>

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#### **PO159**

#### **A CASE OF MUCKLE-WELLS SYNDROME WITH HYPERTROPHIC PACHYMENTINGITIS IN WHICH A NOVEL NLRP3 GENE VARIANT WAS IDENTIFIED**

H. Ida<sup>1,\*</sup>, Y. Hidaka<sup>1</sup>, R. Ogino<sup>2</sup>, K. Izawa<sup>2</sup>, T. Yasumi<sup>2</sup>, R. Nishikomori<sup>3</sup>

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#### **PO160**

#### **VEXAS SYNDROME IN TUNISIA: A RARE DIAGNOSIS, A FIRST REPORT**

O. Chouchene<sup>1</sup>, S. Arfa<sup>2</sup>, M. Ben Brahim<sup>2</sup>, A. Barhoumi<sup>2</sup>, R. Bourguiba<sup>3,\*</sup>, H. Jilani<sup>3</sup>, S. Mbarek<sup>4</sup>, O. Berriche<sup>2</sup>

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#### **PO161**

#### **ANAKINRA IS A FAST AND EFFECTIVE TREATMENT OPTION IN SUBSIDING FMF EPISODES AND THEREBY, IN DECREASING HOSPITALIZATION RATES : FUTURE PROSPECTS FOR READILY AVAILABLE, HOME-USE ANAKINRA INJECTORS**

E. Kilic Konte<sup>1</sup>, N. Akay<sup>1</sup>, U. Gul<sup>1</sup>, E. Ozturk<sup>1</sup>, S. Kaplan Kilic<sup>1</sup>, G. Nuran Cengiz<sup>1</sup>, K. Karaahmetli<sup>1</sup>, E. Aslan<sup>1</sup>, A. Gunalp<sup>1</sup>, F. Haslak<sup>1</sup>, M. Yildiz<sup>1</sup>, A. Adrovic Yildiz<sup>1</sup>, K. Barut<sup>1</sup>, S. Sahin<sup>1,\*</sup>, O. Kasapcopur<sup>1</sup>

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#### **PO162**

#### **CLINICAL SPECTRUM AND GENETIC PROFILE OF MEVALONATE KINASE DEFICIENCY: OUR EXPERIENCE FROM NORTH-WEST INDIA**

D. Suri<sup>1,\*</sup>, V. Joshi<sup>1</sup>, V. Pandiarajan<sup>1</sup>, R. K. Pilania<sup>1</sup>, S. Sharma<sup>1</sup>, M. Dhaliwal<sup>1</sup>, A. Gupta<sup>1</sup>, A. Rawat<sup>1</sup>, S. Singh<sup>1</sup>

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#### **PO164**

#### **PROLONGED URTICARIA AND FEVER IN A LIBYAN TODDLER BOY**

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#### **PO165**

#### **CLINICAL AND GENETIC PROFILE OF ROMANIAN PATIENTS DIAGNOSED WITH MEFV-ASSOCIATED AUTOINFLAMMATORY DISEASE: PRELIMINARY RESULTS OF THE RO\_FMF STUDY**

A. Stieber<sup>1,\*</sup>, A. Ţerban<sup>1</sup>, C. Dumitraşcu<sup>2</sup>, A. Bobircă<sup>2</sup>, A. Andronesi<sup>3</sup>, G. Ismail<sup>3</sup>, C. Jurcuț<sup>1</sup>

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**PO166****CLINICAL PRESENTATION AND GENETICS OF SUSPECTED MONOGENIC AUTOINFLAMMATORY DISEASES IN A SINGLE CENTRE AUSTRALIAN PAEDIATRIC RHEUMATOLOGY COHORT**G. Tiller<sup>1,2,\*</sup>, W. Renton<sup>1,3</sup>, E. Lynch<sup>4,5</sup>, J. Akikusa<sup>1</sup>, S. Mehr<sup>6</sup><sup>1</sup>Rheumatology, The Royal Children's Hospital, Melbourne, <sup>2</sup>Rheumatology, Monash Childrens Hospital, MelbourneMel,<sup>3</sup>Rheumatology, Monash Childrens Hospital, <sup>4</sup>Genetics, The Royal Children's Hospital, <sup>5</sup>Genetics, Murdoch Childrens Research Institute, <sup>6</sup>Immunology, The Royal Children's Hospital, Melbourne, Australia**PO167****PRESENTATION OF PATIENTS WITH NOD2 GENE VARIANTS - A CASE SERIES**H. Cioran<sup>1,2,\*</sup>, A. Stieber<sup>3</sup>, C.-D. Bucșa<sup>4</sup>, R. Vultură<sup>4</sup>, A. Bobirca<sup>5</sup>, C. Jurcut<sup>3</sup>, L. Muntean<sup>1,2</sup>, I. Felea<sup>1</sup>, S.-P. Simon<sup>1,2</sup>, I. Filipescu<sup>1,2</sup>, M. M. Tamas<sup>1,2</sup>, C. Pamfil<sup>1,2</sup>, S. Rednic<sup>1,2</sup>, L. Damian<sup>1</sup><sup>1</sup>Rheumatology, County Emergency Clinical Hospital, Centre for Rare Musculoskeletal Autoimmune and Autoinflammatory Diseases, <sup>2</sup>Rheumatology, Iuliu Hatieganu University of Medicine and Pharmacy, Cluj, <sup>3</sup>Internal Medicine, Dr. Carol Davila Central Military Emergency University Hospital, Bucharest, <sup>4</sup>Pharmacovigilance Research Center Department , Iuliu Hatieganu University of Medicine and Pharmacy, Cluj, <sup>5</sup>Internal Medicine and Rheumatology, Dr. Ion Cantacuzino Clinical Hospital, Bucharest, Romania**PO168****INFLAMMATORY BIOMARKER ANALYSIS CONFIRMS REDUCED DISEASE SEVERITY IN HETEROZYGOUS PATIENTS WITH FAMILIAL MEDITERRANEAN FEVER**I. Elhani<sup>1,\*</sup>, S. Backes<sup>2</sup>, T. Kallinich<sup>3</sup>, G. Amarian<sup>4</sup>, A. Belot<sup>5</sup>, R. Berendes<sup>6</sup>, T. Berger<sup>7</sup>, F. Dressler<sup>8</sup>, D. Foell<sup>9</sup>, S. Fühner<sup>2</sup>, A. Giese<sup>10</sup>, C. Hinze<sup>10</sup>, A. L. Hitzegrad<sup>2</sup>, G. Horneff<sup>11</sup>, A. Jansson<sup>12</sup>, J. Klotsche<sup>13</sup>, E. Lainka<sup>14</sup>, T. Niehues<sup>15</sup>, P. Oommen<sup>16</sup>, J.-P. Haas,<sup>17</sup>, C. Rietschel<sup>18</sup>, K. Theodoropoulos<sup>19</sup>, C. Vinit<sup>20</sup>, V. Hentgen<sup>21</sup>, H. Wittkowsk<sup>22</sup>, E. 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## PO170

### SUSPECTED NLRC4 IN LIBYAN PATIENTS WITH PERSISTENT SKIN RASH

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## PO171

### UNRAVELING INFLAMMASOMOPATHIES: INSIGHTS FROM A SINGLE -CENTER EXPERIENCE IN ARGENTINE

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## PO172

### MULTISYSTEM INFLAMMATORY SYNDROME IN CHILDREN IS ASSOCIATED WITH TGF-B-INDUCED EPSTEIN-BARR VIRUS REACTIVATION

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### PO173

#### LONG-TERM VIRAL PRESENCE IN MONOCYTES CORRELATES WITH DYSREGULATION OF INNATE IMMUNITY IN PATIENTS WITH MIS-C

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### PO174

#### UNDERSTANDING HOST-MICROBIOME RELATIONSHIPS IN THE PATHOPHYSIOLOGY OF BEHÇET SYNDROME: ANALYSIS OF SALIVARY CYTOKINES, SERUM CYTOKINES, AND TRYPTOPHAN METABOLITES CONCENTRATIONS IN PATIENTS FROM THE BEHCETBIOT STUDY.

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### PO175

#### DISSECTING THE HLH IMMUNE SYNAPSE (IS): CRITICAL ROLES FOR IS TERMINATION, CYTOKINE INTENSITY, AND TARGET CELL DEATH

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#### PO176

#### NATURAL KILLER CELL EXHAUSTION AND DYSFUNCTION AS A HALLMARK OF THE INFLAMMATION IN STILL'S DISEASE

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#### PO177

#### MULTI-OMIC STUDY IN PATIENTS WITH SITRAME SYNDROME IDENTIFIES DIFFERENCES IN SYSTEMIC IMMUNE RESPONSES

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#### PO178

#### EVALUATION OF TYPE I INTERFERON SIGNATURE AS A BIOMARKER FOR DISEASE ACTIVITY IN JUVENILE DERMATOMYOSITIS

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#### PO179

#### PLASMA PROTEOMIC PROFILES SEPARATE SURF PATIENTS FROM FMF AND PFAPA: PRELIMINARY DATA FROM THE PERSAIDS PROJECT.

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#### PO180

#### PERSONALIZED MEDICINE FOR SYSTEMIC AUTOINFLAMMATORY DISEASES: THE EUROPEAN MULTICENTER "PERSAIDS" PROJECT

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#### PO181

#### UNVEILING THE UNIQUE IMMUNOPATHOGENESIS OF CHRONIC RECURRENT MULTIFOCAL OSTEOMYELITIS THROUGH SINGLE-CELL RNA SEQUENCING

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#### PO182

#### BRIDGING THE GAP: DISPARITIES IN GENETIC TESTING AND TRAINING FOR AUTOINFLAMMATORY DISEASES BETWEEN EMERGING AND DEVELOPED COUNTRIES: A SURVEY ON 258 DOCTORS FROM 10 FRENCH SPEAKING COUNTRIES

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#### PO183

#### TO ASSESS HEALTH-RELATED QUALITY OF LIFE (HRQOL) IN PEDIATRIC PATIENTS WITH HEREDITARY AUTOINFLAMMATORY DISEASES (HAID) IN INDIA USING KIDSCREEN - 52 QUESTIONNAIRE - PILOT STUDY

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#### **PO184**

#### **INSIGHTS FROM A ONE-YEAR INTERNATIONAL MONTHLY QUIZ ON AA AMYLOIDOSIS CAUSES: ENGAGING 2,567 VOTERS ACROSS FRENCH-SPEAKING COUNTRIES**

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#### **PO185**

#### **CLINICAL FEATURES AND TREATMENT OUTCOMES IN VEXAS SYNDROME: A RETROSPECTIVE SINGLE CENTER EXPERIENCE**

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#### **PO186**

#### **IW-601, A FIRST-IN-CLASS CLINICAL-STAGE MONOCLONAL ANTIBODY TARGETING A NOVEL ADHESION CHECKPOINT ON MYELOID CELLS: POTENTIAL FOR TREATMENT OF AUTOINFLAMMATORY AND AUTOIMMUNE INDICATIONS**

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#### **PO187**

#### **KIKUCHI-FUJIMOTO DISEASE AS PRESENTING SIGN OF INBORN ERRORS OF IMMUNITY: A SINGLE CENTER EXPERIENCE FROM A COUNTRY AT LOW DISEASE PREVALENCE**

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#### **PO188**

#### **A QUANTITATIVE STANDARDIZED STRATEGY FOR CLINICAL APPLICATION OF TYPE I INTERFERON SIGNATURE**

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#### PO189

#### BRIDGING THE GAP: WHEN INSTAGRAM BECOMES A TOOL FOR PATIENT EDUCATION IN FAMILIAL MEDITERRANEAN FEVER

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#### PO190

#### SYSTEMIC AUTOINFLAMMATORY DISEASES IN UKRAINE: CHALLENGES, ACHIEVEMENTS, AND FUTURE PROSPECTS

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#### PO192

#### DATA FROM THE EUROFEVER REGISTRY FOR CENTRAL AND EASTERN EUROPEAN COUNTRIES: AN UPDATE

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#### PO193

#### AA AMYLOIDOSIS IN A FRENCH COHORT OF 312 PATIENTS: A COMPREHENSIVE STUDY ON ETIOLOGIES, DISEASE PROGRESSION AND MORTALITY

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#### PO194

**EMPOWERING PATIENTS WITH RARE AUTOINFLAMMATORY DISEASES: A PIONEER THERAPEUTIC EDUCATION PROGRAM FOR AMYLOIDOSIS, FMF, AND CAPS IN FRANCE**

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**PO195**

**AUTOIMMUNE-LYMPHOPROLIFERATIVE IMMUNODEFICIENCIES: A MONOCENTRIC EXPERIENCE**

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**PO196**

**EFFECTIVENESS OF IL1 INHIBITION WITH ANAKINRA IN ACUTE AND RECURRENT MYOCARDITIS IN CHILDREN**

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**PO197**

**CANADIAN AUTOINFLAMMATORY CASE ROUNDS: AN EDUCATION INITIATIVE TO FOSTER A COMMUNITY OF PRACTICE IN AUTOINFLAMMATION THROUGH CONNECTIVISM**

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**PO198**

**SERUM CYTOKINE PROFILES IN NEUTROPHILIC DERMATOSES**

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**PO199**

**MUTATION OF X-LINKED INHIBITOR OF APOPTOSIS (XIAP) IN A 14-YEAR-OLD GIRL: CHALLENGING MANAGEMENT OF INFLAMMATORY BOWEL DISEASE (IBD) IN XIAP-DEFICIENT FEMALE PATIENTS**

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