References:

- [1] Ben-Zvi I, Kukuy O, Giat E, Pras E, Feld O, Kivity Set al. Anakinra for Colchicine-Resistant Familial Mediterranean Fever: A Randomized, Double-Blind, Placebo-Controlled Trial. *Arthritis Rheumatol*. 2017;69:854-862.

Acknowledgments: We thank our patients and their families

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.6208

AB0979

MEVALONATE KINASE DEFICIENCY AT TREATMENT WITH CANAKINUMAB: RARE BLEATED CUTANEOUS FEATURE

R. Dos Santos Sobrín¹, B. Lopez-Montesinos¹, M. Martí Masanet¹, L. Lacruz Pérez¹, I. Calvo¹. ¹Hospital Universitario y Politécnico La Fe de Valencia, *Reumatología Pediátrica, Valencia, Spain*

Background: Mevalonate Kinase Deficiency (MKD) is an autosomal recessive autoinflammatory syndrome. Diagnostic criteria are based in clinical and genetic features (mutations in MVK gene) and its main treatment consists of blocking IL-1¹⁻⁵.

Objectives: To describe a case-series of a rare bleated cutaneous feature.

Methods: From January 2004 to September 2019, all cases diagnosed of MKD have been reviewed.

Results: 15 patients had MKD diagnosis (11 pathogenic mutations, homozygosis or double heterozygosis). Most common symptoms were oral and genital aphthous, abdominal pain, adenopathies and recurrent fever. Mean age of diagnosis was 10,67 years old (8,67 years later from the beginning of symptoms). Mean time of follow-up was 10 years. 3 patients developed this rare bleated cutaneous feature: suppurative hydrosadenitis. When the first injury appeared, all were at treatment with Canakinumab (mean time of treatment 4 years) and had 7,67 years of MKD course. This comorbidity began as repeating abscesses in folds with apocrine glands and hair follicles (armpits, inguinal, anal and genital folds). 2 patients continued Canakinumab and 1 switched to Adalimumab because of severity of cutaneous involvement.

Conclusion: This is the first case-series showing suppurative hydrosadenitis associated to MKD that has not been described in literature. Both theories have been found: as immune-mediated disease partnering this autoinflammatory syndrome (MKD) or as adverse event of treatment (anti-IL-1, less presumable).

References:

- [1] Van der Meer JW, Simon A. The challenge of autoinflammatory síndromes: with an emphasis on hyper-IgD syndrome. *Rheumatology* 2016;55:23-29.
[2] Arnal C, Modesto C. Síndromes febriles periódicos hereditarios: nuevas perspectivas sobre su conocimiento clínico y genético. 2003;30(2):45-48.
[3] Mulders-Manders C, Simon A. Hyper-IgD syndrome/mevalonate kinase deficiency: what is knew? *Semin Immunopathol* 2015;37:371-376.
[4] Cush, J. Autoinflammatory Syndromes. *Dermatol Clin* 2013;31(3):471-480.
[5] Moreira A, Torres B, Peruzzo J, et al. Skin symptoms as diagnostic clue for autoinflammatory diseases. *An Bras Dermatol* 2017;92(1):72-80.

Disclosure of Interests: Raquel Dos Santos Sobrín: None declared, B Lopez-Montesinos: None declared, Miguel Martí Masanet: None declared, Lucía Lacruz Pérez: None declared, Inmaculada Calvo Grant/research support from: Bristol-Myers Squibb, Clementia, GlaxoSmithKline, Hoffman-La Roche, Merck Sharpe & Dohme, Novartis, Pfizer, Sanofi, Speakers bureau: AbbVie, GlaxoSmithKline, Hoffman-La Roche, Novartis

DOI: 10.1136/annrheumdis-2020-eular.1357

AB0980

CASE-REVIEW ON FAMILIAL MEDITERRANEAN FEVER IN A SPECIALIZED CENTRE

R. Dos Santos Sobrín¹, M. Martí Masanet¹, B. Lopez-Montesinos¹, L. Lacruz Pérez¹, I. Calvo¹. ¹Hospital Universitario y Politécnico La Fe de Valencia, *Reumatología Pediátrica, Valencia, Spain*

Background: Familial Mediterranean Fever (FMF) is a genetic autoinflammatory disorder caused mostly by mutations in MEFV gene. Its inheritance is autosomal recessive and is the most frequent periodic fever syndrome. First-line treatment is based in colchicine use, so biologics (anti-IL-1) are reserved for refractory cases^{1,2}.

Objectives: To account for clinic and treatment features of patients with FMF in a specialized center as opposed to non-referent centers.

Methods: This study was developed in the Pediatric Rheumatology Service in Hospital Universitario y Politécnico La Fe de Valencia. Demographic, clinic and treatment data were collected from patients diagnosed of FMF since January 2004 to September 2019.

Results: 106 patients met last FMF criteria³. 55% had a pathogenic mutation in genetic analysis. 52% were female. Before 10 years old, 71% of patients had the diagnosis (51% before 4 years old). Arthralgia/myalgia (73%), periodic fever (62%) and abdominal pain (54%) were the most common symptoms. Systemic

Juvenile Idiopathic Arthritis (JIA, 6), other forms of JIA (9) and vasculitis (10) were the most prevalent comorbidities. When talking about treatment, 76,4% received Colchicine (60,5% with good response), 22,6% needed a classical disease modifying antirheumatic drug (mostly Methotrexate) and 22 patients got biologic treatment (73% anti-IL-1).

Conclusion: When analyzing this case-review, JIA has a strong association with our patients, so it could explain severe disease activity and more articular involvement. This could be an illustration to the higher use of Methotrexate. Also, the most relevant symptom was arthralgia while fever is the most frequent in literature. Likewise, age of diagnosis has been earlier than other case-series (this would be more frequent in other autoinflammatory syndromes, as literature relates)^{1,2,4}.

References:

- [1] Ozdogan H, Ugurlu S. Familial Mediterranean Fever. *Presse Med*. (2019).
[2] Ozen S, Demirkaya E, Erer B, et al. EULAR recommendations for the management of familial Mediterranean fever. *Ann Rheum Dis* 2016;75:644-651.
[3] Sag E, Demirel D, Demir S, et al. Performance of the new "Eurofever/PRINTO classification criteria" in FMF patients. *Semin Arthritis Rheum*. 2019;19:30369-5.
[4] Rozenbaum M, Rosner I. Severe outcome of juvenile idiopathic arthritis (JIA) associated with familial Mediterranean fever (FMF). *Clin Exp Rheumatol*. 2004;22:S75-8.

Disclosure of Interests: Raquel Dos Santos Sobrín: None declared, Miguel Martí Masanet: None declared, B Lopez-Montesinos: None declared, Lucía Lacruz Pérez: None declared, Inmaculada Calvo Grant/research support from: Bristol-Myers Squibb, Clementia, GlaxoSmithKline, Hoffman-La Roche, Merck Sharpe & Dohme, Novartis, Pfizer, Sanofi, Speakers bureau: AbbVie, GlaxoSmithKline, Hoffman-La Roche, Novartis

DOI: 10.1136/annrheumdis-2020-eular.1355

AB0981

VALIDATION OF THE CAREGIVERS QUESTIONNAIRE (IMPACT OF PEDIATRIC RHEUMATIC DISEASES ON CAREGIVERS MULTIASSESSMENT QUESTIONNAIRE) IN PEDIATRIC RHEUMATIC DISEASES

B. Fortuna¹, F. García-Rodríguez², N. Rubio², A. V. Villarreal², E. Faugier³, G. Reyes⁴, S. Mendieta⁵, I. Peláez-Ballestas⁶, S. Jimenez², S. Rosiles^{2,2}, J. Guadarrama³. ¹Universidad Autónoma de Nuevo León, *Reumatología Pediátrica, Monterrey, Mexico*; ²Universidad Autónoma de Nuevo León, *Monterrey, Mexico*; ³Hospital Infantil de Mexico Federico Gomez, *Ciudad de Mexico, Mexico*; ⁴Hospital de Especialidades de Chihuahua, *Chihuahua, Mexico*; ⁵Instituto de Seguridad Social del Estado de México y Municipios, *Estado de Mexico, Mexico*; ⁶Hospital General de Mexico Dr Eduardo Liceaga, *Ciudad de Mexico, Mexico*

Background: The CAREGIVERS questionnaire is used as a multidimension screening instrument for burden of caregivers, it allows to examine the impact of juvenile idiopathic arthritis (JIA) on several areas of the caregiver's life, looking for specific concerns and needs in which treatment could be required. This instrument has shown a good understanding among participants, reliability and consistency to measure the psychosocial and economic impact on primary caregivers of patients with JIA.

Objectives: The aim of this study is to validate CAREGIVERS questionnaire with the most common pediatric rheumatic diseases, such as JIA, systemic lupus erythematosus (SLE) and juvenile dermatomyositis (JDM).

Methods: The external validation phase of the constructed questionnaire will be carried out following Feinstein's clinimetric and psychometrics methodology, to measure the impact of the disease on the caregiver and create a risk profile for treatment detachment.

Results: A total of 200 questionnaires are applied to caregivers of pediatric patients with rheumatic diseases: 109 caregivers of patients with JIA, 28 caregivers of patients with DMJ and 63 caregivers of patients with SLE. When analyzing all the questions together, a general Cronbach's alpha of 0.6751 was obtained (Table 1).

Table 1. External validation of the Questionnaire

Dimension	Numero de ítems	Alfa de Cronbach
I. Impacto Emocional	6	0.5669
II. Impacto Social	3	0.47
IIIA. Impacto Financiero	4	0.6736
IIIB. Impacto Laboral	3	0.3151
IV. Impacto Familiar	5	0.4948
V. Impacto en la Relación entre Cuidador-paciente	1	NA
VI. Impacto en la relación de pareja	1	NA
VII. Impacto en la Religión/Creencias/Espiritualidad	1	NA
VIII. Impacto de las Redes Sociales	4	0.6642
Total	28	0.6751