Background: We present the first report of high-titer autoantibodies in *NLRP3*-associated autoinflammatory disease (*NLRP3*-AID). Because systemic autoinflammatory disease (SAID) is characterised by the lack of autoreactive T-cells or autoantibodies, we made a systemic review on the theme of autoantibody in SAID to clarify this phenomenon.

Objectives: We present the first report of high-titer autoantibodies *NLRP3*-AID, and discuss autoantibody in classical SAID.

Methods: We collected the clinical data of the patient with *NLRP3*-AID who had high-titer autoantibodies, and made a systemic review about autoantibody in SAID.

Results: A 38-year-old Chinese Han patient was definitely diagnosed as *NLRP3*-AID because of cold-triggered urticaria-like rash and fever, arthralgia, bilateral sensorineural deafness, chronic meningitis, high inflammatory marker and de novo *NLRP3* T348M variant. Figure 1 shows pedigree of the patient. Meanwhile, she had positive antinuclear antibody (ANA) with a nucleolar pattern of 1:160, positive anti-β2GPI antibody 54-68 AU/ml (normal range < 20 AU/ml) and positive lupus anticoagulant 1.24-1.29 (normal range ≤ 1.2). Literature review found that 13 articles reported autoantibodies in Familial Mediterranean fever (FMF), and there was no autoantibody reported in hyperimmunoglobulinemia D syndrome (HIDS), TNF receptor-associated periodic syndrome (TRAPS) and *NLRP3*-AID. The prevalence of ANA, anti-dsDNA, RF and anti-CCP in patients with FMF was similar to healthy controls.

Fig. 1. Pedigree of the patient. The black arrow indicates the proband. The asterisks indicate the individuals who had clinical manifestations and carry the *NLRP3* T348M variant.

Conclusion: Patients with *NLRP3*-AID can have high-titer ANA and APLs by accident. If patients with high-titer autoantibodies have characteristic manifestations of SAIDs instead of typical features of autoimmune diseases, we should make the final diagnosis through detailed investigation and genetic testing.

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.5670

AB1278

A CASE REPORT OF TOCILIZUMAB INDUCE RAPID REMISSION OF ADULT-ONSET STILL'S DISEASE WITH LIFE-THREATENING INTERSTITIAL LUNG DISEASE AND MACROPHAGE ACTIVATION SYNDROME

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Background: Adult-onset still's disease (AOSD) is a systemic inflammatory condition characterized by fevers, polyarthritides, skin rashes and increased leukocyte and neutrophil counts [1]. Sometimes life-threatening complications such as macrophage activation syndrome (MAS), disseminated intravascular coagulopathy (DIC) can be occurred without response to glucocorticoid and disease-modifying antirheumatic drugs (DMARDs).

Objectives: To describe an AOSD patient with life-threatening interstitial lung disease (ILD) and MAS response to tocilizumab.

Methods: This is a case report at our medical practice.

Results: A 43-year-old man was hospitalized for a 2-week history of fever accompanying skin rashes, polyarthritides, sore throat, leukocytosis (the maximum leukocyte count $37.25 \times 10^9/L$ with 89.9% neutrophils), elevated levels of C-reactive protein (190.9mg/L) and ferritin level was very high ($>15000ng/ml$). Several antibiotics were consequently used, but fever was on going. Chest CT displayed diffuse interstitial lung disease. Infectious diseases and lymphoma were excluded and AOSD with ILD was diagnosed. However, MAS occurred the 20 days later after the pulse of intravenous MP (MP 500mg×3days) following MP 40mg/day, which cannot stop the situation deteriorating. The minimum total white blood cells, hemoglobin and platelets went down to $0.25 \times 10^9/l$, 53g/l and $18 \times 10^9/l$. Finally, tocilizumab rescued this patient from desperation.

Conclusion: Tocilizumab was effective in AOSD with life-threatening ILD and MAS.

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.2529

HPR Measuring health (development and measurement properties of PROs, tests, devices)

AB1279-HPR

A DESCRIPTIVE STUDY RELATED TO THE ADHERENCE BEFORE AND AFTER ENROLLING IN A MULTIDISCIPLINARY EDUCATIONAL PROGRAM

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Background: Rheumatoid arthritis (RA) is an inflammatory, chronic disease. It leads to deformity and destruction of joints through the erosion of cartilage and bone. Patients with RA report to suffer symptoms in hands, joints, swelling, loss of motion, muscle weakness among others. (1). Centers of excellence in RA have proposed a multidisciplinary model of care with an initial diagnosis, treatment prescription and follow-up with a rheumatologist, periodic consultations with a physiatrist, psychologist, physiotherapist, occupational therapy, nutrition and a patient focused program (2). With this model of care, the patient is seen as a whole, and the expectation is to achieve the best results in the management of RA. However, if the patient does adhere the model becomes ineffective.

Objectives: The aim of this to report the attendance to a multidisciplinary model of care for patients with RA that attend to a specialized center in Colombia, before and after enrolling in an educational program.

Methods: We performed a descriptive study. Patients enrolled our educational program in July 2019. In our institution patients are followed-up under T2T standards and a multidisciplinary approach, as part of our model of care they have periodic consultations with a rheumatology, physiatrist, psychologist, physiotherapist, occupational therapy and nutrition. We collected sociodemographic data, DAS28, and compare the attendance to each specialty at the beginning and at 6-month follow-up. Descriptive epidemiology was done, we calculated means, and standard deviations for continuous variables and categorical variables were presented as rates. We compared disease activity and adherence at the beginning of the program and after six months of attendance.

Results: We included 229 patients; mean age was 59 years ± 10 ; 93% were female. At the beginning of our program, mean DAS28 was 2.57 ± 1.19 , from all patients 65% were at remission, 11% at low disease activity 19% at moderate disease activity and, 5% at severe disease activity. Regarding adherence to our model, the medical specialty with the highest attendance was rheumatology (30%) followed by, physical therapy (16%) physiatrist consultation (15%) psychology (13%) and, occupational therapy (11%); the specialty with the lowest attendance was nutrition (8%). After six months of attendance to the educational program, we found an increasing number of patients in remission 67%, low disease activity 15%, moderate disease activity 18%, we did not have patients with severe DAS28. Regarding the medical specialties, we found a 3% rise in the attendance to the nutrition consultation and psychology consultation. We did not find statistical association between disease activity and adherence to the model.

Conclusion: These results are a clear example of how an educational program is capable of increasing awareness and improving the clinical outcomes and adherence to a multidisciplinary model for approaching RA. As other studies have shown (3), patient education interventions improve adherence to medication and to attendance to health care specialists.

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Acknowledgments: This project has been funded by a collaboration between the Ministry of Science, Technology and Innovation COLCIENCIAS (contract 746-2018), the Fundación Universitaria de Ciencias de la Salud and Biomab - Center for Rheumatoid Arthritis

Disclosure of Interests: Diana Buitrago-García: None declared, Fernando Rodríguez: None declared, GUILLERMO SÁNCHEZ: None declared, Pedro Santos-Moreno Grant/research support from: I have received research grants from Abbvie, Biopas-UCB, Janssen, Novartis, Pfizer., Speakers bureau: I have been a speaker for Abbvie, Biopas-UCB, Janssen, Lilly, Novartis, Pfizer, Roche, Sanofi.