is an underrecognized adverse effect that can potentially cause significant morbidity and mortality.

Objectives: To study incidence, presentation, and characteristics of HCQ-induced AVB in IMID.

Methods: Open-label single center study of 293 patients with IMID treated with HCQ for at least 3 months. Electrocardiograms were analyzed at baseline and during HCQ treatment. In addition, a comparative study between patients with and without AVB was conducted.

Results: We studied 293 patients (270 women/ 23 men; mean age 59.7±14.7 years). Underlying IMID were Systemic Lupus Erythematosus (SLE) (n=109, 40.6%); undifferentiated connective-tissue disease (n=70, 23.9%), Sjögren's syndrome (SS) (n=70, 23.9%), antiphospholipid syndrome (n=31, 10.6%) and other IMID (n=13, 4.4%). HCQ was used for 4.1±3.5 years.

After 11.8±8.9 years of follow-up (HCQ mean cumulative dose: 979.7±272.1 g). AVB was observed in 19 out of 293 (6.5%) patients: 16 (84.2%) were first-degree AVB and 3 (15.8%) complete AVB. 4 (21.2%) patients with AVB were treated with a permanent pacemaker.

Presentation of atrioventricular block according to hydroxychloroquine incidence rate and treatment duration is shown in Figure 1. Most of AVB happened in the first 40 months, regardless of HCQ cumulative dose.

Figure 1. Presentation of atrioventricular block according to hydroxychloroquine incidence rate and treatment duration.

A comparative study between patients with and without AVB was performed (Table 1). Patients with AVB were older ($p=0.014$) and had a higher incidence of diabetes mellitus ($p=0.033$). HCQ cumulative dose and duration of IMID were similar in both groups ($p>0.05$).

Other HCQ adverse effects were retinopathy (n=16, 5.4%), gastrointestinal alterations (n=14, 4.8%), cutaneous alterations (n=14, 4.8%), allergic reactions (n=4, 1.4%) and myopathy (n=1, 0.3%).

Conclusion: AVB was observed in 6.5% of patients with IMID treated with HCQ. Most AVB were first-degree AVB. HCQ increased the risk of developing an AVB in patients with IMID regardless of HCQ cumulative dose or underlying disease duration. Older patients with diabetes mellitus had a higher risk of developing an AVB.

Table 1. Comparative study between patients with and without AVB.

	with AVB n= 19	without AVB n= 274	p
General features at HCQ onset			
Age, years mean±SD	57.4±14.5	46.4±16.8	0.014
Female, n (%)	17(89.5)	239(87.4)	0.877
Hypertension, n (%)	14 (73.7)	149 (54.4)	0.118
Dyslipemia, n (%)	11 (58.9)	117 (42.7)	0.265
Diabetes Mellitus, n (%)	5 (26.3)	24 (8.7)	0.033
Renal impairment, n (%)	5 (26.3)	29 (10.7)	0.073
Ischemic cardiopathy, n (%)	2 (10.5)	11 (3.9)	0.228
CRP, mg/dL median [IQR]	0.3 [0.1-0.5]	0.4 [0.3-0.8]	0.343
ESR, mm/h median [IQR]	17.0 [15.0-22.3]	13.0 [7.0-29.5]	0.206
Data at last visit (11.8±8.9 years of follow-up)			
IMID duration, years mean±SD	11.8±8.9	14.3±10.3	0.288
# Use of other cardiotoxic drugs, n (%)	2 (10.5)	11 (4.0)	0.723
HCQ cumulative dose, g mean±SD	813.2±206.0	996.3±266.9	0.527

(beta blockers, calcium channel blockers, tricyclic drugs, other AVB inducers)

Disclosure of Interests: Alba Herrero-Morant: None declared, Adrián Margarida-de Castro: None declared, Raquel Pérez-Barquín: None declared, Jon Zubiaur-Zamacola: None declared, Miguel A. González-Gay Speakers bureau: AbbVie, Pfizer, Roche, Sanofi, Lilly, Celgene and MSD, Grant/research support from: AbbVie, MSD, Jansen and Roche, Ricardo Blanco Speakers bureau: AbbVie, Pfizer, Roche, Bristol-Myers, Janssen, Sanofi, Lilly and MSD, Grant/research support from: AbbVie, MSD, and Roche

DOI: 10.1136/annrheumdis-2021-eular.3295

POS1371 BIOLOGICAL THERAPY IN REFRACTORY NEUROBEHÇET'S DISEASE. MULTICENTER STUDY OF 42 PATIENTS

A. Herrero-Morant¹, J. L. Martín-Varillas², S. Castañeda³, I. González-Mazón¹, O. Maiz⁴, A. Blanco⁴, J. Sánchez⁵, N. Ortego⁶, E. Raya⁶, A. Olive⁷, A. Brandy-García⁷, Á. Prior-Español⁷, C. Moriano⁸, E. Díez Álvarez⁸, R. Melero⁹, J. Graña¹⁰, Á. Seijas-López¹⁰, A. Urruticoechea-Arana¹¹, A. Ramos Calvo¹², C. Delgado Beltrán¹³, M. Loreda Martínez¹³, E. Salgado-Pérez¹⁴, F. Sivera¹⁵, I. Torre-Salaberri¹⁶, J. Narváez¹⁷, J. L. Andréu Sánchez¹⁸, O. Martínez González¹⁹, R. Gómez de la Torre²⁰, S. Fernández²¹, S. Romero-Yuste²², G. Espinosa²³, M. Á. González-Gay¹, R. Blanco¹. ¹Hospital Universitario Marqués de Valdecilla, Rheumatology, Santander, Spain; ²Hospital de Sierrallana, Rheumatology, Torrelavega, Spain; ³Hospital Universitario La Princesa, Rheumatology, Madrid, Spain; ⁴Hospital Universitario de Donostia, Rheumatology and Ophthalmology, San Sebastián, Spain; ⁵Hospital 12 de octubre, Rheumatology, Madrid, Spain; ⁶Hospital San Cecilio, Rheumatology and Internal Medicine, Granada, Spain; ⁷Hospital Universitario Germans Trias i Pujol, Rheumatology, Barcelona, Spain; ⁸Hospital de León, Rheumatology, León, Spain; ⁹Complejo Hospitalario de Vigo, Rheumatology, Vigo, Spain; ¹⁰Hospital Universitario de A Coruña, Rheumatology, A Coruña, Spain; ¹¹Hospital Can Misses, Rheumatology, Ibiza, Spain; ¹²Complejo Hospitalario de Soria, Rheumatology, Soria, Spain; ¹³Hospital Clínico Lozano Blesa, Rheumatology, Zaragoza, Spain; ¹⁴Complejo Hospitalario Universitario de Ourense, Rheumatology, Ourense, Spain; ¹⁵Hospital General Universitario de Elda, Rheumatology, Elda, Spain; ¹⁶Hospital de Basurto, Rheumatology, Bilbao, Spain; ¹⁷Hospital Bellvitge, Rheumatology, Barcelona, Spain; ¹⁸Hospital Universitario Puerta de Hierro, Rheumatology, Madrid, Spain; ¹⁹Hospital Clínico Universitario de Salamanca, Rheumatology, Salamanca, Spain; ²⁰Hospital Universitario Central de Asturias, Internal Medicine, Oviedo, Spain; ²¹Hospital Universitario de Cabueñes, Rheumatology, Gijón, Spain; ²²Complejo Hospitalario Universitario de Pontevedra, Rheumatology, Pontevedra, Spain; ²³Hospital Clínic de Barcelona, Internal Medicine, Barcelona, Spain

Background: Neuro-Behçet's disease (NBD) is a severe complication of Behçet's disease (BD). Despite well-established therapies with glucocorticoids and conventional immunosuppressants (cIS), a significant proportion of patients are refractory.

Objectives: To assess efficacy and safety of biologic therapy (BT) in NBD refractory to glucocorticoids and at least one cIS.

Methods: Open-label multicenter study of refractory NBD from 23 different referral Spanish Hospitals. Main outcome was neurological response. Secondly, analytical efficacy was measured by Erythrocyte Sedimentation Rate (ESR), C-Reactive Protein (CRP) and Hemoglobin (Hb) at baseline, 6 months, 1 year and 2 years.

Results: We studied 42 patients (21 women/ 21 men; mean age 40.4±10.8 years). HLA B51 was positive in 15 out of 37 (40.5%) patients tested. Non-neurological manifestations were oral ulcers (n=41, 97.6%), genital ulcers (n=31, 73.8%), skin lesions (n=28, 66.7%), arthralgia (n=27, 64.3%), uveitis (n=21, 50.0%), arthritis (n=9, 21.4%), venous thrombosis (n=9, 21.4%) and arterial thrombosis (n=4, 9.5%). The underlying neurologic manifestation were parenchymal (n=34, 81.0%) and non-parenchymal (n=17, 40.5%) involvement (Table 1). The first BT used was infliximab (n=20), adalimumab (n=13), golimumab (n=3), tocilizumab (n=3) and etanercept (n=2).

After 58.2±51.4 months since initiation of BT, neurological response was complete (n=27; 64.3%), or partial (n=11, 26.1%) (Figure 1). Only 4 (9.5%) patients did not respond. After 6 months of BT, ESR improved from 31.5±25.6 to 15.3±11.9 mm/h ($p=0.005$), CRP from 1.4 [0.2-12.8] to 0.3[0.1-3] mg/dL ($p=0.002$) and Hb from 13.1±1.6 to 13.8±1.3 g/dL ($p=0.005$).

Table 1. Neurologic manifestation of 42 patients with refractory neurobehçet's disease treated with biologic therapy.

Parenchymal subtype, n (%)	34 (81.0)
-Hemiparesis	8 (19.1)
-Polineuropathy	8 (19.1)
-Encephalopathy	6 (14.3)
-Cognitive impairments	4 (9.5)
-Optic neuropathy	4 (9.5)
-Ophthalmoparesis	4 (9.5)
-Other cranial nerve involvement	3 (7.1)
-Hemihypoesthesia	3 (7.1)
-Cerebellar dysphasia	1 (2.4)
-Cerebellar involvement	1 (2.4)
-Non-steroidal psychosis	1 (2.4)
Non-parenchymal subtype, n (%)	17 (40.5)
-Aseptic meningitis	12 (28.6)
-Thrombosis	4 (9.5)
-Intracranial hypertension	1 (2.4)

Figure 1. Neurological clinical response to biological therapy.

Primary failure was observed in 16 (38.1%) patients due to inefficacy (n=11, 68.8%) or adverse effects (n=5, 31.3%). Similarly, causes of secondary failure (n=6, 14.3%) were inefficacy (n=5, 83.3%) and adverse effects (n=1, 16.7%). No serious adverse effects were observed.

Conclusion: BT, especially monoclonal anti-TNF drugs, seems to be effective and safe in refractory NBD.

Disclosure of Interests: Alba Herrero-Morant: None declared, José Luis Martín-Varillas Grant/research support from: AbbVie, Pfizer, Lilly, Janssen, and Celgene, Santos Castañeda: None declared, Iñigo González-Mazón: None declared, Olga Maiz: None declared, Ana Blanco Speakers bureau: AbbVie, Julio Sánchez: None declared, Norberto Ortego: None declared, Enrique Raya Speakers bureau: MSD, Grant/research support from: AbbVie, Alejandro Olive: None declared, Anahy Brandy-García: None declared, Águeda Prior-Español: None declared, Clara Moriano: None declared, Elvira Díez Álvarez: None declared, Rafael Melero: None declared, Jenaro Graña: None declared, Álvaro Seijas-López: None declared, ANA URRUTICOECHA-ARANA: None declared, Angel Ramos Calvo: None declared, Concepción Delgado Beltrán: None declared, Marta Loredo Martínez: None declared, Eva Salgado-Pérez: None declared, Francisca Sivera: None declared, Ignacio Torre-Salaberri: None declared, J. Narváez Speakers bureau: Bristol-Myers Squibb, José Luis Andréu Sánchez: None declared, Olga Martínez González: None declared, Ricardo Gómez de la Torre: None declared, Sabela Fernández: None declared, Susana Romero-Yuste: None declared, Gerard Espinosa: None declared, Miguel Á. González-Gay Speakers bureau: AbbVie, Pfizer, Roche, Sanofi, Lilly, Celgene and MSD, Grant/research support from: AbbVie, MSD, Jansen and Roche, Ricardo Blanco Speakers bureau: AbbVie, Pfizer, Roche, Bristol-Myers, Janssen, Sanofi, Lilly and MSD, Grant/research support from: AbbVie, MSD, and Roche

DOI: 10.1136/annrheumdis-2021-eular.3407

POS1372

EVALUATION OF E148Q AND CONCOMITANT AA AMYLOIDOSIS SECONDARY TO FAMILIAL MEDITERRANEAN FEVER AFTER ADJUSTED CLINICAL-DEMOGRAPHIC CHARACTERISTICS

Z. S. Arici¹, M. Romano², D. Piskin³, F. Guzel⁴, M. Yilmaz⁵, E. Demirkaya².

¹Sanliurfa Training and Research Hospital, Paediatric Rheumatology, Sanliurfa, Turkey; ²Schulich School of Medicine & Dentistry, University of Western Ontario, Paediatric Rheumatology, London, Canada; ³Lawson Health Research Institute,

Pediatrics, London, Canada; ⁴Gentera Biotechnology, Biotechnology, Istanbul, Turkey; ⁵Epigenetic Health Solutions, Unit of Nephrology, Ankara, Turkey

Background: Amyloid A (AA) amyloidosis, previously known as secondary or reactive amyloidosis, is a long-recognized severe complication of some chronic inflammatory diseases. The pathogenesis and risk factors for amyloidosis in Familial Mediterranean Fever (FMF) remain partially understood (1). The development of AA amyloidosis is dependent on ethnicity and country of residence (2). In the pre-colchicine era, renal AA-amyloidosis was largely reported patients of Turkish (67%) and Sephardic Jewish ancestry (26.5%) (2,3). Currently it's well known that the MEFV M694V variant associated with high risk of amyloidosis however, mutations on exon 2, specifically E148Q variant remained controversial.

Objectives: To evaluate the E148Q mutation variant and concomitant AA Amyloidosis secondary to FMF after adjusted clinical-demographic characteristics.

Methods: Patients were recruited from the renal unit at Epigenetic Health Center outpatient clinic in Turkey between September 2003 and February 2020. Patients who had biopsy confirmed FMF related AA amyloidosis were included the study. Tel-Hashomer criteria were applied in the diagnosis of FMF. The clinical characteristics of FMF patients and medication history were recorded by the physician at the time of registry entry. All patients had detailed baseline assessment of clinical features, renal functions, genetic testing, histopathological diagnosis of amyloidosis, and treatment received. We performed multiple comparisons according to the age of diagnosis, demographic features, disease phenotype, allele frequency, type of mutation and mortality. Statistical analysis was performed with Statistical Package of Social Science (SPSS) for Windows, version 15.0 (SPSS Inc, Chicago, IL).

Results: Our registry consists of 195 patients with a diagnosis of AA amyloidosis. Complete information on 169 patients (lost to follow up, n=26) were included. The median age was 36 (19-49) years; male/female ratio was 1.6 (104/65). The median follow-up duration was 15.0 years (4-17 years). There were 101 patients diagnosed with FMF <18 years of age and 68 patients diagnosed ≥18 years of age. All participants developed renal amyloidosis before the age of 32 years. Family history of FMF was documented in 56 patients (33.1%) and family history of amyloidosis was present in 41 patients (24.3%). The three most common clinical symptoms were fever (84.6%), abdominal pain (71.6%) and arthritis (66.9%). During the follow-up, 5 patients started dialysis treatment and 9 patients had kidney transplantation. The most common allele frequency across patients was M694V (60.6%), E148Q (21.4%) and M680I (10.3%). The most frequent mutations were M694V/M694V (63.3%), M694V/E148Q (20.8%) and E148Q/E148Q (15.8%). During the follow up period, 15 patients (10 male, 5 female) died. In those that died, the mutations in 14 had M694V/M694V and one demonstrated E148Q/E148Q.

Conclusion: Patients with FMF related AA amyloidosis have an increased risk for mortality. This study confirmed the association between M694V and FMF-associated AA amyloidosis, which has been reported in many studies. Close clinical follow-up and further evaluation of patients with the E148Q mutation is warranted specifically if residing in FMF endemic areas. The possible relationship between E148Q and AA amyloidosis need to be confirmed in other cohorts.

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2021-eular.3420

POS1373

DEFINING EAR CHONDROITIS: DATA FROM 685 PATIENTS WITH RELAPSING POLYCHONDROITIS

M. Ferrada¹, N. Costedoat-Chalumeau², G. Moulis³, N. Linn⁴, E. Rose¹, P. Grayson¹. ¹NIH, NIAMS, Bethesda, United States of America; ²APHP, Université de Paris, Paris, France; ³Toulouse University Hospital, Department of Internal Medicine, Toulouse, France; ⁴Relapsing Polychondritis Foundation, Relapsing Polychondritis Foundation, New York, United States of America

Background: Ear chondritis is often considered the pathognomonic feature of relapsing polychondritis (RP). Although painful redness and swelling of the pinna and a resultant cauliflower ear are universally recognized as chondritis, the complete spectrum of symptoms associated with ear chondritis have not been well described.