included use of opioids. The prescription patterns were similar to those observed in other German cohort studies (Albrecht et al. Lupus Sci Med. 2014;**1**:e000059 and 2021;8:e000526).

## PO.6.141 EFFICACY OF MYCOPHENOLATE MOFETIL (MMF) IN PEDIATRIC PATIENTS WITH DEFINED AND UNDEFINED TYPE I INTERFERONOPATHIES

[1]MF Natale*, [2]C Udina, [3]CGL Raffaele, [1]V Messia, [1]C Celani, [1]C Bracaglia, [1]S Federici, [1]G Prencipe, [1]F De Benedetti, [1]A Insalaco. *[1]Bambino Gesù Children's Hospital ~ Rome ~ Italy; [2]Ospedale Santa Maria degli Angeli-Pordenone ~ Italy; [3]Ospedale San Benedetto-Alatri ~ Frosinone ~ Italy*

10.1136/lupus-2022-elm2022.161

**Purpose** Interferonopathies are conditions characterized by excessive production of type 1 interferon. Several diseases, autoinflammatory or autoimmune, are classified as type1 interferonopathies such as Aicardi-Goutieres, Sting associated vasculopathy with onset in Infancy, monogenic Systemic lupus erythematosus (SLE) and Dermatomyositis. The aim of this study is to evaluate the role of MMF in modulating the activation of the interferon pathway describing its clinical and laboratory effects in a cohort of patients with defined and undefined interferonopathy

**Methods** We included patients aged 0–18 years with defined (genetically confirmed) and undefined interferonopathy (evocative clinical picture, interferon signature (IS)> 2 in at least 3 samples 3 months apart, negative genetic test) followed at Bambino Gesù Children's Hospital treated with MMF. For each patient demographic, clinical, laboratory parameters and IS were collected every 3 months starting from the year before the MMF treatment until the date of the last follow up. In vitro the effect of MMF on the IFN-pathway was evaluated by adding MMF to peripheral mononuclear cells of the patients.

**Results** Nine patients were enrolled, 4 with defined and 5 with undefined interferonopathy. When therapy was started, 78% of patients had recurrent febrile episodes, 57% polyarthritis, 67% skin and/or neurological involvement. A pathological brain MRI was found in 44% of patients. Persistent anemia, lymphopenia and autoantibody positivity were detected in 44% of patients and increased ESR in 67%. All patients had a positive IS(median 46.9; IQR 23.5–133.8). Three months after the beginning of MMF we observed a resolution of febrile episodes and skin manifestations in 86% of cases, remission of joint involvement in all patients, improvement of neurological symptoms in 1 patient, resolution of the anemia and normalization of the ESR in 50% and 66% respectively. During MMF therapy, as reported in patients with SLE, ANA titers decline or normalized in 75% of patients and anti dsDNA in 66% of patients. Cerebral MRI improved in patient with radiological alterations. MMF therapy allowed to completely withdraw glucocorticoid therapy in 4 patients and to reduce the dosage in other 3. No significant reduction in IS was detected during a median follow-up of 36 months (IQR 25–45). Nevertheless these data contrasted with our ex vivo experiments in which the incubation with MMF of peripheral mononuclear cells of our patients (and of patients with other interferonopathies including SLE and dermatomyositis) showed a significant reduction in the INF-a pathway activation after cells stimulation

**Conclusions** This is the first study evaluating the effects of MMF in a cohort of patients with type I interferonopathy. The results, although preliminary, seem to suggest a role of the drug in improving clinical, laboratory and radiological findings of our patients, allowing also a significant glucocorticoid sparing. These data, if confirmed in larger and prospective studies, should encourage the use of MMF especially for those patients in which standard treatment with JAK inhibitor could be contraindicated or ineffective

# Friday 07 October 2022 from 13:00 to 14:10

# PO.7 E- poster session 7: patient views and reported outcomes, registries and cohorts

## PO.7.142 HEALTH-RELATED QUALITY OF LIFE ACROSS THE SPECTRUM OF CONNECTIVE TISSUE DISEASES: A LATENT PROFILE ANALYSIS

[1]S Dyball*, [2]J Reynolds, [1]IN Bruce, [3]B Parker. *[1]University of Manchester ~ UK; [2]Rheumatology Research Group, Institute of Inflammation and Ageing, College of Medical and Dental Sciences, University of Birmingham ~ UK; [3]The Kellgren Centre for Rheumatology, Manchester University NHS Foundation Trust ~ Manchester ~ UK*

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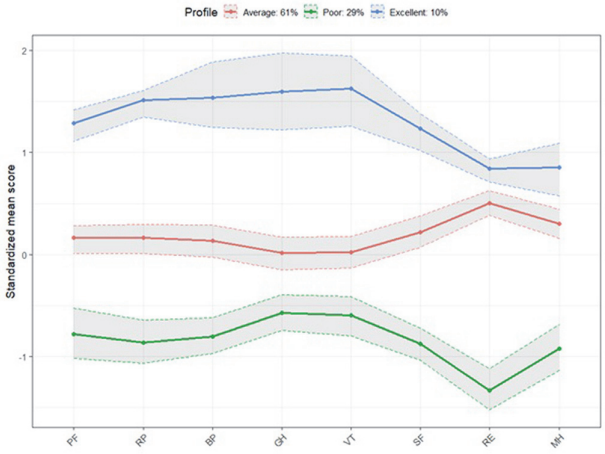
**Purpose** Poor health-related quality of life (HR-QoL) is well recognised within patients with connective tissue diseases (CTD). We hypothesised that subgroups of patients across the spectrum of CTD experience different HR-QoL patterns, and aimed to determine patient-level characteristics associated with subgroup membership.

**Methods** The medical outcomes short-form 36 (SF-36) questionnaire was used, and the eight continuous domains of the SF-36 questionnaire were derived which range from 0 to 100,




**Abstract PO.7.142 Figure 1** Latent profile analysis using eight domains of the SF-36 across connective tissue diseases (n=309). PF, physical function; RP, role physical; BP, body pain; GH, general health; VT, vitality; SF, social function; RE, role emotional; MH, mental health