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

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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

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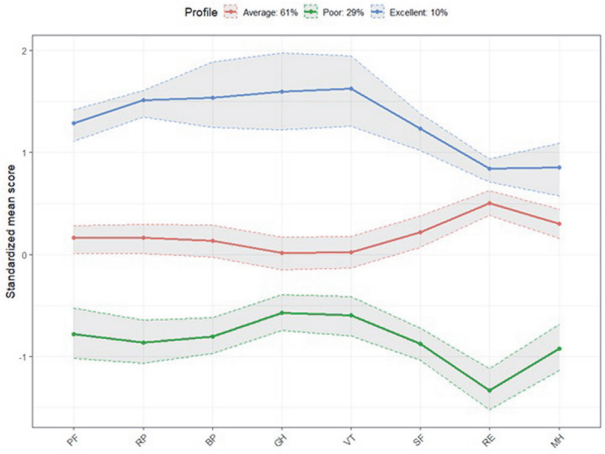
10.1136/lupus-2022-elm2022.162

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

with higher scores reflecting better HR-QoL. We used the 'mclust 5.4.9' model-based clustering package in R V4.0.4 to identify latent profiles (LP) of patients who experienced distinct HR-QoL patterns. Variances were equated and covariances fixed to zero. Missing values were imputed using methodology suggested by the SF-36 manual. Number of comorbidities included osteoarthritis, chronic kidney disease, ischaemic heart disease, cancer, diabetes, reflux disease, chronic liver disease, obstructive airway disease or interstitial lung disease, thyroid disease, stroke, vitiligo, Addison's disease and atrial fibrillation. Multivariable ordinal logistic regression was used to determine patient-level characteristics associated with membership in the HR-QoL subgroups. Models were adjusted for ethnicity, sicca, fibromyalgia, and number of comorbidities.

Results 309 patients with a variety of CTDs recruited into LEAP completed the SF-36 questionnaire, (280 [90.6%] women, mean [SD] age 48.9 [12.9] years). There were 115 (37.2%) patients with lupus, 72 (23.3%) undifferentiated CTD, 56 (18.1%) primary Sjögren's syndrome and 66 (21.4%) patients with systemic sclerosis, idiopathic inflammatory myopathy or an overlap syndrome (SSc-IIM). Three LP were identified with poor (n=89), average (n=190) and excellent (n=30) HR-QoL (figure 1). In multivariable models, LP were not associated with diagnostic grouping (Sjogren's: OR 0.97 [95% CI 0.46–2.06]; undifferentiated CTD: OR 1.05 [95% CI 0.57–1.92]; SSc-IIM: OR 0.89 [95% CI 0.48–1.68]). Black ethnicity (OR 0.26 [95% CI 0.10–0.70]) or Asian ethnicity (OR 0.41 [95% CI 0.21–0.81]), concomitant fibromyalgia (OR 0.44 [95% CI 0.22–0.86]), sicca phenomenon (OR 0.56 [95% CI 0.35–0.91]) and multi-morbidity (OR 0.80 [95% CI 0.63–1.00]) were associated with membership of a lower HR-QoL LP.

Conclusion CTD patients can be clustered into distinct HR-QoL subgroups agnostic of their clinical diagnosis which may have meaningful implications in practice for stratifying therapy. This study highlights that sicca syndrome, ethnicity, fibromyalgia and multi-morbidity are key areas to focus on in improving HR-QoL in patients with a CTD, irrespective of clinical diagnosis or antibody profile.

PO.7.143 VALUE OF FATIGUE: PATIENTS VS. PHYSICIANS – TIME TRADE-OFF APPROACH RESULTS FROM A PATIENT SURVEY

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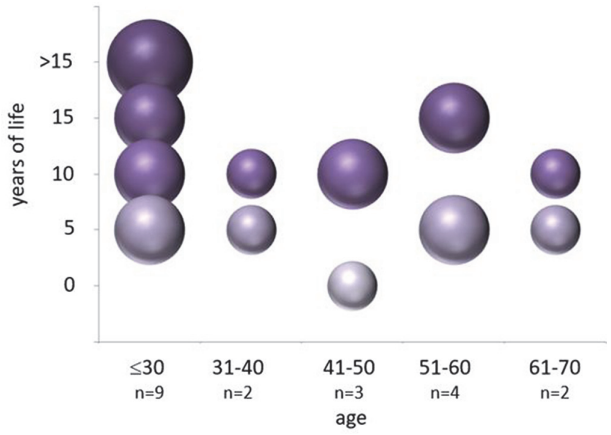
10.1136/lupus-2022-elm2022.163

Purpose Fatigue is a very common symptom in SLE patients and often held responsible for their reduced quality of life. With this survey, we wanted to examine the degree of suffering in SLE patients with fatigue and determine, whether they would trade in years in order to live without fatigue or if they would trade in fatigue for other disease manifestations (skin, joints or kidney). We also looked at the number of patients, who were regularly asked about fatigue by their treating physician.

Methods Our patient survey took place during a meeting with patients from the German SLE self help community on World Lupus Day, May 10th 2019 in Düsseldorf, Germany. We used Edivote®, which is an anonymous, audience response system working with PowerPoint® for our survey. Patients' demographics (age, gender and disease duration) were documented separately. We asked if patients had been questioned by their treating physicians about fatigue (1: always, 2: sometimes, 3: never), how many years of life they would sacrifice in order to live without fatigue (0, 5, 10, 15, >15 years, respectively) and who would trade in fatigue for another disease manifestation (skin, joints or kidney).

Results 26 patients (25 female) agreed to take part in our survey and completely or in part responded to the questions asked. Most patients were ≤50 years old (n=16, 61.5%). Most patients had a disease duration of >5 years (6–10 years: n=5; >10 years: n=16). Six patients were regularly asked about fatigue, four patients were sometimes asked about fatigue and eight patients had never been asked about fatigue. The results of the time trade-off question are shown in figure 1, sorted by patients' age. In our cohort, five patients were willing to trade in fatigue for a skin manifestation (consistent redness on exposed skin). One patient was willing to trade in fatigue for a kidney manifestation (50% reduction of kidney function, accompanied by edema and fluid restriction). Twelve patients did not want to trade in fatigue for one of the mentioned disease manifestations.



Abstract PO.7.143 Figure 1 Results from our time trade-off survey, sorted by patients, age; n=20, absolute numbers; (Size of circles represents 1 patient [small], 2 patients [medium] and 3 patients [large])

Conclusion 42% of responding patients in our cohort (n=19) had never been asked about fatigue by their treating physicians. To our surprise, almost all patients were willing to trade in at least 5 years and more for a life without fatigue. This result underlines patients' degree of suffering and should raise more awareness for SLE patients living with fatigue. Our results clearly indicate the discordant assessment of the importance of fatigue by patients and physicians. Although our cohort is small and relatively young, even some patients between the age of 61 and 70 were willing to trade in up to 10 years for a life without fatigue. Future research must be conducted to better understand fatigue in SLE patients and to develop appropriate treatment strategies. Our results with respect to time trade-offs should be validated in a larger cohort.