groups in any of the included variables. Most patients reported mild pain (NRS median 3) and moderate disability. Thirty-six had additional affection of interphalangeal joints. At follow-up, the control group showed no improvements, while the intervention group showed significant improvement in all pain and functional measures, except flexion deficit. When adjusting for baseline value and time to follow-up, there were significant differences between the two groups at follow-up for pain at rest, pain following grip, self-reported function (MAP-Hand and Quick-DASH) and grip strength, with the intervention group showing greater improvement ($p < 0.01$).

Conclusion: Occupational therapy had significant positive short-term effects on pain and function in patients referred to surgical consultation for CMC1 OA.

Disclosure of Interests: None declared

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OP0162-HPR

THE NEED FOR PERSONALIZED, NON-PHARMACOLOGICAL INTERVENTION PROGRAMMES IN AUTOIMMUNE CONNECTIVE TISSUE DISORDERS: RESULTS OF A EULAR-FUNDED SCOPING REVIEW WITH A NESTED, DESCRIPTIVE META-ANALYSIS

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Background: Autoimmune Connective Tissue Disorders (CTDs), including Mixed Connective Tissue Disease (MCTD), systemic sclerosis (SSc) and systemic lupus erythematosus (SLE) can lead to Raynaud phenomenon, involvement of internal organs, pain in joints, skin abnormalities and muscle weakness. Consequently, people with CTDs experience limitations in activities of daily life and restrictions in participation. Programmes promoting patient education, self-management, physical activity, and healthy life style, provided by health professionals are therefore essential to these patients. However, there is a substantial gap of knowledge about the content which should be included in non-pharmacological intervention programmes.

Objectives: To identify, map and appraise the content of existent face-to-face, digital and/or distance-learning non-pharmacological intervention programmes for patients with MCTD, SSc and/or SLE to inform a EULAR funded project with the overall aim to develop a new distance learning programme realizable with digital technologies.

Methods: A scoping literature review with a nested descriptive meta-analysis was performed in MEDLINE [PubMed], EMBASE [OVID], CINAHL [EBSCO], PsycINFO [ProQUEST], the Cochrane Database of Systematic Reviews, OTSeeker, PEDro, and SciELO in September 2018. Furthermore, an international task-force identified additional grey literature and ongoing research projects. Eligibility checks of the articles based on pre-defined in- and exclusion criteria and data extractions were done independently by two researchers. The interventions contained in these programmes were extracted using the "Better reporting of interventions: template for intervention description and replication (TIDieR) checklist and guide". A Sankey diagram and descriptive statistics were used to illustrate the relationships between the interventions and other characteristics of the studies. The protocol was pre-published on ResearchGate.

Results: Of 6667 identified records, 94 papers were eligible. Sixty-one studies (65%) were conducted between 2000 and 2018, mainly in the USA (35 [37%]), Brazil (11 [12%]) and Italy (10 [11%]). Thirty-three studies (35%) were randomised controlled trials, 17 (18%) were one-group pre-post-test designs and 12 (16%) clinical trials without randomisation (Figure 1). Overall, 4274 patients participated; 2955 had SLE [69%] and 1319 had SSc [31%], thus no studies with MCTD were found. The main content of the intervention programmes included exercises (22 [23%]), self-management (9 [10%]) and education (7 [7%]), with treatment goals focusing mainly on physical function (27 [29%]), quality of life (19 [20%]) and fatigue (12 [16%]). The interventions (67 [72%]) included or were based on an initial face-to-face component.

Figure 1. Sankey diagram. This diagram illustrates the relationships between the different study characteristics. The width of the grey connections lines between the bars are reflecting a higher congruency between studies (higher congruency is displayed with wider lines).

Conclusion: There was a variety of programmes content and interventions due to different consequences regarding activity limitations and participation restrictions in autoimmune CTDs. This highlights the need for personalized, multicomponent, non-pharmacological interventions.

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Other orphan diseases

OP0163

LONG-TERM EFFECTIVENESS OF CANAKINUMAB IN PATIENTS WITH CRYOPYRIN-ASSOCIATED PERIODIC SYNDROME

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Background: Treatment options for autoinflammatory diseases include anti-interleukin (IL)-1 therapies since the IL-1 pathway is crucial in immune dysregulation in patients with monogenic periodic fever syndromes.

Objectives: To gain further insights from routine clinical practice with respect to long-term effectiveness and safety of canakinumab (CAN) in pediatric and adult patients with CAPS (cryopyrin-associated periodic syndrome, including Muckle-Wells syndrome [MWS], familial cold autoinflammatory syndrome [FCAS], neonatal onset multisystem inflammatory disease [NOMID]/chronic infantile neurological cutaneous and articular syndrome [CINCA]), FMF (familial Mediterranean fever), TRAPS (tumor necrosis factor receptor-associated periodic syndrome) and HIDS/MKD (hyperimmunoglobulinemia D syndrome/mevalonate kinase deficiency).

Methods: RELIANCE is a prospective, non-interventional, multi-center, observational study based in Germany with a 3-year follow-up enrolling patients aged ≥ 2 years with clinically confirmed diagnoses who routinely received CAN. 6-monthly clinical assessment and evaluation of patient-reported outcomes is aligned with routine clinic visits. CAPS disease activity was determined by physicians' and patients' assessment. Endpoints were effectiveness and safety of CAN under standard clinical practice conditions.

Results: The interim analysis includes 52 CAPS patients with prior long-term CAN treatment (43.1% females) enrolled by September 2018. 44.2% of the patients participated in the β -Confident study previously. The mean age was 20.7 years (4.0–79.0 years) at baseline and the mean duration of prior CAN treatment was 5.4 years (0.0–11.0 years). 40 patients (76.9%) were diagnosed with MWS, the other patients had FCAS (2), NOMID/CINCA (7) or atypical CAPS (1) (subtype diagnosis of 2 patients not available). Mutations in Nod-like receptor family pyrin domain-containing-3 were identified in 34 patients (68.0%) including E311K (7), Q703K (5), V198M (5), T348M (5), A439V (4). A baseline screening revealed sensorineural hearing loss (62.0%), papillitis (84.0%) and neurological symptoms (71.4%) in patients which had not been detected previously. The majority of patients had no disease related symptoms at baseline and 6 months demonstrating sustained remission in patients receiving long-term CAN treatment. The following disease related symptoms (mild/moderate and severe) were observed in the analysis cohort (N=31) at baseline and 6 months, respectively – disease symptom: baseline (mild/moderate, severe) vs. 6 months (mild/moderate, severe) – urticarial skin rash: 19.4%, 6.5% vs. 25.8%, 0.0%, arthralgia: 32.3%, 0.0% vs. 29.0%, 9.7%, myalgia: 9.7%, 0.0% vs. 16.1%, 0.0%, headaches: 22.6%, 9.7% vs. 19.4%, 19.4%, conjunctivitis: 32.3%, 3.2% vs. 12.9%, 6.5%, abdominal pain: 9.7%, 6.5% vs. 22.6%, 9.7%. Patients' assessment of disease activity and fatigue did not change between baseline and 6 months. However, at baseline 45.5% and after 6 months 76.0% of patients had no impairment of social life by the disease. Serious adverse events were reported for 2 patients (tonsillitis, delivery at week 31).

Conclusion: The RELIANCE study longitudinally monitors the stability of effectiveness of CAN in patients with monogenic periodic fever syndromes. An initial interim analysis including the CAPS subgroup which had prior CAN treatment showed that CAN is an effective and safe treatment in those patients.

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OP0164

LEFLUNOMIDE IN COMBINATION WITH GLUCOCORTICOIDS IS SUPERIOR TO CONVENTIONAL GLUCOCORTICOID MONOTHERAPY IN PREVENTING RELAPSE IN IGG4-RELATED DISEASE: A RANDOMIZED, OPEN-LABEL, CONTROLLED TRIAL

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Background: Glucocorticoids (GCs) was recommended as first-line therapy for the induction of remission of IgG4-related disease (IgG4-RD), whereas, a large proportion of patients may fail to maintain remission with GCs monotherapy. Considering that recurrent flares might lead to irreversible organ damage, immunosuppressive agents should be added. However, opinions among the experts were split on this issue [1]. Previously, we preliminarily reported the efficacy and safety of GCs and leflunomide (LEF) combination therapy for maintaining the remission of IgG4-RD [2].

Objectives: To evaluate the efficacy and safety of GCs and LEF combination therapy compared with GCs monotherapy for the management of IgG4-RD.

Methods: This study was a 12-month, randomized, open-label, controlled trial (NCT02703194). Patients were randomly allocated to GCs and LEF combination therapy or GCs monotherapy with 1:1 group allocation ratio. The dosage of LEF was 20 mg/d. All received GCs with a predefined taper regimen from a starting dosage of 0.5–0.8mg/kg/d. The primary outcome was time to relapse and the main secondary outcomes were time to complete response (CR) and time to remission.

Results: A total of 66 patients were randomly assigned (33 in each group) (Figure 1). The demographic and disease characteristics of the patients in the ITT population were similar between the treatment groups. The mean initial dosages of GCs were similar (GCs and LEF: 0.68 mg/kg/d, GCs: 0.70mg/kg/d, $P=0.529$). The cumulative relapse rates at 12 months were 21.2% in the GCs and LEF group and 50.0% in the GCs group ($P=0.023$). GCs and LEF combination therapy was significantly superior to GCs monotherapy with respect to the time to relapse (HR, 0.35; 95% CI, 0.13 to 0.90; $P=0.030$) (Figure 2), and was superior to GCs monotherapy with respect to the time to remission (HR, 1.74 95% CI, 0.97 to 3.10; $P=0.063$). These two groups were similar when analyzing the time to CR (HR, 1.57 95% CI, 0.91 to 2.74; $P=0.108$). At 12 months, 14 (42.4%) patients in the GCs group and 22 (66.7%) patients in the GCs and LEF group successfully tapered GCs to 10 mg/d or less with no relapse ($P=0.048$), 6 (18.2%) patients in the GCs group and 18 (54.5%) patients in the GCs and LEF group to 5 mg/d or less with no relapse ($P=0.002$). The relapsing organs of these relapsing patients were salivary glands ($n=5$), pancreas ($n=5$), bile ducts ($n=5$), retroperitoneal fibrosis ($n=3$), lacrimal gland ($n=1$), and skin ($n=1$). The incidences of new-onset diabetes mellitus (GCs and LEF: 21.21%, GCs: 27.27%, $P=0.566$) and the infections (GCs and LEF: 18.18%, GCs: 12.12%, $P=0.566$) were similar between two treatment groups.

Conclusion: The efficacy of LEF in combination with GCs therapy was significantly superior to conventional GCs monotherapy in preventing relapse in patients with IgG4-RD. LEF can be used as a steroid-sparing agent in the management of IgG4-RD.

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Figure 1. Treatment Assignments and Withdrawal in the Intention-to-Treat Population.

Figure 2. Kaplan-Meier Analysis for Time to Relapse.

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