Background: Subjective sleep problems, including difficulties falling asleep, waking up, un-restorative sleep and daytime sleepiness are highly prevalent in patients with juvenile fibromyalgia (JFM). Sleep disturbances has been consid- ered a consequence of severe pain and depression, but also in healthy indi- viduals sleep deprivation is also a risk factor for the development of chronic widespread pain, tenderness and fatigue, suggesting the important role of sleep in pain control and in the pathophysiology of fibromyalgia.

Objectives: To estimate the incidence of polysomnographic alterations in JFM and to explore the relationship between sleep problems and the musculoskeletal pain, fatigue and mood and anxiety disorders.

Methods: 21 patients (M 3; F 18; mean age 16.1) with JFM were included. The objective sleep quality was measured by overnight polysomnography (PSG) (using the EMBELTTA MPR PG device). PSG data were compared to age and sex-matched controls. The subjective sleep disturbances were assessed by the Sleep Condition Indicator (SCI). Musculoskeletal symptoms were evaluated by using the widespread pain index (WPI). Pain intensity was evaluated on a 0-10 visual analogical scale (PVAS). Fatigue was assessed by using the Symptom Severity (SS) questionnaire. Mood and anxiety disorders were evaluated by using the Children Depression Index (CDI) and the Multidimensional Anxiety Scale for Children (MASC). Comparison of categorical data was performed by means of the Fisher’s Exact test. The relationship between sleep quality and clinical symptoms were assessed using Spearman’s rank order correlation coeffi- cient (rs). All statistical test were 2-sided and p values less than 0.05 were con- sidered statistically significant.

Results: Nineteen out of 21 (90.5%) patients complained subjective sleep distur- bances and un-restorative sleep. Seven out of 21 (33.3%) patients had mood and anxiety disorders. Eight out of 21 patients (38.1%) showed an electroencepha- lographic pattern of alpha wave intrusion in slow wave sleep (SWS). SCI was significantly correlated to CDI score rs = -0.775 (p=0.001), MASC 0.61 (p=0.005), WPI -0.731 (p=0.001), SS 0.492 (p=0.038), PVAS -0.590 (p=0.006).

Conclusion: A substantial percentage of JFM patients experience sleep distur- bances, which are, correlated with the severity of the musculoskeletal symptoms and mood and anxiety disorders. One third of JFM patients have alpha intru- sion in the SWS. The important role of sleep in pain control suggests that the development of treatments to improve sleep quality may lead to more effective management of fibromyalgia in the future.

References:

Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2020-eular.6112

AB0094 A NEW ALTERNATIVE IN THE TREATMENT OF PATIENT WITH MUTATION OF GEN LACC1

R. M. Alcobendas1, C. Quintana1, J. Arostegui2, C. Udaondo1, S. Murias Loza3, A. Reme sal1. 1University Hospital La Paz, Madrid, Spain; 2Hospital, Barcelona, Spain

Background: Few patients have been described in the literature with mutations in the Lacasa Domain containing one (LACC1) gene. Its clinical presentation usually associates sustained systemic inflammation associated with chronic polyarticular erosive arthritis. Until now, there have been multiple treatments described to try to control the disease, however, they are generally unsuccessful in the long term.

Objectives: Describe the clinical course of a patient as well as the different treat- ments used

Methods: Clinical chart review

Results: Female 18-year-old born from a consanguineous Moroccan couple. Mother, brother and sister with similar conditions. She started at 3 years with fever, anemia, intense elevation of acute phase reactants and symmetric polyarthritis (knees, elbows, carps, shoulders, hands and ankles). Subsequent whole exome sequencing identified c.128,129delGT mutation in the LACC1/ FAMIN gene. During the course of her illness, she has received treatment with oral, intravenous and infiltrated corticosteroid, metotrekate and etanercept, without getting adequate control of the disease. In 2016, she started treatment with tocilizumab (8 mg / kg every two weeks), obtaining an acceptable control of the disease (requiring periodic infiltrations every 2-3 months due to persistent arthritis). Nonetheless, in April 2019, she consulted for clinical worsening of the arthritis and laboratory test (C reactive protein 99.7 mg / L, erythrosedimenta- tion rate 53 mm / h, leukocytes 13.500/L and neutrophils 10,930/L). At that time, she discontinued therapy with tocilizumab and started tofacitinib 5 mg every 12 hours with good evolution. Since its introduction, it has not required joint infiltration again and the inflammatory parameters (persistently elevated previously) have normalized.

Conclusion: The jak kinases inhibitors may be a treatment option in those patients with bad response to conventional therapy.

References:

Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2020-eular.5555

AB0095 JUVENILE ONSET SYSTEMIC LUPUS ERYTHEMATOSUS WITH SJÖGREN’S SYNDROME: CLINICAL AND LABORATORY FEATURES.

M. Maleka1, L. Nikiforov3, 1V.A. Nasonova Research Institute of Rheumatology, Pediatric Department, Moscow, Russian Federation; 2V.A. Nasonova Research Institute of Rheumatology, Pediatric Department, Moscow, Russian Federation

Background: Systemic lupus erythematosus with juvenile onset (SLE) with Sjögren’s syndrome (SS) in children is a poorly studied and rare combination, the frequency of which, according to the literature, is 7.5-10.0%1.

Objectives: To study demographic data, specific features of SLE with SS in single center.

Methods: Retrospective study of all consequently patients (pts) of single-center in pediatric department with combination of SLE and SS.

Results: SS was verified in 14 pts with SLE (14.3% were boys), which amounted to 15.5% of all pts with SLE. The median age of SLE onset was 13.5 y.o. [9.3; 14.9]. The median of disease duration at the time of SS verification was 1.3 y [0.6; 2.9].

References:


The youngest brother has started his therapy by methotrexate. It should be noted that the family has the eldest brother (20 y.o.), who has been suffering from arthritis since an early childhood with similar clinical picture. We are going to perform genetic analysis of the NOD2/CARD15 gene for the eldest brother.

Conclusion: Our clinical case shows that extremely rare BS may be misdiagnosed as JIA. Lack of efficacy of the etanercept therapy and uveitis de novo developing may be caused by genetic (non-idopathic) nature of disease. Classic triad of boggy-arthritis, granulomatous uveitis and/or skin lesions without acute phase markers is required to perform genetic assay for the detection of a pathogenic mutation of the NOD2/CARD15 gene. This case is remarkable by the presence of BS in two (or 3) children of the family.

References:

Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2020-eular.6239