Assessing the children’s health related quality of life measure, adherence to therapy, comorbidities as well as motivation on individual basis.

Disclosure of Interest: None declared


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Other orphan diseases

**THU0606**

**EFFECT OF INTERLEUKIN-1 ANTAGONISTS ON THE QUALITY OF LIFE IN FAMILIAL MEDITERRANEAN FEVER PATIENTS**

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**Background:** Familial Mediterranean Fever (FMF) is characterised by febrile inflammatory painful attacks of peritonitis, pleuritis, arthritis and erysipelas erythema. Colchicine is the mainstay of treatment in FMF but about 10% patients do not respond well to colchicine. IL-1 antagonists have been shown to be effective in the prevention of attacks in colchicine resistant FMF (crFMF) patients. Herein we investigated effect of these agents on quality of life of crFMF patients.

**Objectives:** To investigate effect of IL-1 antagonists on quality of life of crFMF patients.

**Methods:** Data is derived from Gazi FMF cohort which was established in year 2010. Since then data of patients with FMF who were diagnosed according to the Tel Hashomer criteria were registered. Co-morbidities, detailed attack characteristics, treatments, laboratory parameters and impact of FMF on their life in terms of quality of life were recorded. A retrospective cohort analysis was made from records of patients who were treated with IL-1 inhibitors. SF-36 form was filled before and 3 months after the IL-1 antagonist treatment. Wilcoxon test was used for the analyses and a p value equal or less than 0.05 is considered as statistically significant.

**Results:** There were 41 patients (24 women and 17 men). Anakinra was used in 33 patients and 8 patients received canakinumab. There was a statistically significant reduction in the frequency, severity and the duration of attacks after treatment with IL-1 antagonists (p<0.001, for each). A statistically significant improvement was observed in all domains of SF-36 (figure 1).

**Conclusions:** IL-1 inhibitor therapy reduces frequency, severity and duration of attacks and significantly improves the quality of life of crFMF patients.

Disclosure of Interest: None declared


**THU0605**

**FACILITATING PATIENT CENTRED CARE: THE DEVELOPMENT OF ILLUSTRATED MULTIDIMENSIONAL PATIENT REPORTED OUTCOME MEASURE FOR CHILDREN WITH JUVENILE IDIOPATHIC ARTHRITIS**

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**Background:** The advances in paediatric rheumatology management have mandated a drastic change in the way children with juvenile arthritis are assessed and monitored. As a consequence, there has been a call for new outcome measures that reflect a more holistic approach to day to day standard management. Such an approach can assess construct outcome measures of children with arthritis and acknowledges the critical link between physical and psychological health as well as adherence to therapy amongst the children living with inflammatory arthritis.

**Objectives:** To assess validity; reliability and responsiveness to change of an illustrated child/parent Multidimensional Patient Reported Outcome Measures questionnaire which can assess construct outcome measures of children with juvenile inflammatory arthritis.

**Methods:** 106 children with juvenile inflammatory arthritis were included in this web-based multicentre study. The questionnaire was developed by integrating information obtained from children living with JIA as well as their parents. The questionnaire included 5 main categories which are patient-centred: Health related quality of life: functional ability (children health assessment questionnaire) and quality of life (10-items reflecting psychological, social, school and behavioural issues as well as the patient’s own perception). 2. Disease activity measure: pain intensity, the child’s overall well-being, measure of fatigue and morning stiffness using a 0–10 numerical visual analogue scale, 3. self-reported joint tenderness and swelling. 4. Current medication, side effects as well as adherence to therapy (2 questions using a 0–10 numerical visual analogue scale); 4. Comorbidities as well as 5. Patient motivation. All the items were supported by illustrations to explain the question and make it easier to understand. The questionnaire has parent and patient versions. The disease activity status was assessed using JADAS-27.

**Results:** The questionnaire was reliable as demonstrated by a high-standardised alpha (0.890–0.978). The questionnaire items correlated significantly (p<0.01) with clinical parameters of disease activity. The patient reported tender joints correlated significantly with the physician’s scores (0.842). Changes in functional disability quality of life as well as the motivation score showed significant variation (p<0.01) with diseases activity status in response to therapy. The illustrated PROMs questionnaire showed also a high degree of comprehensibility (9.6).

**Conclusions:** Integrating patient reported outcome measures into standard clinical practice is feasible and applicable. This version of illustrated multidimensional questionnaire was found to be valid and reliable. It provides informative quantitative measure for the disease activity core set data, and in the meantime, facilitates assessing the children’s health related quality of life measure, adherence to therapy, comorbidities as well as motivation on individual basis.

Disclosure of Interest: None declared

Objectives: Describe the clinical characteristics, genetic variants of different autoinflammatory diseases of a cohort of adult patients with follow-up in a 3rd level hospital.

Methods: We carried out a descriptive cross-sectional study of adult patients with follow-up in reference hospital consultations with suspicion and/or diagnosis of autoinflammatory disease. Clinical, demographic, and treatment variables were characterised. A descriptive analysis was carried out by subgroup of pathology. The qualitative variables were expressed in frequency and percentages and the quantitative variables in median and interquartile range (IQR). The statistical software IBM SPSS v.18 was used for the analysis

Results: A total of 51 patients were included, 31 women (60.8%). Overall, the median age at diagnosis was 28 years (RI 15–40). The most used treatment was colchicine (60.8%), steroids (43.1%), FAMES (21.6%), anti IL-1 (17.6%). By sub-group of pathologies the most prevalent with 31 cases (60.7%), 16 women (51.6%) was familial Mediterranean Fever (FMF) presenting 17 patients (49.4%) pathogenic mutations, 8 patients (25, 8%) low penetrance mutations and the rest (25.8%) genetic polymorphisms. The median age at diagnosis was 30.6 years (IQR16–44). The most common symptoms were fatigue (80,6%), acute phase reactants (APR) elevation (71%), fever and myalgias (67.7%), arthralgias (64%), abdominal pain (48.4%), rash (31%), arthritis (29%), lymphadenopathy (29%), serositis (12.9%).

With regard to TRAPS, 10 patients were diagnosed (19.6%), 7 women (70%), with a median age at diagnosis of 26.5 years (IQR 15–31.5). The most common symptoms were arthralgias, fatigue and myalgias (90%) with elevated APR, fever (80%), abdominal pain (70%), rash (60%) and arthrits (50%). There were 2 cases of women (4%) with Schnitzler syndrome, with arthrits symptoms, elevated APR and chest pain (in 1 case). There were 6 cases (11.8%) of disease associated with NOD2 gene with a median age at diagnosis of 30 years (IQR 12.5–40), 4 were women (66.7%). The most common symptoms were arthralgias, fever and myalgias (83.3%) with increased APR, arthrits (66.7%), abdominal pain, oral thrush and rash (66.7%).

In the disease associated with NLPR12 gene there were 2 cases (3.9%), both cases were women. The most common symptoms were arthralgias, arthrits, fever, fatigue, increased APR and serositis (in one case). There was a case of sickness associated with the NLPR3 gene (cryopyrinopathy) (2%) in a woman with fever and refractory pyoderma gangrenosum and another woman with a diagnosis of Muckle-Wells (2%) with clinical signs of deafness since childhood, conjunctivitis, fever and arthrits. There was a case of PFAPA (2%) in a woman with fever, myalgias, rash and lymphadenopathy.

Conclusions: In our series, the most frequent pathology was FMF followed by TRAPS. The most prevalent symptoms were systemic manifestations (fever, fatigue, APR elevation) and musculoskeletal manifestations.

Disclose of Interest: None declared


THU0608 THE RELATIONSHIP BETWEEN SERUM SOLUBLE KLOTHO, FGF-23 LEVELS AND FLOW-MEDIATED DILATATION (FMD) IN PATIENTS WITH FAMILIAL MEDITERRANEAN FEVER (FMF)

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Background: Familial Mediterranean fever (FMF) is a disease having inflammatory attacks. Systemic inflammation has an important role in the etiology of endothelial dysfunction. Fibroblast Growth Factor 23 (FGF-23) is expressed by osteoblasts and needs serum soluble klotho. FGF-23 provides phosphate regulation. Increased FGF-23 is associated endothelial dysfunction. An invasive way of measuring endothelial dysfunction is Flow-Mediated dilation (FMD).

Objectives: To investigate the possible relationship between FGF23, serum soluble klotho levels and FMD in patients with FMF and healthy subjects.

Methods: Between March 2017 - September 2017, sixty -FMF patients that following up at Cumhuriyet University Medical Faculty Rheumatology-Internal Medicine Department and thirty healthy volunteers were included into the study. Blood samples were taken from all participants and serum soluble klotho, FGF23 values were measured. Clinical findings of all patients were recorded. Blood tests were examined by Elisa method in Cumhuriyet University Department of Biochemistry. FMD assessed by Doppler ultrasound in all participants.

Results: Mean serum FGF 23 level was measured as 221.01 pg/ml pg in FMF group and 99.4 pg/ml in healthy control (HC) group. There was statistically significant difference between two groups (p=0.05). The mean serum levels of serum soluble Klotho was measured as 3.0 pg/ml FGF23 group and 8.25 pg/ml in the HC group. There was statistically significant difference between the two groups (p=0.05) (figure 1). The mean percentage of FMD was measured as 10.1 in FMF group and 18.6 in the HC group. There was statistically significant difference between the two groups (p=0.05).

Abstract THU0608 – Figure 1 Serum Levels of FGF-23 and Serum Soluble klotho in FMF patients and control group

Conclusions: We found that high FGF 23 levels in FMF patients caused a significant decrease in FMD. In addition, FGF23 may be a parameter for the early diagnosis and prognosis of possible cardiovascular events showing significant change in FMD which demonstrates endothelial dysfunction indirectly.

REFERENCES:

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THU0609 CLINICAL CHARACTERISTICS AND OUTCOME OF SPANISH PATIENTS WITH ACUTE MYELITIS ASSOCIATED WITH AUTOIMMUNE DISEASES

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Background: Acute myelitis (AM) is a focal inflammatory disorder of the spinal cord characterised by motor, sensory, and autonomic dysfunctions that usually develop during a short period (several hours to days) and may resolve over several weeks to months. Acute has been reported as an unusual complication of autoimmune diseases (AD), mainly in systemic lupus erythematosus (SLE) and Sjögren’s syndrome (SS).

Objectives: To analyse the frequency of concomitant AD in patients with AM seen in a Spanish tertiary centre and to compare their clinical characteristics and outcome with those of AM patients without other AD.

Methods: We performed a retrospective study including all the patients diagnosed with AM in our centre between January 1989 and December 2017. Patients with previous history of spinal disease (multiple sclerosis (MS), compression, trauma, arteriovenous malformation, radiotherapy) and children were excluded. Demographics, clinical, laboratory, imagerenology, therapeutic and outcome data were obtained from their medical records.

Results: During the study period 144 cases of AM were identified, 76 of them had complete data and were analysed. Most of the patient were women (47, 62%), with a mean age at diagnosis 42±17 years. The main causes of AM in our series were MS (35%), AD (18%), postviral myelitis (9%) and idiopathic (34%). The AD diagnosed in these patients were: SLE7, SS2, undifferentiated connective tissue disease,2 Graves’ disease,1 ulcerative colitis,1 polymyalgia rheumatic,1 cryoglobulinemic vasculitis1 and sarcoidosis.1 A patient with MS had concomitantly another AD (inflammatory myopathy). Most of the patients with AD (12, 80%) were diagnosed of these disorders before the AM episode (median 48 months, range: 24–120). No differences regarding to sex, age and anatomical extension of AM were detected between AD and non-AD patients. The recurrence of AM was more frequent in non-AD patients (7% vs 54%, p<0,001) and a tendency to less severe clinical course was found in AD patients (67% vs 41%, p=0.08).

Conclusions: AD were an important cause of AM in our series and this usually occurred after diagnosis of AD, so it is important to consider AM when these patients develop neurological dysfunctions that suggest spinal cord damage. The outcome of AM seem to be better in patients with concomitant AD.