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 considered a consequence of severe pain and depression, but also in healthy individuals 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 EMBLETTA 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 analogue 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 coefficient (rs). All statistical test were 2-sided and p values less than 0.05 were considered statistically significant.

Results: Nineteen out of 21 (90.5%) patients complained subjective sleep disturbances 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 electroencephalographic pattern of alpha wave intrusion in slow wave sleep (SWS). SCI was significantly correlated to CDI score rs -0,775 (p≤0,0001), MASC 0,61 (p=0,005), WPI -0,731 (p=0,001), SSI 0,492 (p=0,038), PVAS -0,590 (p=0,006).

Conclusion: A substantial percentage of JFM patients experience sleep disturbances, which are, correlated with the severity of the musculoskeletal symptoms and mood and anxiety disorders. One third of JFM patients have alpha intrusion 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.

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AB0993

A CASE SERIES OF KAWASAKI DISEASE FROM KENYA

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Background: Kawasaki disease (KD) has been described across the globe, including the African continent, but none yet from Kenya.

Objectives: To describe the clinical features and management strategies of pediatric patients diagnosed with Kawasaki Disease at a tertiary referral hospital in Kenya.

Methods: A retrospective chart review was undertaken for the period January 2013 to December 2017 for all pediatric patients admitted at Aga Khan University Hospital Nairobi, Kenya. All medical records with a discharge diagnosis of Kawasaki disease were reviewed, de-identified and data extracted using a data collection tool.

Results: Among the 15 cases identified, 8 (53%) had complete KD. The mean age was 1 year 10 months with a slight increase in males (53.3%). The mean duration of symptoms at diagnosis was 7.2 days (range 1-11 days). Fourteen patients (93.3%) received both intravenous immunoglobulin and aspirin but dosing varied from high dose aspirin (80-90mg/kg/day) to low dose aspirin (3mg/kg/day). Baseline cardiac evaluations were done among these 14 (93.3%) and one patient was found to have bilateral dilated coronaries. Only 5 patients (33.3%) had repeat echo examinations within 6 weeks after diagnosis all of which were normal.

Conclusion: The challenges faced in the management of KD in Kenya include awareness of the disease, access and expertise to pediatric echocardiography, follow-up, access and cost of IVIG. Increasing awareness and improving health care resources is important in improving outcomes of KD in Kenya.

Keywords: Kawasaki Disease, Pediatric Rheumatology, Kenya, Global Health, Vasculitis

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AB0994

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

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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 treatments 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, methotrexate 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, erythrocyte sedimentation rate 53 mm / h, leukocytes 13,500/μL and neutrophils 10,930/μL). At that time, she discontinued therapy with tocilizumab and started tofacitinib 5mg 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 kinase inhibitors may be a treatment option in those patients with bad response to conventional therapy.

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AB0995

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

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Background: Systemic lupus erythematosus with juvenile onset (jSLE) with Sjogren'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 jSLE with SS in single center.

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

Results: SS was verified in 14 pts with jSLE (14.3% were boys), which amounted to 15.5% of all pts with jSLE. The median age of jSLE 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].

Expressed constitutional disorders (fever, weight loss) observed in 13 pts (91.7%). 11 pts (78.6%) had acute cutaneous lupus, 5 pts (35.7%) – chronic cutaneous lupus, 3 (21.4%) – oral and nasal ulcers, 6 (42.8%) – nonscarring alopecia, 9 pts (64.3%) – polyarthritis, 2 pts (14.3%) – renal involvement, 2 pts (14.3%) – serositis, 1 (7.1%) – interstitial lung disease, 5 pts (35.7%) – neuropsychiatric disorder, including psychosis in 2 (14.3%). 4 patients had skin lesions atypical for SLE (1 - annular erythema, 1 - erythema nodosum, 2 - Ro-associated skin vasculitis). 12 pts (85.7%) had generalized lymphadenopathy. 12 pts (85.7%) had various hematological disorders: anemia – in 5 pts (35.7%), leukopenia – in 9 (64.3%), isolated lymphopenia in 1 (7.1%), thrombocytopenia – in 4 pts (28.6%). 13 pts had isolated involvement of salivary glands, 1 – combined with lacrimal glands. The decrease in salivary gland function was recorded in 50% of cases, hypolacrimia – in one case. Recurrent parotitis was present in only one case (7.1%). ANA were detected in 100% pts, anti-dsDNA – in 10 pts (71.4%), anti-Sm – in 7 pts (50.0%), anti-Ro – in 10 (71.4%), anti-La – in 7 (50%), RNP-70 – in 5 pts (35.7%), RF+ – in 6 pts (42.9%), hypocomplementemia – in 3 pts (21.4%). The most common was the combination of positive ANA, anti-dsDNA, antiRo with acute cutaneous lupus, polyarthritis, generalized lymphadenopathy and expressed constitutional disorders – 8 pts (57.1%). 4 pts (28.6%) had polyclonal hypergammaglobulinemia. 3 pts (21.4%) had concomitant autoimmune non-rheumatic disease; 1 - autoimmune hepatitis, 1 - type 1 diabetes mellitus, 1 - autoimmune thyroiditis. Median disease activity by SLEDAI at the time of jSLE verification was 11.5 scores [9.25;15.7].

Conclusion: According to our results, the frequency of detection of secondary SS in jSLE was higher than the literature data. The clinical features include a high frequency of constitutional disorders, lymphadenopathy, skin manifestations, high frequency of antiRo with a significantly lower incidence of kidney involvement, serositis than jSLE without SS. In pts with a diagnosis of SLE, the possibility of developing secondary SS should be considered (specially in girls with antiRo positive), the early detection of which affects the choice of therapy and prognosis.

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AB0996 FAMILY CASE OF BLAU SYNDROME IN 3 CHILDREN, INITIALLY DIAGNOSED AS JUVENILE IDIOPATHIC ARTHRITIS

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Background: Juvenile Idiopathic Arthritis (JIA) is heterogeneous group of diseases which may include genetically determined conditions. Extremely rare monogenic hereditary autoinflammatory disease, such as Blau syndrome (BS) is usually difficult to recognize and JIA is initially established. BS is caused by a mutation in the NOD2/CARD5 gene and phenotypically characterized by triad of granulomatous polyarthritis, uveitis and skin rash.

Objectives: To present the family case of genetically confirmed BS in 2 siblings, initially diagnosed as patients with JIA.

Methods: Two brothers of 15 and 3 years old were examined in our clinic. Additional genetic assay was performed because of unusual clinical picture.

Results: *Case report.* Older brother, 15 y.o. developed arthritis of both wrist joints at the age of two. There was erythematous maculo-micropapular scaly rash on the trunk and extremities before the onset of arthritis. By 2009 (5 y.o.), the knees, ankles, and three PIP joints of the left hand were involved. Polyarthritis was characterized by severe effusion and periarticular tissues swelling. He was treated in regional hospital by NSAID, methotrexate, cyclosporine A without significant positive effect. Since 2012 etanercept was added for treatment with variable result. Inactive status of the disease is never reached. Repeated intra-articular GC injections were needed from 4 to 10 times per year. The patient was admitted to our clinic in November 2019. He was suffering from severe polyarthritis and uveitis de novo (granulomatous chorioretinitis) was detected. At the same time his younger 3 y. o. brother with recently started disease was hospitalized in our clinic. He had polyarthritis with typical features of "boggly" synovitis and tenosynovitis of wrists, ankles and knees, anterior uveitis was determined. The onset of arthritis was preceded by a small-spotted rash with desquamation. This classical clinical features in younger brother let us suggest the BS. There was no increasing of ESR and CRP in both sibs throughout the course of the disease. A molecular genetic study of the NOD2/CARD15 gene in both brothers revealed the same mutation of c.1001G>A (p. Arg334Gln). Because of the absence of specific treatment for this disease and due to insufficient effect of etanercept with uveitis de novo therapy was changed to golimumab and good initial effect is reached.

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-idiopathic*) 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.

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AB0997 IS HEIGHT ADJUSTMENT NECESSARY IN PEDIATRIC DENSITOMETRY IN ALL CHILDREN?

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Background: The current guidelines of the International Society for Clinical Densitometry (1) recommend that in children with linear growth or maturational delay, Z score results should be adjusted. Height for age Z score (HAZ) adjustment is valid and can be calculated using the formula the formula proposed by Zemel et al(2). It is possible that pediatric populations without linear growth or maturational delay, also benefit from HAZ, to prevent bone size from influencing the final Z score.

Objectives: To evaluate Z score variability adjusted and without adjusting for height for age.

Methods: We analysed data from densitometry performed on patients 2-20 years of age, from 2016 to 2018, assessed in the pediatric rheumatology office of our hospital for presenting risk factors for low bone mass/osteoporosis. The HAZ was calculated according to Zemel's formula.

Results: Data from 103 patients are presented. Its characteristics are summarized in Table 1

Table 1.

Mean age	9,8 years
Female	52,4%
Height Percentil ≤ 3	6,8%
Height Percentil ≥ 97	4,9%
LBM (Z score ≤ -2) spine	8,2%
LBM HAZ spine	6,4%
LBM whole body	10,5%
LBM HAZ whole body	7,2%

The table shows that the proportion of patients with BMD decreases in both the spine region and the whole body when adjusting for HAZ.

When evaluating the relationship between densitometric measurements we found that spine Z score (ZsS) and whole body Z score (ZsWB) had a correlation coefficient of 0,73 (p<0,001). There were no differences between their averages (p=0,170).

At the LBM cut-off point (Z score ≤ -2) there were discrepancies in 7%, where 5% presented LBM in ZsWB but not in ZsS. The concordance index at this point was 0,557.

When comparing these measures with their HAZ adjusted equivalents, we observe:

HAZ adjusted ZsS vs ZsS without adjusting: There were no differences between their averages (p=0,913) with a correlation coefficient of 0,78 (p<0,001). Concordance index at cut-off point for LBM was 0,498, with a discrepancy of 7%, where 2% had LBM according to HAZ adjusted ZsS, but not to ZsS without adjusting.

HAZ adjusted ZsWB vs ZsWB without adjusting: There were no differences between their averages (p=0,367) with a correlation coefficient of 0,82 (p<0,001). Concordance index at cut-off point for LBM was 0,557, with a discrepancy of 7%, where 2% had LBM according to HAZ adjusted ZsWB, but not to ZsWB without adjusting.

Conclusion: There are discrepancies at the LBM cut-off point depending on the HAZ adjustment.

The pediatric population without linear growth or maturational delay, can also benefit from HAZ adjustment, especially those with high height percentiles in which their size can hide a diagnosis of LBM.