B6 mice (<0.0001), reflecting increased anxiety. There were no differences motor coordination, supporting an anxiety phenotype leading to decreased movement in the open field. On novel object placement testing, there were no differences in time spent with the novel object. Lastly, the forced swim test revealed that B6.Nba2 mice spent more time immobile than B6 mice (p<0.02), suggesting a depressive-like behavior in B6.Nba2 mice.

Conclusions Our data suggest that the B6.Nba2 mouse model expresses a strong anxiety phenotype, a depressive phenotype, but no deficits in spatial memory. This observation warrants further exploration of the B6.Nba2 as a mouse model of NPSLE along with analyses of brain histology and morphology and future evaluations of the relationship between IFN-, autoantibody levels and neurological disease manifestations.

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108 EVALUATION OF PSYCHOMETRIC PROPERTIES OF THE PATIENT-REPORTED OUTCOMES MEASUREMENT INFORMATION SYSTEM PHYSICAL FUNCTION 10-ITEM SHORT FORM IN SYSTEMIC LUPUS ERYTHEMATOSUS

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Background While the PROMIS (Patient-Reported Outcomes Measurement Information System) physical function short form 10a (PF10a) is both practical and acceptable for implementation in routine clinical practice, its psychometric properties have not been evaluated in Systemic Lupus Erythematosus (SLE). We examined the validity and responsiveness of PF10a in SLE among a racially/ethnically diverse clinic population and developed estimates of the minimally important difference (MID).

Methods Data were derived from electronic health records for all SLE patients seen in a university-based rheumatology clinic between 2013 and 2018. We evaluated the PF10as floor and ceiling effects among different racial/ethnic groups. Construct validity was assessed by examining Spearmans correlation coefficients between the PF10a and other patient-reported (pain (scale 0–10) and pain visual analogue scale (VAS) (scale 0–100)), physician-reported (SLE disease activity index (SLEDAI)) and laboratory (erythrocyte sedimentation rate (ESR)) measures. Known-group validity was assessed by evaluating effect size (Cohens d) between categories of pain (no pain vs. moderate-severe pain). We used standardized response means to examine the responsiveness of the PF10a to longitudinal changes in pain and SLEDAI. MID was estimated using distribution based and anchor-based methods.

Results We included 612 patients in cross-sectional analyses of validity and 462 patients in longitudinal analyses of responsiveness. Mean age was 40.5±14.6, 87% were female and 32% Caucasian. The PF10a had ceiling effects above the commonly accepted criteria of 15% among Caucasian (23%), Asian (23%) and Other (17%) race/ethnicities, and no floor effects. Construct validity analyses showed strong correlations (=0.66, p<0.05) with pain VAS, moderate correlations (=0.58, p<0.05) with pain, and weak correlations with ESR (=0.25, p<0.05) and SLEDAI (=0.16, NS). Known-group validity analyses showed large differences among pain groups (Cohens d=1.49, p<0.05). The PF10a was responsive to improvements in pain (SRM=0.5) and SLEDAI (0.49), but less so to deteriorations in pain (SRM=−0.42) or SLEDAI (SRM=−0.24). Distribution-based MIDs were +8 for improvement and −7 for deterioration. Anchor-based MIDs were +2 for improvement, −3 for deterioration with pain as anchor and +5 for improvement, −5 for deterioration with SLEDAI as anchor.

Conclusions Although the PF10a showed some ceiling effects, it had good validity in this young racially/ethnically diverse sample with SLE. The PF10a was responsive to improvements in pain and disease activity. The anchor-based MIDs appear to be similar to those reported for PF10a in rheumatoid arthritis. This information supports the use of the PF10a in SLE and provides important information to facilitate interpretation of scores.

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109 DERMAL LYMPHATIC CHARACTERIZATION AND PHOTOSENSITIVITY IN THE MRL/LPR LUPUS MODEL

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Background Proper function of lymphatic vessels is needed to limit the magnitude and duration of tissue inflammation, and in direct regulation of immune cell activity. Inflammatory states such as rheumatoid arthritis and psoriasis are associated with lymphatic dysfunction, but lymphatics in lupus models have not been well characterized. SLE patients are photosensitive, developing inflammatory skin lesions upon exposure to even ambient ultraviolet radiation (UVR). We hypothesized that lymphatic dysfunction may contribute to photosensitivity in lupus.

Methods MRL/MpJ-Faslpr/lpr (lpr) mice and age-/sex-matched MRL controls were evaluated at baseline, 1 day, 1 week and 1 month after exposure to 2000–2500 J/m^2 of UVB radiation. Lymphatic function was assessed with an intradermal injection of 1 µL of 2% evans blue (EB) to the ear, followed by measurement of EB concentration in the draining lymph node (dLN) 1 min later. Flow cytometry of ears and dLN allowed quantification of resident cell populations. Kruskal-Wallis test, followed by Dunns test for multiple comparisons were used to compare the groups; data is presented as mean ±SE, with a two-tailed p-value of <0.05 considered significant.

Results At baseline, the effective flow of EB, calculated as EB concentration in the dLN, per ear lymphatic endothelial cell (LEC) number, is comparable between the 2 groups. Following UVR, in both lpr and control mice, there is an increase in the effective flow of EB to the dLN, that starts as early as 1 day, and continues to increase at 1 week, in similar levels between the 2 groups. At 1 month post-UVR, however, the flow continues to improve in the MRL control mice, but there is no similar improvement in the lpr mice. In fact, while at baseline, 1 day and 1 week the mean relative lymphatic flow in the lpr mice ranges from 1.6-, 2.5- and 0.9- fold of controls, respectively, at 1 month the effective flow in the lpr mice is only 5% that of controls (p=0.007).

Conclusions Dermal lymphatic network of lupus models have never been characterized, despite evidence for a major role of lymphatics in the regulation of chronic inflammation and autoimmunity. Here we provide indications to impaired long-term response of lymphatic drainage in the lpr lupus strain, which can lead to reduced lymphatic regulation of the immune response, contributing to the unchecked skin inflammation known to occur in SLE.

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110 IMPACT OF THE DIAGNOSIS OF JUVENILE SYSTEMIC LUPUS ERYTHEMATOSUS IN PATIENTS AND THEIR PARENTS

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Background The influence of psychosocial aspects on Juvenile Systemic Lupus Erythematosus (JSLE) is known both in the triggering of the disease and in reactivation. Since it is a chronic disease in pediatric patients, its diagnosis has an important impact on both the patient and the family. The understanding of the diagnosis, evolution and treatment of the disease leads to a better adherence to the treatment, consequently better evolution.Objective: To evaluate the impact of the diagnosis of JSLE in the life of patients and parents and the degree of understanding about the disease and treatment.

Methods Pilot study with application of a questionnaire containing epidemiological data, questions about the understanding of the disease, psychological impact on diagnosis and currently and association with a stressor event. This questionnaire was applied to the jSLE patients and their parents accompanied at pediatric rheumatology department of Santa Casa de São Paulo. Qualitative data were submitted to exploratory descriptive analysis.

Results 24 patients and 21 parents answered the questionnaire. 84% of the patients were female and 95% of the relatives were mothers. The age ranged from 9 to 17 and from 28 to 46 years for the patients and the parents, respectively. The follow-up time was 33.3±5.7 months. Only 21% of the patients and 38% of the parents were able to define SLE as an autoimmune disease. Regarding the cause of SLE, 29% of parents and 13% of children do not know but many parents associated with the emotional issue, while patients related to altered immune system. Both parents and patients associated a stressor event with the onset of jSLE. Regarding treatment both children and parents demonstrated an awareness of the need for appropriate medication and follow-up, but also described the importance of sun protection.

The main feeling reported at the time of diagnosis was sadness for both parents and patients. Already today, 50% of children and parents express a feeling of conformity and tranquility, knowing how to deal better with the disease, but still have some degree of concern.

Conclusions jSLE is a disease that brings a sense of sadness and there is an association of a stressor event for both patients and parents. The health team must be able to clarify the lupus disease, considering limitations of understanding of the patients and parents, which allows a better control of the disease, bringing tranquility.

Funding Source(s): None

111 KIKUCHI-FUJIMOTO DISEASE IN CHILDHOOD ONSET LUPUS: A CASE SERIES FROM A TERTIARY CARE CENTER IN NORTH INDIA

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Background Kikuchi-Fujimoto disease (KFD) is a rare disorder described mostly in adolescents and young adults. It is a great mimicker, has a diverse clinic-laboratory profile and appears to be an under-recognized disease entity, especially in children.

Methods We report 6 children who presented with fever and lymphadenopathy and had findings on fine needle aspiration cytology (FNAC) and/or histopathology that were compatible with a diagnosis of KFD. Two amongst these 6 patients developed lupus.

Results Case 1: A 7 year old boy was admitted with complaints of high-grade fever and left submandibular lymph node swelling for 2 weeks. A possibility of suppurative lymphadenitis was kept and he was treated with antimicrobials. He was the youngest born to a consanguineously married couple who had lost two children previously to a lupus-like illness. The striking family history suggested the possibility of a monogenic form of lupus (early complement deficiency). C1q levels were found to be significantly low (C1q level: 0.27 mg/L; normal (102–170 mg/L)) Genetic analysis revealed a nonsense mutation in the C1QA gene (c.622C>T Q208X). Investigations revealed mild anemia (hemoglobin:107 gm/L) and elevated erythrocyte sedimentation rate (ESR:68 mm) in 1 st hour and C-reactive protein (CRP:8 mg/dl). An excision biopsy of the lymph node showed necrotizing histiocytic lymphadenitis consistent with KFD. Immunological work-up revealed Antinuclear antibodies (ANA) 2+speckled pattern on immunofluorescence; no anti ds-DNA antibodies; no antiphospholipid antibodies. He was treated with immunosuppressants and twice-daily fresh frozen plasma. Fever subsided in 5 days and lymph nodes regressed in 2 weeks.

Case 2: A 12-year-old boy presented with fever of 7 months duration and generalized seizures followed by altered sensorium. He was treated in lines of subacute meningo-encephalitis. As there was no response to treatment he was referred to our institute. On examination, he had malar rash, frontal alopecia and bilateral, multiple enlarged cervical lymph nodes. Laboratory investigations revealed anemia, elevated ESR, high CRP and low levels of C3 and C4. ANA was positive (3+speckled); anti-dsDNA titers were 130 IU/ml (normal-<60 IU/ml); FNAC from lymph node revealed features consistent with KFD. A diagnosis of lupus and KFD was made and he was started on oral prednisolone along with