

symptoms could only be found in patients with an established diagnosis of SLE. In addition, a few rheumatologists were concerned about labeling Type 2 symptoms as ‘SLE’ since they attributed these symptoms to co-morbid conditions. Conversely, two providers noted that Type 2 symptoms could sometimes be part of Type 1 SLE activity.

**Conclusion** The Type 1 & 2 SLE model was well accepted by both patients and rheumatologists and considered as a useful approach to identifying and treating SLE manifestations. The concern that SLE could be mis-diagnosed in patients with only Type 2 symptoms indicates the importance of limiting the use of this model to patients meeting classification criteria for SLE. Many patients and several physicians suggested that the connection between Type 1 and 2 SLE may reflect an inflammation-driven subset of Type 2 symptoms.

### 1116 LONG-TERM OCULAR SAFETY OF HYDROXYCHLOROQUINE IN PATIENTS WITH CHILDHOOD-ONSET SLE (CSLE) FOLLOWED INTO ADULTHOOD

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**Background** In recent years there has been increasing concerns about the long-term ocular toxicity of hydroxychloroquine which has resulted in increased vigilance of the use of a maximum of 5 mg/kg/day. The aim of this study was to determine the long-term ocular safety of the use of hydroxychloroquine in patients with cSLE.

**Methods** The SLE Clinic database at SickKids hospital was searched for all patients followed since its inception in 1985 diagnosed and followed who were treated with hydroxychloroquine. Follow-up data was obtained into adulthood by reviewing the records of patients following transfer to care at an adult SLE clinic. While treated at SickKids all patients were treated with hydroxychloroquine at a dose 5.5-6 mg/kg/day (maximum 400 mg/day).

However, following the recent recommendations for lower dosing of hydroxychloroquine, some of the patients had their dose lowered while followed in adulthood. All patients had regular hydroxychloroquine ophthalmologic follow-up at recommended intervals of 6-12 months. Exclusion criteria were: cSLE not diagnosed at SickKids Hospital, <2 follow visits, no documentation of ophthalmology follow-up or no documentation of hydroxychloroquine dose. This study was approved at local ethics boards at all participating centers.

**Results** A total of 718 patients with cSLE diagnosed until the end of 2019 were found in the SickKids Clinic database. 15 were not diagnosed with cSLE at SickKids or had <2 follow-up visits; 4 were eliminated as the hydroxychloroquine was stopped within one month for systemic side-effects. The study cohort there consisted of 699 patients who were followed for a total of 5815.5 person years. The mean follow-up time was 8.33 years (SD 6.15 years) (minimum 0.2 years and maximum of 35.9 years). 456/699 (65%) had  $\geq 5$  year follow-up, 218 (31%)  $\geq 10$  years; and 41(6%)  $\geq 20$  years. During the follow-up time one patient stopped hydroxychloroquine for

deposition in the retina without visual changes. A second patient had hydroxychloroquine stopped based on an optometrist's examination but when reviewed by an ophthalmologist, no retinal changes were noted.

**Conclusions** In this long-term follow-up study of patients with cSLE treated hydroxychloroquine at a dose of 5.5-6.0 mg/kg/day (maximum 400 mg/day) there was no evidence of visual changes after long-term follow-up of 5815.5 person years (patients followed for up to 35.9 years). We suggest that, unlike the potential for ocular toxicity of hydroxychloroquine found in studies of adult at doses  $> 5$ mg/kg/day, patients with cSLE can be treated with 6 mg/kg/day prior to transfer to adult care.

### 1117 SLE PHENOTYPES FORMED FROM MACHINE LEARNING AND THEIR ASSOCIATIONS WITH COGNITIVE IMPAIRMENT

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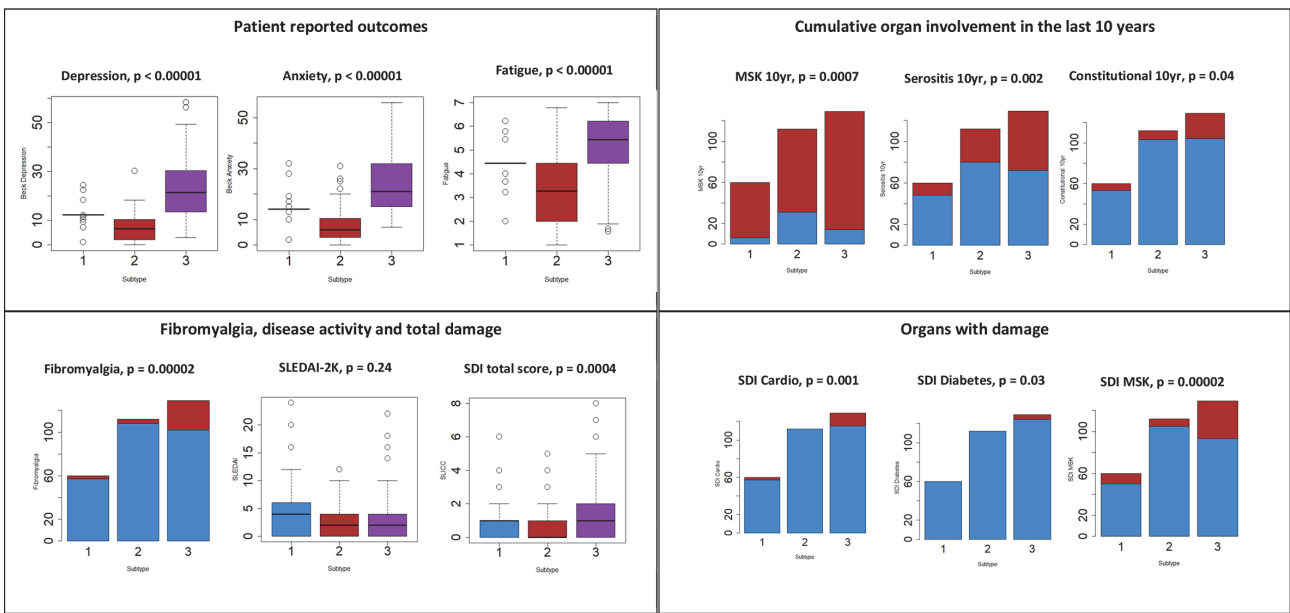
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**Background** Cognitive impairment (CI) in SLE is highly prevalent. Several factors are associated with CI: depression, pain, fatigue, medications, as well as more specific SLE factors such as disease damage, and autoantibodies. We aimed to phenotype CI in SLE using machine learning techniques to enable personalised targeted treatments.

**Methods** SLE patients aged 18-65 years attending a single completed the ACR Neuropsychological Battery (ACR-NB) cognitive assessment. Z-scores on all 19 tests of ACR-NB. ACR-NB tests were reduced using principal component analysis (PCA) to generate a factor score (CI Factor Score).

Demographic, clinical data, and patient reported outcomes including, SF-36, LupusQoL, the PDQ-20 (perceived cognitive deficits), Beck Depression Inventory-II, Beck Anxiety Inventory, and the fatigue severity scale (FSS) were analysed using similarity network fusion (SNF) to identify patient subtypes. Differences between the SNF identified subtypes were evaluated using Kruskal-Wallis tests and chi-square tests.

**Results** Of 301 patients, 89% were women, mean age and disease duration at study visit  $40.9 \pm 12.1$  years. The CI Factor score accounted for 28.8% of the variance and was associated predominantly with executive function and verbal memory. The SNF defined three subtypes (1, 2 and 3 with 60, 112, and 129 patients respectively) with distinct patterns in health-related quality of life (HRQoL), depression, anxiety, fatigue, fibromyalgia, medication usage, and damage. The CI Factor Score was significantly different between the subtypes. Examining specific cognitive domains revealed the most significant differences in the language processing and executive function tests. Subtype 3 performed worst on the majority of cognitive domains). Further exploration revealed statistical differences with depression, anxiety, fatigue, and fibromyalgia between the subtypes (figure 1). Differences were also found relating to organ involvement within the last ten years and damage within specific organs. No differences were found for SLE disease activity. Subtype 3 had higher levels of all conditions and



**Abstract 1117 Figure 1** Variables with significant differences between the three phenotyped subtypes. Box and whisker plots: blue=subtype 1, red=subtype 2 and purple=subtype 3. Bar charts: red=number of participants with variable and blue=number without

disease damage, Subtype 2 had lower levels and Subtype 1 mixed levels.

**Conclusion** The subtype with the greatest psychiatric and disease burden and reduced HRQoL performed worse on cognitive testing, specifically in domains of language processing and executive function. This subtype also had more musculoskeletal (MSK) and cardiovascular involvement. MSK involvement affects pain levels, which can impact cognition. Cardiovascular damage may be linked to cerebral small vessel disease, which is known to affect cognitive function SLE patients. Overall, these results aid with phenotyping CI in SLE and provide a baseline for our future longitudinal results.

1118

**INCIDENCE OF SYSTEMIC LUPUS ERYTHEMATOSUS IN THE UNITED STATES: ESTIMATES FROM A META-ANALYSIS OF THE CENTERS FOR DISEASE CONTROL AND PREVENTION NATIONAL LUPUS REGISTRIES**

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**Background** Epidemiologic data on systemic lupus erythematosus (SLE) are limited, particularly for racial/ethnic subpopulations in the United States (U.S.). This meta-analysis leveraged data from the Centers for Disease Control and Prevention (CDC) National Lupus Registry network of population-based SLE registries to estimate the general and by sex, race/ethnicity incidence of SLE in the U.S.

**Methods** The CDC registries were established in Michigan, Georgia, California, New York and through the Indian Health

Service (IHS). Registries used the 1997 revised ACR classification criteria for SLE as their case definition, and the surveillance time periods ranged from 2002-2009. Age-standardized incidence rates were stratified by sex and race/ethnicity from the state-based registries; the American Indian/Alaska Native (AI/AN) estimate was based only on the IHS registry that covered multiple states. For pooling data across the four sites with data on different racial/ethnic groups, we used Cochran’s Q and I<sup>2</sup> statistic to test for heterogeneity across sites. Due to significant heterogeneity, we used a random effects model to calculate pooled incidence, which allows for more variation across sites. We then extrapolated to the 2018 Census population data according to sex and race-stratified groups, including data from the IHS registry, and summed the stratum-specific estimates to provide a total population estimate of incident SLE cases in the U.S.

**Results** The registries contributed 1,057 classified cases of SLE from a mix of urban and rural areas. From the meta-analysis of the four state-based registries, the overall incidence was 5.1 (95%CI4.6,5.6) per 100,000 person-years. The incidence among females was about 7 times higher than males (8.7 vs 1.2). In the meta-analysis, the incidence rate was highest among Black females (15.9,95%CI12.5,20.3), followed by Asian/Pacific Islander females (7.6,95%CI5.5,10.4), Hispanic females (6.8,95%CI6.2,7.6), and White females (5.7,95%CI4.9,6.7). Among males, the incidence rate was highest among Black males (2.4,95%CI1.8,3.0) followed by Hispanic males (0.9,95%CI0.4,1.9), White males (0.8,95%CI0.6,1.1), and Asian/Pacific Islander males (0.4,95%CI0.2,0.6). The AI/AN incidence estimates, had the second highest rates of SLE among females (10.4,95%CI6.6,14.6) and highest for males (3.8, 95%CI1.6,7.8). Applying our sex- and race-specific incidence estimates to the corresponding population denominators from 2018 Census data, we estimated that 14,263 new persons (12,560 females and 1,703 males) in the U.S. were diagnosed with SLE and fulfill the ACR classification criteria, table 1.