Clinical Sciences

CS-01

DIFFERENTIAL EXPRESSION OF THE FSTL-1 PROTEIN AMONG PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS

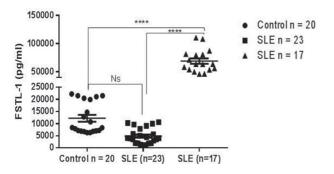
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Background There is an increasing prevalence of insulin resistance and metabolic syndrome in Systemic Lupus Erythematosus (SLE). Follistatin-like 1 (FSTL-1) is a secreted glycoprotein recently identified as a proinflammatory cytokine and emerging as a potential mediator of inflammation. Its plasma levels are elevated in rheumatoid arthritis and correlate with disease activity. In fact, FSTL-1 has been proposed as a marker of autoimmune diseases. However, it is not clear whether this protein plays a role in SLE. We aimed to evaluate the plasma FSTL-1 value of patients with SLE and the interactions between their plasma concentration, metabolic syndrome variables, markers of activity and disease damage.

Methods Plasma concentration of FSTL-1 of 40 female SLE patients and 20 healthy controls was measured by enzymelinked immunosorbent assay (ELISA). Metabolic variables, activity and disease damage were measured and analyzed. The activity of SLE was determined by Systemic Lupus Erythematosus Disease Activity Index (SLEDAI-2K). Damage accumulated was calculated by the Index of Damage SLICC/ACR (SDI Damage index). A one-way ANOVA test was performed to assess the difference between the mean FSTL-1 values in different groups. Interactions between variables of the database and their effect on FSTL-1 was evaluated using a regression tree technique.

Results Healthy controls and SLE females attended on an outpatient basis had a mean of 38 ± 16 and 40 ± 13 years old respectively. SLE patients had low levels of activity and damage measured both by the scores and by the individual parameters of inflammatory activity. The mean value of FSTL-1 was 12213 ± 1424 pg/ml for healthy individuals. Of the SLE patients 57% had a mean value of 4855 ± 662.8 pg/ml and 43% had a mean of 68000 ± 4789 pg/ml. There was a significant difference between the subgroup of SLE with high value of FSTL-1 compared to the control (p-value < 0.001) and the subgroup with



Abstract CS-01 Figure 1

low value of FSTL-1 (p-value < 0.001) (figure 1). From the regression analysis, we found a correlation between increasing plasma concentrations of FSTL-1 and presence of variables of metabolic syndrome (insulin, cholesterol and leptin) in patients with SLE. No correlation was found between FSLT-1 value and the activity or damage of the disease.

Conclusions FSTL-1 is significantly elevated in patients with SLE compared to controls. Moreover, elevated levels were observed in SLE patients with elevated leptin, cholesterol or insulin. More studies are needed to determine if there is a pathogenic role, and if this population have a worse long-term cardiovascular prognosis.

CS-02

THE LUPUS COHORT IN THE NEW CARRA REGISTRY: THE FIRST YEAR OF ENROLLMENT

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Background The Childhood Arthritis and Rheumatology Research Alliance (CARRA) aims to collect meaningful clinical data on all children affected by rheumatic disease in North America as a basis for performing high quality clinical and translational research. The new CARRA Registry (building on a prior CARRA registry with a minimal dataset) has enrolled pediatric patients with systemic lupus erythematosus (SLE) and related conditions since March 2017. We sought to describe the popula-

Abstract CS-02 Table 1 Components of SLEDAI Present within 30 days of Baseline Visit (n=184)

| SLEDAI Component | N (%) |
|--------------------------|----------|
| Seizure | 1 (0.5%) |
| Psychosis | 2 (1%) |
| Organic Brain Syndrome | 1 (0.5%) |
| Visual Disturbance | 1 (0.5%) |
| Cranial Nerve Disorder | 1 (0.5%) |
| Lupus Headache | 8 (4%) |
| Cerebrovascular Accident | 0 |
| Vasculitis | 6 (3%) |
| Arthritis | 24 (13% |
| Myositis | 8 (4%) |
| Urinary Casts | 25 (14% |
| Hematuria | 13 (7%) |
| Proteinuria | 13 (7%) |
| Pyuria | 10 (5%) |
| Rash | 32 (17% |
| Alopecia | 13 (7%) |
| Mucosal Ulcers | 19 (10% |
| Pleurisy | 0 |
| Pericarditis | 0 |
| Low Complement | 72 (39% |
| Increased DNA Binding | 50 (27% |
| Fever | 14 (8%) |
| Thrombocytopenia | 8 (4%) |
| Leukopenia | 22 (12% |

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tion enrolled thus far and to demonstrate the breadth and potential value of the data generated by the new CARRA Registry.

Methods We requested de-identified counts of several fields collected from the case report forms for subjects with SLE. Patients were eligible for enrollment in the new CARRA registry if they were diagnosed with SLE prior to the age of 18 and had either 1) a new diagnosis of SLE or 2) a flare of lupus nephritis within two years prior to the baseline visit. IRB approval was not required for this data request.

Results To date, 184 patients (pts) have been enrolled; 156 (85%) are female. There are 46 black pts, 45 Hispanic pts, 47 white pts, 18 Asian pts and 16 pts were >1 race. Over half the pts have private health insurance (n=95, 52%) and 60 pts (33%) have Medicaid. Autoantibody positivity was prevalent: 175 pts (95%) were ANA positive, 109 (59%) dsDNA positive, and 87 (47%) anti-Smith positive. Positivity for anti-RNP, anti-Ro, anti-La, and APLs ranged from 15% to 51%. At the baseline visit, the mean Systemic Lupus Erythematosus Disease Activity Index (SLEDAI, n=166) score was 5.5 ± 6.3 , median=4 (range 0-37; IQR 0.25-8). The mean Systemic Lupus International Collaborating Clinic Damage Index (SLICC DI, n=150) score was 0.4, median=0 (range 0-7). Approximately one quarter of pts (n=50) were being treated for lupus nephritis at the time of the baseline visit. Manifestations of SLE at the baseline visit were varied (table 1) but serologic disease, mucocutaneous disease and active nephritis were the most prevalent. Conclusions Nearly 200 SLE pts have been enrolled in the new CARRA Registry to date. This is a multi-racial cohort with moderate disease activity and varied disease manifestations. Further enrollment will continue to build a robust data source to study disease course and outcomes in a pediatric SLE inception cohort.

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CS-03 HYDROXYCHLOROQUINE IN LUPUS PREGNANCY: A META-ANALYSIS OF INDIVIDUAL PARTICIPANT DATA

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Background Our current knowledge about how to treat lupus in pregnancy derives from small prospective or retrospective cohorts. The goal of this individual participant meta-analysis was to pool data from multiple prospective cohorts to answer

the clinical question of whether hydroxychloroquine (HCQ) treatment affects pregnancy outcomes.

Methods The literature was searched for prospective cohorts of pregnancies among women with lupus. HCQ use was defined as use any time during pregnancy. Outcomes of interest included fetal loss, preterm birth, high disease, and preeclampsia. Data from each cohort were collected and analyzed individually. Pooled ORs were calculated by random-effect models in Review Manager. Due to multiple pregnancies per patient, one pregnancy was randomly selected per patient. Primary analysis included only women with first trimester visits (6 cohorts). Subgroup analyses were stratified by a history of nephritis, APS, and disease activity at first clinic visit.

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Conclusions Our results suggest that among patients with lupus nephritis, HCQ use may decrease the risk of fetal loss. The heterogeneity of data collection suggests the need for a unified approach to identify larger cohorts of lupus pregnancies.

CS-04 ASSOCIATED FACTORS OF LONG-TERM CARDIAC DYSFUNCTION IN A LONGITUDINAL COHORT OF NEONATAL LUPUS

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Background There are no longitudinal studies regarding the long term cardiac health of children with cardiac manifestations of neonatal lupus (NL). This study was performed to evaluate risk factors for morbidity and provide evidence-based guidance regarding the course of cardiac NL.

Methods Echocardiograms throughout life were evaluated in 240 individuals born with cardiac NL from the Research Registry for Neonatal Lupus: 142 were available from ages 0–1 years, 174 from ages 1–17 years, and 65>17 years. A composite adverse outcome defined as qualitatively decreased left ventricular (LV) function or concurrent use of cardiac

Abstract CS-03 Table 1 Pooled odds ratios for the association of hydroxychloroquine use and pregnancy outcomes

| | Fetal Loss | Preterm Birth | High Disease Activity | Preeclampsia |
|---|------------------|------------------|-----------------------|------------------|
| | OR (95% CI) | OR (95% CI) | OR (95% CI) | OR (95% CI) |
| Overall | 0.50 (0.27-0.94) | 0.95 (0.58–1.55) | 0.69 (0.35–1.39) | 1.19 (0.62-2.30) |
| Lupus Nephritis History | 0.24 (0.07-0.83) | 0.81 (0.35-1.89) | 0.47 (0.21–1.09) | 0.70 (0.24-2.03) |
| No Lupus Nephritis History | 0.70 (0.33-1.46) | 1.04 (0.57-1.89) | 0.98 (0.45-2.17) | 1.36 (0.58-3.16) |
| APS | 0.39 (0.10-1.47) | 0.82 (0.23-2.96) | 1.30 (0.16–10.48) | 0.55 (0.12-2.45) |
| No APS | 0.61 (0.31-1.20) | 0.96 (0.56-1.64) | 0.70 (0.40-1.22) | 1.28 (0.58-2.84) |
| High Disease Activity at 1 st Visit | 0.61 (0.13-2.89) | 1.53 (0.42-5.62) | - | 0.93 (0.12-7.14) |
| No High Disease Activity at 1 st Visit | 0.46 (0.21–1.02) | 0.81 (0.45–1.44) | 0.73 (0.29–1.87) | 1.07 (0.50–2.31) |

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