flare over the last 3 months (31% vs. 26%) among those who had transferred to adult rheumatology vs. those who had not. Those who remained in paediatric care were significantly more likely to have seen a rheumatologist in the past year (94% v. 68%, p = 0.002) and more likely to be taking immunosuppressive medications (89% v. 34%, p < 0.001).

Conclusions Many individuals in this cohort of young adults with cSLE continue with active lupus. In spite of similar disease activity among those who had left paediatric care and those who had not, young adults who had transferred to adult care were significantly less likely to access routine rheumatology care or take immunosuppressive medication, and more likely to encounter difficulty obtaining health insurance coverage. Improving access to adult rheumatology care may be important to prevent poor health outcomes in cSLE.

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CE-42 SLE PRESENTING IN ASSOCIATION WITH HUMORAL IMMUNODEFICIENCY

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Rational Humoral immunodeficiency syndromes including common variable immune deficiency (CVID) are not uncommonly associated with autoimmune features seen in SLE. Studies were undertaken at an academic centre managing both disorders to determine the relative prevalence, clinical and immunologic features, and outcomes of SLE associated with humoral immunodeficiency.

Methods A retrospective review of records identified using an electronic medical record search of diagnosis codes for SLE and hypogammaglobulinemia seen between 2011 and 2016 was undertaken. The clinical and immunologic profile was determined for patients with confirmed or suspected SLE who also had undergone evaluation for humoral immunodeficiency.

Results We identified 40 patients meeting ACR criteria for SLE with inadequate response to pneumococcal vaccine challenge (failure to generate protective antibody titer to ≥5/14 pneumococcal vaccine antigens) and/or low serum IgG levels (<700 mg/dl) not attributable to antecedent immunosuppressive therapy. This comprised 5.0% of our SLE patients meeting ACR SLE criteria in active follow-up. An additional 37 patients with SLE clinical features but not meeting SLE ACR criteria were identified with low serum IgG and/or inadequate vaccine responses. Among the 40 identified patients meeting ACR SLE criteria, serum immunoglobulin levels ranged from 459–760 mg/dl; 36 (90%) had serum IgG levels <700 mg/dl, while 24 (60%) had inadequate response to pneumococcal vaccine challenge, including the four patients with serum IgG > 700 mg/dl. Frequent upper/lower respiratory infections requiring antibiotic treatment (≥3 episodes/year) were reported in 25/40 (63%) patients. SLE features developed 2–26 years (mean = 8.9 years) prior to the recognition of low serum IgG in 27 (68%) patients, whereas initial SLE features were noted concurrently with or 3–4 years following first confirmed low IgG levels in 13 (33%). Arthritis (75%), photosensitivity (81%), malar rash (63%) and mucosal ulcers (56%) were the most prevalent SLE features. Only 9 (23%) of patients had low complement C3 or C4 levels, 6 (15%) had cytopenias, and 2 (5%) had elevated levels of anti-dsDNA. The majority of patients were managed with antimalarials (86%), with 8/40 (20%) also using methotrexate; 18/40 (45%) were on treatment with IVIG and 6/40 (15%) were on treatment with belimumab. Disease activity was low (SLEDAI score ≤ 2) in 37/40 (93%) at the last noted follow-up assessment. In the four patients for whom sera was available for testing, levels of BLyS(BAFF) were elevated relative to those noted in 20 control patients without autoimmunity or immunodeficiency, but less than that noted in 20 SLE patients without noted humoral immunodeficiency.

Conclusion SLE may be a presenting feature of patients with humoral immunodeficiency. Serum immunoglobulin levels and assessment of the response to pneumococcal vaccination for patients with low or low normal serum IgG levels should be included as part of the evaluation for suspected SLE. Favourable outcomes are seen in the context of standard of care treatment for SLE combined with immunoglobulin replacement therapy and/or belimumab.

CE-43 FACTORS ASSOCIATED WITH NEUROPSYCHIATRIC INVOLVEMENT IN 1193 LATIN AMERICAN PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS

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Introduction Neuropsychiatric (NP) manifestations in SLE are a major cause of morbidity, mortality and long term consequences.

Factors related to their occurrence in patients with short disease duration, both early in the course of the disaease and during follow up have not been clearly established.

Purpose To identify disease and non-disease related factors associated with NP manifestations in early SLE.

Methods We included 1193 patients from the GLADEL inception cohort free of NP involvement at cohort entry. We examined the relationship between socio-demographic, clinical and laboratory data as well as disease activity and damage with NP involvement during follow-up. Data were recorded in ARTHROS database. We excluded all the secondary NP manifestations (metabolic, drug induced, infectious, etc). **Statistical methods** The time from cohort entry to first NP manifestation was examined using a Cox proportional hazard regression model. Patients without NP manifestations were censored at last study visit. Independent factors associated with NP involvement were identified using a multivariable Cox regression model. Variables included in the final model were selected using a backward selection algorithm with α-level to stay in the model set to 0.05. Results are summarised as hazard ratios with 95% confidence intervals.

Results During a median follow-up time of 52 months, 238 (20 %) patients had NP involvement. The cumulative incidence estimate of NP involvement at 1, 3 and 5 years was 8.3%, 17.8% and 24.7%, respectively. In the univariable analysis some variables like ethnic origin were found to be more frequent in mestizos as compared to patients in the other ethnic groups. Factors