however the numbers evaluated were small and the patients studied were young. This study was initiated to ascertain the current prevalence of autoimmune disease in NL children and their unaffected siblings, and to evaluate whether fetal or maternal factors associated with the development of future autoimmunity.

Materials and methods A retrospective cohort of family members from the RRNL were contacted to evaluate for autoimmune disease. Follow-up questionnaires were completed which included 35 items describing symptoms and diagnoses associated with autoimmunity in 138 cardiac NL children, 74 cutaneous NL children, and 134 unaffected siblings. Medical records were obtained and evaluated from the patient's physicians to confirm diagnoses. Maternal diagnosis of systemic lupus and/or Sjogren's syndrome and fetal cardiac disease severity based on a previously described severity score were associated with postnatal autoimmune diseases using chi square and Mann-Whitney analyses.

Results Seventeen (8.0%) of NL affected children developed an autoimmune disease at the time of follow up (mean age 11.6 ±9.0 years). These included 3 patients with SLE, 1 with JIA, 3 with thyroid disease, 5 with psoriasis, 1 with IBD, 1 with uveitis, 1 with UAS and 2 with type 1 DM. Six (4.5%) unaffected siblings developed an autoimmune disease (mean age 10.6±7.1 years), which included 1 with JIA, 1 with Sarcoidosis/ITP, 1 with Myasthenia Gravis/Celiac disease, 1 with psoriasis, 1 with UAS and 1 with type 1 DM,. There was a significant association of between having an autoimmune disease and having advanced heart block (11.0% vs. 4.2%, p = 0.03) and a trend towards an association with cardiac NL disease severity score (4.43±4.89 vs. 2.58±4.24, p = 0.06). Mother's diagnosis of SLE or Sjogren's did not associate with the children's development of autoimmune disease (p = 0.828).

Conclusions Fetuses that develop advanced congenital heart block as a manifestation of NL may be at greater risk for developing autoimmune diseases later in life. This could potentially relate to a genetic component that makes a Ro exposed fetus both more prone to inflammatory effects of passive immunity and predisposes to future autoimmunity, independent of the mother's rheumatic disease status.

CE-41 DISEASE ACTIVITY AND HEALTH CARE UTILISATION AMONG YOUNG ADULTS WITH CHILDHOOD-ONSET LUPUS TRANSITIONING TO ADULT CARE: DATA FROM THE PAEDIATRIC LUPUS OUTCOMES STUDY

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Background Individuals with childhood-onset systemic lupus erythematosus (cSLE) must transfer from paediatric- to adult-oriented health care as they enter adulthood. However, few studies have assessed outcomes of the transition from paediatric to adult care in cSLE. The goal of this study is to examine disease activity and health care utilisation among young adults with cSLE who are undergoing or have recently completed the transition to adult care.

Materials and methods Data derive from the baseline interview of the Paediatric Lupus Outcomes Study, an annual longitudinal telephone survey of 91 diverse English- and Spanish-speaking participants age 18–30 with confirmed cSLE (age of onset <18 years). Subjects were recruited from paediatric and adult rheumatology clinics; diagnosis of cSLE was confirmed by chart review.

Abstract CE-41 Table 1 Demographics, disease characteristics and healthcare utilisation among young adults with cSLE in paediatric rheumatology care vs. young adults with cSLE who have transferred to adult care

	Paediatric rheumatology care (n = 47) *N (%) or Mean (SD)*	Adult rheumatology care (n = 38) *N (%) or Mean (SD)*	*P*
Demographics			
Age (years)	19 (1)	24 (3)	<0.001
Female	41 (87)	36 (95)	NS
Non-white ethnicity	35 (74)	29 (11)	<0.001
Age at diagnosis (years)	13 (3)	13 (2)	NS
Disease Characteristics			
Renal biopsy ever	25 (54)	18 (47)	NS
Cyclophosphamide use ever	7 (15)	19 (50)	<0.001
Disease activity (SLAQ)*	11 (11)	9 (10)	NS
Self-reported flare in the past 3 months	14 (31)	10 (26)	NS
Health Care Utilisation			
Current medications			
Plaquenil	41 (89)	24 (63)	0.005
Steroid	36 (76)	7 (18)	<0.001
DMARD	33 (70)	12 (32)	<0.001
Biologic	0 (0)	0 (0)	NS
Insurance coverage	45 (96)	34 (89)	NS
Difficulty obtaining insurance	5 (11)	13 (34)	0.008
Rheumatology visit in the past year	44 (94)	26 (68)	0.002
General MD visit in the past year	23 (51)	15 (41)	NS
ED visit in the past year	18 (38)	9 (24)	NS
Hospitalization in the past year	14 (30)	9 (24)	NS

*SLAQ = Systemic Lupus Activity Questionnaire (0–47)

To define a cohort undergoing transition from paediatric to adult care, we included respondents who received care from a paediatric rheumatologist currently or in the past (N = 85). We assessed disease activity according to the Systemic Lupus Activity Questionnaire (SLAQ), self-reported SLE flare in the past 3 months, current immunosuppressive medication use (any steroid, DMARD or biologic medication), current health insurance coverage, and health care utilisation over the past year. Bivariate analyses were used to compare individuals cared for by adult rheumatologists to those who continue in paediatric care.

Results Mean baseline age was 21 ± 3 years, and mean age at diagnosis was 13 ± 3 years. Ethnicities included White (48%), Black (5%), Asian (20%), Latino (24%), multi-ethnic (5%) and other (6%). 38 respondents (45%) had transferred care out of paediatric rheumatology (Table 1). Most respondents were currently insured (93%), however those who had transferred were more likely to report difficulty obtaining insurance (34% v. 11%, p = 0.008). 32% had visited an emergency department and 27% had received inpatient care in the past year, with similar rates in adult and paediatric care groups. There was no difference in disease activity (SLAQ score 9 v. 11) or likelihood of self-reported

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