modified SELENA-SLEDAI Flare Index. Safety and PK were assessed. Analyses were performed on the intent-to-treat population. The study was not powered to test for differences between groups; p-values were not calculated.

Results: 93 patients were included (BEL, n=53; PBO, n=40). Groups (BEL vs PBO) were balanced at baseline for age (mean [standard deviation] 13.5 [2.59] vs 14.8 [2.17] years, respectively) and SELENA-SLEDAI score (10.3 [3.34] vs 10.4 [3.63], respectively). Compared with PBO, there were more SRI4 responders (including all 3 components of SRI4), and PRINTO/ACR 30 and 50 responders in the BEL group (**Figure**). Likewise, more BEL than PBO recipients had sustained improvement of SRI and patient well-being (ParentGA) (**Figure**). Changes in cSLE core response variables are shown in the **Table**. Severe flares were 62% less frequent with BEL vs PBO (hazard ratio 0.38 [95% CI 0.18, 0.82]). PK: BEL exposures in cSLE were similar to adult SLE studies. 9/53 (17%) BEL patients had ≥ 1 serious adverse event vs 14/40 (35%) PBO patients. One PBO patient died of acute pancreatitis.

Conclusion: The benefit:risk profile of BEL IV plus SoC in cSLE is generally consistent with BEL in adult SLE. The 10 mg/kg IV dose used in adults may be an appropriate dose in cSLE.

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I perform consultancy activities on behalf of the Gaslini Institute for the companies listed below:

AbbVie, Biogen, Boehringer Ingelheim, Bristol-Myers Squibb, EMD Serono, Janssen, Novartis, Pfizer, R-Pharm.

The money received for these activities are directly transferred to the Gaslini Institute's bank account. Before March 2016, I was the head of the Pediatric Rheumatology Department at the G. Gaslini Hospital, where the PRINTO Coordinating Centre is located. For the coordination activity of the PRINTO network, the Gaslini Hospital received contributions from the industries listed in this section. This money has been reinvested for the research activities of the hospital in fully independent manners besides any commitment with third parties., Daniel J Lovell Consultant for: Consulting fees and/or honoraria from Astra Zeneca, Wyeth Pharma, Amgen, Abbott, Pfizer, F. Hoffmann-La Roche, Novartis, UBC, Takeda, GSK, Boehringer, and Celgene, Hermine Brunner Grant/research support from: Bristol-Myers Squibb, Pfizer, Consultant for: Pfizer, Bristol-Myers Squibb, Janssen, Novartis, Lilly, Roche, GlaxoSmithKline, Sanofi, Speakers bureau: Novartis, Roche

Figure. Primary and major secondary endpoints at Week 52 (N=93)

*At least 50% improvement in 2 of 5 cSLE core response variables, with no more than 1 of the remaining worsening by more than 30%; †at least 30% improvement in 3 of 5 cSLE core response variables, with no more than 1 of the remaining worsening by more than 30%; ‡Weeks 44–52; §Defined as increase of <0.30 points from baseline

ACR, American College of Rheumatology; BILAG, British Isles Lupus Activity Group; CI, confidence interval; OR, odds ratio; ParentGA, parent global assessment; PGA, Physician's Global Assessment; PRINTO, Pediatric Rheumatology International Trials Organisation; SELENA-SLEDAI, Safety of Estrogens in Systemic Lupus Erythematosus-National Assessment Trial-Systemic Lupus Erythematosus Disease Activity Index; SRI, SLE Responder Index

Table. Percent change from baseline in cSLE core response variables at Week 52

PRINTO/ACR variable	Placebo (n=40)	Belimumab (n=53)
ParentGA, median (range)	-23.6 (-95, 600)	-53.9 (-100, 900)
n	38	47
PGA, median (range)	-47.9 (-100, 61)	-76.1 (-100, 117)
n	40	53
SELENA-SLEDAI, median (range)	-60.0 (-100, 58)	-60.0 (-100, 150)
n	39	53
PedQL (physical functioning), median (range)	12.5 (-55, 575)	10.5 (-100, 280)
n	40	53
Proteinuria, median (range)	7.1 (-91, 570)	-2.1 (-85, 1682)
n	39	53

PedQL, Pediatric Quality of Life Inventory; PGA, Physician's Global Assessment

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FRI0182 FREQUENCY OF HYDROXYCHLOROQUINE RETINOPATHY IN THE HOPKINS LUPUS COHORT

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Background: The Kaiser-Permanente study projected hydroxychloroquine (HCQ) retinopathy rates of 40% after 20 years of use (1).

Objectives: We have prospectively followed SLE patients in the Hopkins lupus cohort to compare.

Methods: Patients in the Hopkins cohort are seen quarterly for assessment of disease activity and lupus complications. Yearly ophthalmology examinations are requested. Patients, if insurance allows, are referred to the Wilmer Retina group. Four tests are performed: OCT, ERG, MP-1 and FAF.

Results: Patients had a variety of retinal testing done, with optical coherence testing most frequent. Table 1 shows the concordance of a test abnormality with the retina expert opinion.

Table 1. – Performance of retinal imaging modalities relative to expert diagnosis

Test	Retinopathy N (%)	No Retinopathy N (%)
Ocular Coherence Tomography (OCT)		
Abnormal	25 (93%)	133 (16%)
Normal	2 (7%)	721 (84%)
Total	27	854
Electroretinogram (ERG)		
Abnormal	15 (100%)	195 (49%)
Normal	0 (0%)	201 (51%)
Total	15	396
Microperimetry (MP1)		
Abnormal	17 (100%)	144 (30%)
Normal	0 (0%)	331 (70%)
Total	17	475
Fundal Autofluorescence (FAF)		
Abnormal	19 (83%)	165 (24%)
Normal	4 (17%)	536 (76%)
Total	23	701

The concordance of the test abnormality with the retina expert opinion showed the following sensitivity and specificity, respectively: OCT 93%, 84%; ERG 100%, 51%; MP1 100%, 70%; and FAF 83%, 76%.

The frequency of retinopathy increased with years of HCQ use [number of retinopathies per total patients (percent frequency)]: 5 years or less, 1/103 (0.97%); 6-10 years, 2/109 (1.83%); 11-15 years, 3/91 (3.30%); 16-20 years, 11/96 (11.46%); and 21 or more years, 6/75 (8.00%).

Conclusion: In agreement with the American Academy of Ophthalmology, OCT appears to be the optimum test for yearly monitoring (2). The frequency of retinopathy was much lower in our prospective study than estimated by the Kaiser-Permanente study. Our data also show the need for ophthalmologists with retinopathy expertise to interpret retina testing, as screening tests are frequently abnormal due to causes other than HCQ retinopathy. Stopping HCQ based on an abnormal test without confirmation from a retinopathy expert could needlessly deprive an SLE patient of an important medication.

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FRI0183

STUDY DESIGN FOR A MULTICENTER, RANDOMIZED, DOUBLE-BLIND, PLACEBO-CONTROLLED STUDY TO ASSESS THE EFFICACY AND SAFETY OF REPOSITORY CORTICOTROPIN INJECTION IN PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS DESPITE MODERATE-DOSE CORTICOSTEROIDS

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Background: Repository corticotropin injection (RCI) is a complex mixture containing purified porcine pituitary adrenocorticotrophic hormone (ACTH)-

analogue with potential anti-inflammatory and immunomodulatory effects through the melanocortin receptors (MC1R, MC3R-MC5R) and endogenous steroid production. RCI is approved by the US FDA for the treatment of acute exacerbations (ie, disease flares) or as maintenance therapy in patients with systemic lupus erythematosus (SLE). In a pilot study, RCI was well tolerated with improvements in disease activity for lupus patients treated with RCI compared to those who received placebo (Furie et al, 2016).

Objectives: To discuss the design, demographics, and baseline characteristics of a clinical trial evaluating the role of RCI in the treatment of patients with inadequately controlled SLE disease activity.

Methods: This is a multicenter, double-blind, randomized, placebo-controlled, 24-week clinical trial aiming to determine the effect of RCI in reducing disease activity for patients with persistently active SLE despite moderate-dose corticosteroids (CSs). Patients are required to have SLE (≥ 4 of 11 ACR criteria from 1997) and active disease (moderate to severe rash and/or arthritis) despite stable dose CSs ≥ 4 weeks prior to screening (7.5 - 30 mg/day of prednisone or equivalent). Patients are treated with RCI 1 mL (80 U) subcutaneously (SC) or placebo 1 mL, every other day for 4 weeks followed by twice per week for 20 weeks. Randomization is stratified by study site location and prednisone or equivalent dose (≤ 20 and > 20 mg/day). Patients remain on stable doses of CSs through Week 16, with tapering encouraged between Weeks 16 and 24. Efficacy is evaluated using the SLE Responder Index-4 (SRI-4) and changes from baseline in SLE Disease Activity Index-2000 (SLEDAI-2K), British Isles Lupus Assessment Group-2004 (BILAG-2004) scores, and Physician's Global Assessment (PGA). The primary efficacy endpoint is the proportion of SRI-4 responders at Week 16. Secondary and exploratory endpoints include changes in disease activity scores over time, prednisone dose, and biomarkers (ie, complements, autoantibodies, inflammatory, bone turnover).

Results: As of 14 December 2018, 124 patients comprised the intent-to-treat (ITT) population. Target enrollment is 270 patients, of which 162 patients are expected to be randomized at approximately 60 sites globally. Demographic and baseline characteristics (Table) were collected, including circulating lymphocyte profiles (CD19+ B Cells, CD3+ total T cells, and CD4 Treg cells), which are consistent with the pilot study of RCI (Furie et al, 2016).

Characteristic	Patients (ITT Population, N=124)
Age, n, mean (SD)	40.6 (12.5)
Female, n (%)	114 (91.9)
Race, n (%)	
Caucasian	45 (36.3)
African American	15 (12.1)
American Indian or Alaska Native	25 (20.2)
Other	39 (31.3)
Ethnicity, n (%)	
Hispanic or Latino	80 (68.5)
Weight, kg, mean (SD)	73.3 (19.2)
SLEDAI-2K Total Score, mean (SD)	9.8 (3.0)
BILAG Total Score, mean (SD)	88.4 (6.6)
Physician's Global Assessment (100 mm), mean (SD)	62.1 (12.3)
Prednisone or Equivalent Daily Dosage, mg, mean (SD)	11.5 (5.2)
Anti-malarials, n (%)	71 (57.3)
Immunosuppressants, n (%)	73 (58.9)
Bone Turnover Markers	
PTH, ng/mL, mean (SD)	44.0 (23.4)
CTX-L, ng/mL, mean (SD)	0.347 (0.175)

Conclusion: This on-going study assesses the efficacy and safety of RCI for the treatment of refractory SLE in a racially and ethnically diverse population. The rapid onset of action of RCI allows for a 16-week primary endpoint. Patients enrolled in this trial have moderate or high disease activity (as shown by the mean SLEDAI-2K, BILAG, and PGA) and are likely to provide valuable data on the role of RCI in refractory lupus. Bone turnover markers will further define the safety profile of RCI in lupus.

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