**Conclusion:** Serious infections are a major cause of mortality and damage accrual in SLE. Constitutional symptoms, gastrointestinal involvement, current and cumulative steroid dose and cyclophosphamide use predict serious infections. TB prophylaxis in patients with SLE should be considered in endemic areas, especially when using high-dose steroid therapy.

**Table 1. Cox proportional hazard models for predictors of infection**

| Covariates | Hazard ratio | 95% CI |
|---|---|---|
| **Fever*** | **8.51** | **1.17-61.45** |
| Serositis | 0.81 | 0.41-1.59 |
| **Gastrointestinal involvement*** | **4.76** | **1.94-19.94** |
| Major organ manifestation | 1.07 | 0.49-2.31 |
| SLEDAI-2K | 0.99 | 0.95-1.05 |
| **Daily steroid dose (/10mg)*** | **1.36** | **1.14-1.62** |
| **Mean cumulative steroid dose*** | **1.004** | **1.002-1.005** |
| Albumin | 1.01 | 0.66-1.55 |
| Absolute lymphocyte count | 0.99 | 0.99-1 |
| **Cyclophosphamide use*** | **2.22** | **1.11-4.46** |

*-significant baseline predictors of serious infection on follow up. Major organ manifestation referes to presence of nephritis or neuropsychiatric lupus.

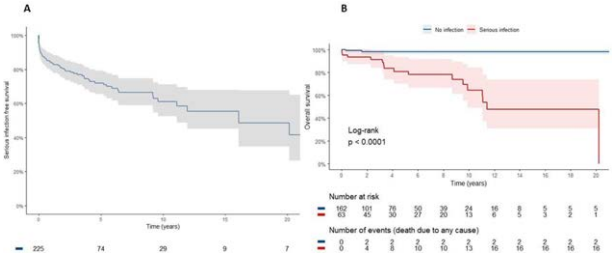


**Figure 1.** A. The Kaplan meier survival curve depicts time to first serious infection. B. The Kaplan meier survival curve shows the overall survival difference between those with any serious infection ever compared to those with no serious infection in the disease course.

**REFERENCES: NIL.**
**Acknowledgements: NIL.**
**Disclosure of Interests: None Declared.**
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## POS0138 EFFICACY AND SAFETY OF BELIMUMAB IN ADULTS WITH CHILDHOOD-ONSET SYSTEMIC LUPUS ERYTHEMATOSUS: PRELIMINARY RESULTS FROM A PROSPECTIVE OBSERVATIONAL STUDY

**Keywords:** Outcome measures, bDMARD, Systemic lupus erythematosus

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**Background:** Childhood-onset Systemic Lupus Erythematosus (cSLE) is a rare autoimmune disease with multi-system manifestations, more severe disease course and higher frequency of morbidity than adult-onset SLE. Belimumab is the first treatment for cSLE approved for children ≥5 years of age.
**Objectives:** To investigate the efficacy and safety of Belimumab in adult patients with cSLE.
**Methods:** A prospective observational (non-interventional) study, involving adult patients with cSLE was conducted. During the 9-yearstudy period (01/2015 to 12/2022), adults with **a** cSLE diagnosis and Belimumab receivers (by intravenous or subcutaneous administration) for >12 consecutive months were enrolled. All patients met the revised 1997 American College of Rheumatology (ACR) or 2012 Systemic Lupus International Collaborating Clinics (SLICC) classification criteria and were followed at regular intervals (up to 6 months) in the **T**ransition **R**heumatology **O**utpatient **C**linic. SLE activity was defined according to SLEDAI-2K and the SELENA-SLEDAI Physician Global Assessment (PGA), scale 0–3 [1]. At the last follow-up visit, response to therapy was assessed by Lupus Low Disease Activity State (LLDAS), remission state and SLE Responder Index (SRI4). LLDAS was defined as: i) SLEDAI-2K ≤4, with no activity in major systems ii) no new lupus disease activity compared to previous evaluation iii) a SELENA-SLEDAI PGA ≤1 iv) current prednisolone (or equivalent) dose ≤7.5 mg daily and v) standard maintenance doses of immunosuppressive drugs. [2]. Remission was defined as i) clinical SLEDAI-2K=0 ii) dose of prednisone ≤5 mg/day according to the DORIS definition [3]. SRI4 was defined as i) ≥4-point reduction from baseline in SELENA-SLEDAI score, ii) no worsening in PGA and iii) no new BILAG A organ domain score or 2 new BILAG B organ domain scores compared with baseline [4].
**Results:** A total of 15 patients (14 females) were enrolled in the study. At baseline, the patients' current mean (SD) age was 25.4 (7) years and the interval from disease onset to first Belimumab administration 12.4 (7.3) years. Half of the patients had a history of lupus nephritis and 42.8% were aPL positive. All patients were under hydroxychloroquine treatment and 80% of them were additionally receiving a second immunosuppressive agent (Methotrexate, MMF, Azathioprine). Glucocorticoids (GCs) were concomitantly administered in 13 (86.6%) patients in a dose of ≥7.5mg in 57.1% of them. The patients' SLEDAI-2K mean (SD) score before Belimumab initiation was 12.1 (2.3), indicating high disease activity. The mean (SD) duration of Belimumab administration was 36.2 (1.9) months. At the last follow up visit, ongoing therapy with Belimumab was recorded in 66.7% of the cohort. SLEDAI-2K mean (SD) score was reduced to 3.5 (2.3), 1 patient (6.6 %) was in remission, 9 (60%) patients had mild and none high disease activity. The majority (78.5%) met the LLDAS definition and 78.5% were SRI4 Responders. Half of the patients (53.8%) achieved lower doses or discontinuation of GCs and 26.7% accomplished a reduction of the immunosuppressant's dose. Reasons for Belimumab discontinuation included pregnancy issues, infection and low adherence (3, 1, 1 patient respectively). None of the patients experienced a serious adverse event.
**Conclusion:** In this cohort of adult patients with cSLE, Belimumab was well-tolerated and effectively reduced disease activity allowing the downstream of GCs's dose.
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**Acknowledgements: NIL.**
**Disclosure of Interests: None Declared.**
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## POS0139 DIFFUSE JUVENILE SYSTEMIC SCLEROSIS PATIENTS SHOW DISTINCT ORGAN INVOLVEMENT, ANTIBODY PATTERN AND HAVE SIGNIFICANTLY MORE SEVERE DISEASE IN THE LARGEST JSSC COHORT OF THE WORLD. RESULTS FROM THE JUVENILE SCLERODERMA INCEPTION COHORT

**Keywords:** Systemic sclerosis

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**Background:** Juvenile systemic sclerosis (jSSc) is an orphan disease with a prevalence of 3 in 1 000 000 children. In adult patients there are significant differences between the clinical presentation of diffuse and limited subtypes. We reviewed clinical differences in presentation of subtypes in patients in the juvenile systemic scleroderma inception cohort (jSScC).
**Objectives:** To study the clinical presentation of jSSc patients with diffuse (djSSc) and limited (ljSSc) subtypes.
**Methods:** We reviewed the baseline clinical characteristics of the patients, who were recruited to the jSScC till December 2022. jSScC is a prospective cohort of jSSc patients, who developed the first non-Raynaud´s symptom before the age of 16 years and are under the age of 18 years at the time of inclusion.
**Results:** The JSScC included 232 patients, 68% (n=159) had diffuse subtype. The median age at onset of Raynaud phenomenon was 10.4 years (7.3-12.9), at the first non-Raynaud symptom 10.9 years (7.3-13.0) and median disease duration 2.5 years (1.0-4.6). The female/male ratio was significantly lower in the djSSc subtype

(3:1 versus 5:1, p<0.001). Antibody profile was similar, with the exception of a significantly higher number of anticentromere positive patients in the ljSSc. Decreased FVC<80% was found in approximately 30% and decreased DLCO<80% was found in around 40% in both subtypes. Abnormal HRCT findings were found in 44% of patients. Pulmonary hypertension assessed by ultrasound occurred in approximately 5% in both groups and gastrointestinal involvement in 43% of djSSc and 36% in ljSSc (p=0.303). Patients with djSSc had significantly higher modified Rodnan Skin Score, more frequently sclerodactyly, a history of digital ulceration active ulceration, telangiectasia, a decreased Body Mass Index z score ≤ -2 and decreased joint range of motion. Patients with ljSSc had significantly higher rate of cardiac involvement. Regarding patient related outcomes assessed by VAS 0-100 djSSc patients had more severe disease also physician related outcome assessed by VAS 0-100 were significantly higher in djSSc (see Table 1).

**Table 1. Comparison of subtypes at time of inclusion in the cohort**

| | Whole Group N=232 | Diffuse Subtype N=159 | Limited Subtype N=73 | P value |
|---|---|---|---|---|
| Anticentromere | 5% (7/156) | 2% (2/106) | 10% (5/50) | 0.022 |
| MRSS, median (IQR) | 10 (4 – 20) | 16 (8 - 27) | 4 (0 – 8) | 0.001 |
| Gottron Papules | 26% (59/228) | 31% (48/155) | 15% (11/73) | 0.011 |
| Sclerodactyly | 75% (165/219) | 85% (127/150) | 55% (38/69) | <0.001 |
| Telangiectasia | 37% (77/209) | 44%(62/141) | 22% (15/68) | 0.002 |
| History of ulceration | 52% (119/229) | 62% (98/158) | 30% (21/71) | <0.001 |
| Active ulceration | 17% (39/229) | 21% (33/158) | 8% (6/71) | 0.021 |
| Only Cardiac involvement | 5% (12/232) | 3% (4/159) | 11% (8/73) | 0.007 |
| BMI < - 2 z score | 15% (33/217) | 20% (29/148) | 6% (4/69) | 0.008 |
| Joints with decreased range | 59% (136/231) | 64% (101/158) | 48% (35/73) | 0.022 |
| **Physician Reported (Median, IQR)** | | | | |
| Physician global disease activity | 30 (20 – 45) n=197 | 35 (20– 50) n=138 | 20 (10 – 30) n=59 | 0.001 |
| Physician global disease damage | 30 (15 – 40) n=195 | 30 (20 – 45) n=138 | 20 (5 – 30) n=57 | 0.004 |
| Physician ulceration activity | 0 (0 – 16) n=216 | 5 (0 – 20) n=154 | 0 (0 – 0) n=62 | 0.018 |
| **Patient Reported (Median, IQR)** | | | | |
| Patient global disease activity | 40 (20 – 50) n=178 | 40 (20 – 50) n=129 | 30 (15 – 55) n=49 | 0.024 |
| Patient global disease damage | 30 (15 – 60) n=177 | 40 (20 – 60) n=128 | 25 (5 – 55) n=49 | 0.001 |
| Patient Raynaud activity | 30 (10 – 60) n=202 | 30 (10 – 60) n=145 | 15 (0 – 55) n=57 | 0.001 |
| Patient ulceration activity | 0 (0 – 30) n=203 | 10 (0 – 30) n=145 | 0 (0 – 20) n=58 | 0.001 |

**Conclusion:** In the largest jSSc cohort in the world, djSSc patients have a significantly more severe disease. Patients and physician related outcomes were significantly more severe in djSSc group. Interestingly, we found no differences regarding interstitial lung disease, pulmonary hypertension or gastrointestinal involvement, although the number of patients with decreased BMI ≤ -2 z score was significantly higher in the djSSc patients.

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**Acknowledgements: NIL.**

**Disclosure of Interests: None Declared.**

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POS0140

## PATIENT AND PHYSICIAN REPORTED OUTCOMES OF JUVENILE SYSTEMIC SCLEROSIS PATIENTS SIGNIFICANTLY IMPROVE OVER 24 MONTHS OBSERVATION PERIOD IN THE JUVENILE SYSTEMIC SCLERODERMA INCEPTION COHORT

**Keywords:** Systemic sclerosis

I. Foeldvari[1], J. Klotsche[2], O. Kasapcopur[3], A. Adrovic[3], K. Torok[3], M. T. Terreri[3], A. P. Sakamoto[3], J. Antón[3], B. Feldman[3], R. Khubchandani[3], M. Kostik[3], T. Lehman[3], E. Marrani[3], M. Katsikas[3], D. Nemcova[3], M. J. Santos[3], F. R. Sztajnbok[3], S. Appenzeller[3], C. Battagliotti[3], L. Berntson[3], J. Brunner[3], L. Harel[3], G. Horneff[3], S. Johnson[3], T. Kallinich[3], K. Minden[3], M. Moll[3], F. Nuruzzaman[3], A. Patwardhan[3], D. Schonenberg[3], N. Helmus[1]. [1]*Hamburg Centre for Pediatric and Adolescence Rheumatology, An der Schön Klinik Hamburg Eilbek, Hamburg, Germany*; [2]*German Rheumatism Research Center, German Rheumatism Research Center, Berlin, Germany*; [3]*jSSc collaborative group, Hamburg Centre for Pediatric and Adolescence Rheumatology, Hamburg, Germany*

**Background:** Juvenile systemic sclerosis (jSSc) is an orphan disease with a prevalence of 3 in 1 000 000 children. The Juvenile Systemic Scleroderma Inception cohort (jSScC) is the largest cohort of jSSc patients in the world. The jSScC collects data prospectively in jSSc, allowing the evaluation of the development of organ involvement and patient and physician reported outcomes in jSSc over time.

**Objectives:** To review the changes in the clinical characteristics, and patient and physician reported outcomes, over a 24 month observation period from the time of inclusion into the cohort.

**Methods:** The jSScC cohort enrolls jSSc patients who developed the first non-Raynaud´s symptom before the age of 16 years and are under the age of 18 years at the time of inclusion. We reviewed jSScC patient clinical data and patient and physician reported outcomes, who had 24 months follow up from the time of inclusion until 1st of December 2022.

**Results:** We extracted data from 90 patients, 77% of them had the diffuse subtype. The female/male ratio was 3.5:1. Median age of onset of Raynaud´s was 9.4 years and the median age of onset of non-Raynaud´s was 10.0 years. Eighty-nine percent of the patients were treated with disease modifying anti-rheumatic drugs (DMARDs) at time of inclusion in the cohort (T0) and 96% after 24 months (T24). Median disease duration was 2.4 years at T0. No patient died during the follow up. Antibody profile stayed unchanged. Only 3 clinical parameters changed and all improved significantly, the median modified Rodnan skin score improved from 11 to 8 (p=0.021), number of patients with joints with pain on motion decreased from 21% to 10% (p=0.04) and the number of patients with muscle weakness decreased from 13% to 4% (p=0.03). All other organ involvement did not show any statistically significant change from T0 to T24. All collected patient reported outcomes improved significantly from T0 to T24: the patient reported disease activity by VAS 0-100 from 40 to 20 (p=0.001), the patient reported disease damage by VAS 0-100 from 35 to 20 (p=0.027), patient reported ulceration activity by VAS 0-100) from 8 to 0 (p=0.001) and the patient reported Raynaud activity by VAS 0-100 from 20 to 10 (p=0.002). Two of the three physician reported outcomes improved significantly, the physician global disease activity by VAS 0-100 from 30 to 20 (p=0.001) and physician reported ulceration activity by VAS 0-100 from 5 to 0 (p=0.017). (Table 1.)

**Table 1. Items with significant improvement over 24 months observation period in the cohort**

| | 0 month N=90 | 24-month follow-up N=90 | P Value |
|---|---|---|---|
| MRSS, median (IQR) | 11 (5 – 20) | 8 (2 – 20) | 0.021 |
| Presence of joints with pain on motion | 21% (19/89) | 10% (9/89) | 0.040 |
| Muscle Weakness | 13% (10/75) | 4% (3/81) | 0.030 |
| Physician global disease activity | 30 (20 – 45) n=71 | 20 (10 – 33) n=83 | 0.001 |
| Physician ulceration activity | 5 (0 – 19) n=86 | 0 (0 – 10) n=89 | 0.017 |
| Patient global disease activity | 40 (20 – 54) n=66 | 20 (10 – 30) n=76 | 0.001 |
| Patient global disease damage | 35 (20 – 60) n=66 | 20 (10 – 35) n=76 | 0.027 |
| Patient Raynaud activity | 20 (0 – 50) n=83 | 10 (0 – 26) n=84 | 0.002 |
| Patient ulceration activity | 8 (0 – 28) n=84 | 0 (0 – 10) n=84 | 0.001 |

**Conclusion:** Skin and musculoskeletal clinical features improved over 24 months, with almost all patients on DMARDs, supporting the response of these features to therapy. It was promising that internal organ involvement, like cardiac, lung and gastrointestinal, did not significantly worsen or increase. The most striking observation is the positive direction and improvement all patients and two of the three physician reported outcome measures over 24 months in this large international cohort.

**REFERENCES: NIL.**

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**Disclosure of Interests: None Declared.**

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POS0141

## APPLICATION OF CRISS SCORE, REVISED CRISS SCORE AND RCID SCORE IN PATIENTS WITH JUVENILE SYSTEMIC SCLEROSIS

**Keywords:** Systemic sclerosis

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