Background and aims To assess outcomes of childhood systemic lupus erythematosus (cSLE) in three different age groups evaluated at last visit: group A early-onset disease (<6 years), group B school age (6–12 years) and group C adolescence (12–18 years).

Methods Observational cohort study of 10 Paediatric Rheumatology centres, including 847 cSLE patients.

Results Group A had 39 (4%), B 395 (47%) and C 413 (49%). Median disease duration was higher in group A compared to groups B and C (3.3 vs. 2.3 vs. 2.3 years, p<0.0001). The median SLICC/ACR-DI (0–9) vs. 0 (0–6) vs. 0 (0–7), p=0.065) was comparable in all groups. Further analysis of organ/system damage revealed that frequencies of neuropsychiatric (21% vs. 10% vs. 7%, p=0.007), skin (10% vs. 1% vs. 3%, p=0.002) and peripheral vascular involvements (5% vs. 3% vs. 0.3%, p=0.008) were more frequent in group A compared to B and C. Frequencies of severe cumulative lupus manifestations such as nephritis, thrombocytopenia and autoimmune hemolytic anaemia were similar in all groups (p>0.05). Mortality rate was higher in group A compared to groups B and C (15% vs. 10% vs. 6%, p=0.028). Out of 69 deaths, 33/69 (48%) occurred within the first two years after diagnosis. Infections accounted for 54/69 (78%) of the deaths and 38/54 (70%) had concomitant disease activity.

Conclusions: This large multicenter study provided evidence that early-onset cSLE group had distinct outcomes, with higher mortality rate and neuropsychiatric/vascular/skin organ damages in spite of comparable frequencies of severe cumulative lupus manifestations. We also identified that overall death in cSLE patients was an early event mainly attributed to infection associated with disease activity.