Clinical Characteristics of Pre-pubescent Patients with Systemic Lupus Erythematosus (SLE)

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Background:
Systemic Lupus Erythematosus (SLE) is a chronic autoimmune condition that affects multiple organ systems, including cutaneous, renal, neurological, and hematological disorders. Pediatric SLE (pSLE) has a prevalence of 3.3-8.8 cases per 100,000 children. Children with SLE (cSLE) have higher disease severity than adults, but the evidence on cSLE is mostly from pubertal patients, and the reasons for the variation on presentation is unclear. There is limited data on prepubertal SLE, as SLE rarely affects pre-pubescent patients. This study aims to outline the clinical characteristics, disease activity and organ damage of pre-pubescent patients with SLE.

Methods:
A retrospective study was performed on prospectively collected multi-center data from 2005-2015. Patient data, including race, ethnicity, sex, age of presentation and diagnosis, Systemic Lupus Erythematosus Disease Activity Index (SLEDAI) score, and Systemic Lupus International Collaborating Clinics (SLICC) damage index was collected using REDcap.

Results:
This study included 43 pre-pubescent SLE patients, with an average age at diagnosis of 9.7 years and 10.7 years at the study's start. Organ systems with the highest prevalence rates were musculoskeletal (Arthritis – 69.77%) and cutaneous (Malar rash – 58.14%). The organ systems with the highest involvement frequency, according to the SLICC damage index, were skin (alopecia - 9.3%), ocular (retinal changes/optic atrophy/cataracts - 9.3%), gastrointestinal (pancreatic insufficiency – 4.65%), neuropsychiatric (cognitive impairment – 4.65%), and renal (reduced glomerular filtration rate – 4.65%). Throughout the study, all patients had SLEDAI scores ranging from no activity to high activity, with 55.8% having mild activity, 27.9% having moderate activity, and 14% having high activity.

Conclusion:
Similar to SLE studies on pediatric populations, our study found that cutaneous, gastrointestinal, and renal manifestations were common. However, we found a high prevalence of damage in prepubertal patients, in particular ocular and cognitive impairment. This highlights the need to prioritize ocular exams and cognitive assessments in patients with prepubertal SLE.