THE CHILL-NL STUDY:
VERY LONG TERM OUTCOMES OF CHILDHOOD-ONSET SLE - DISEASE ACTIVITY, DISEASE DAMAGE AND QUALITY OF LIFE

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Introduction

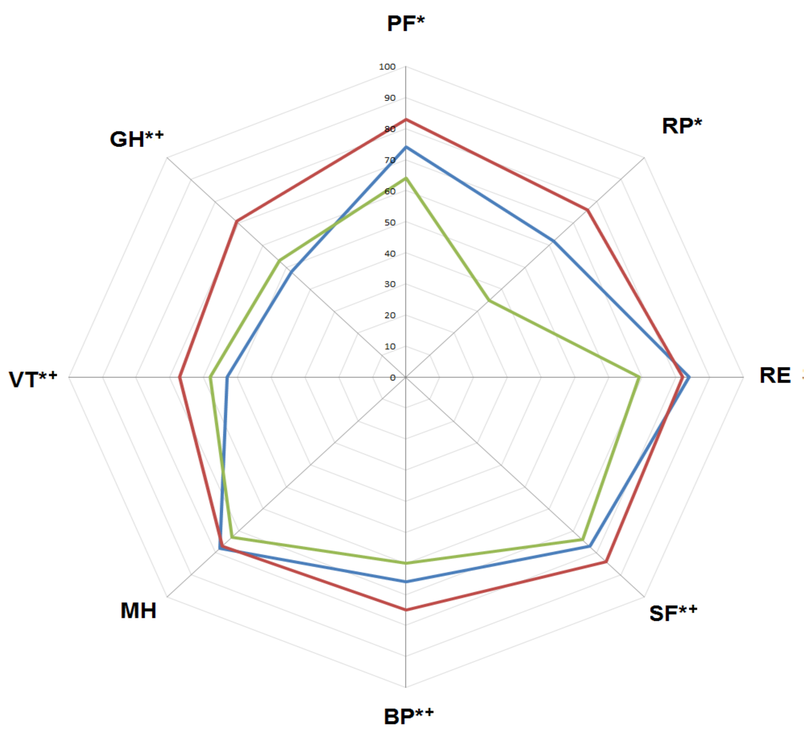
Childhood-onset systemic lupus erythematosus (cSLE) is a severe, chronic multi-system autoimmune disease. Little is known regarding outcomes in adult life. This study addresses these very long-term outcome of cSLE.

Results

Characteristic (n = 47)	Outcome	
Ethnicity White	68%	
% females	91%	
Age at diagnosis (median+range)	14 yrs (7 – 17)	
Disease duration (median+range)	16 yrs (1 – 36)	
Current SLEDAI-score (median+range)	4 (0 – 14)	
Medication use	**Ever**	**Current**
Prednisone	98%	67%
Hydroxychloroquine	81%	63%
MMF	46%	22%
Azathioprine	65%	24%
ACE-I or ARB	64%	43%
SLICC damage index (median + range)	1 (0 – 8)	
% SLICC damage index ≥ 1	62%	
Renal involvement ever	51% (24/47)	
Renal damage in SDI	21% (10/47)	
End stage renal disease	13% (6/47)	
Renal transplant	13% (6/47)	
CNS involvement ever	38% (18/47)	
CNS damage on SDI	23% (11/47)	
Hospitalization due to infections	49%	

Health-Related Quality of Life in cSLE patients, compared to cancer patients and to Dutch normative data

PF : Physical Functioning
RP : Role Physical
RE : Role Emotional
SF : Social Functioning
BP : Bodily Pain
MH: Mental Health
VT : Vitality
GH : General Health



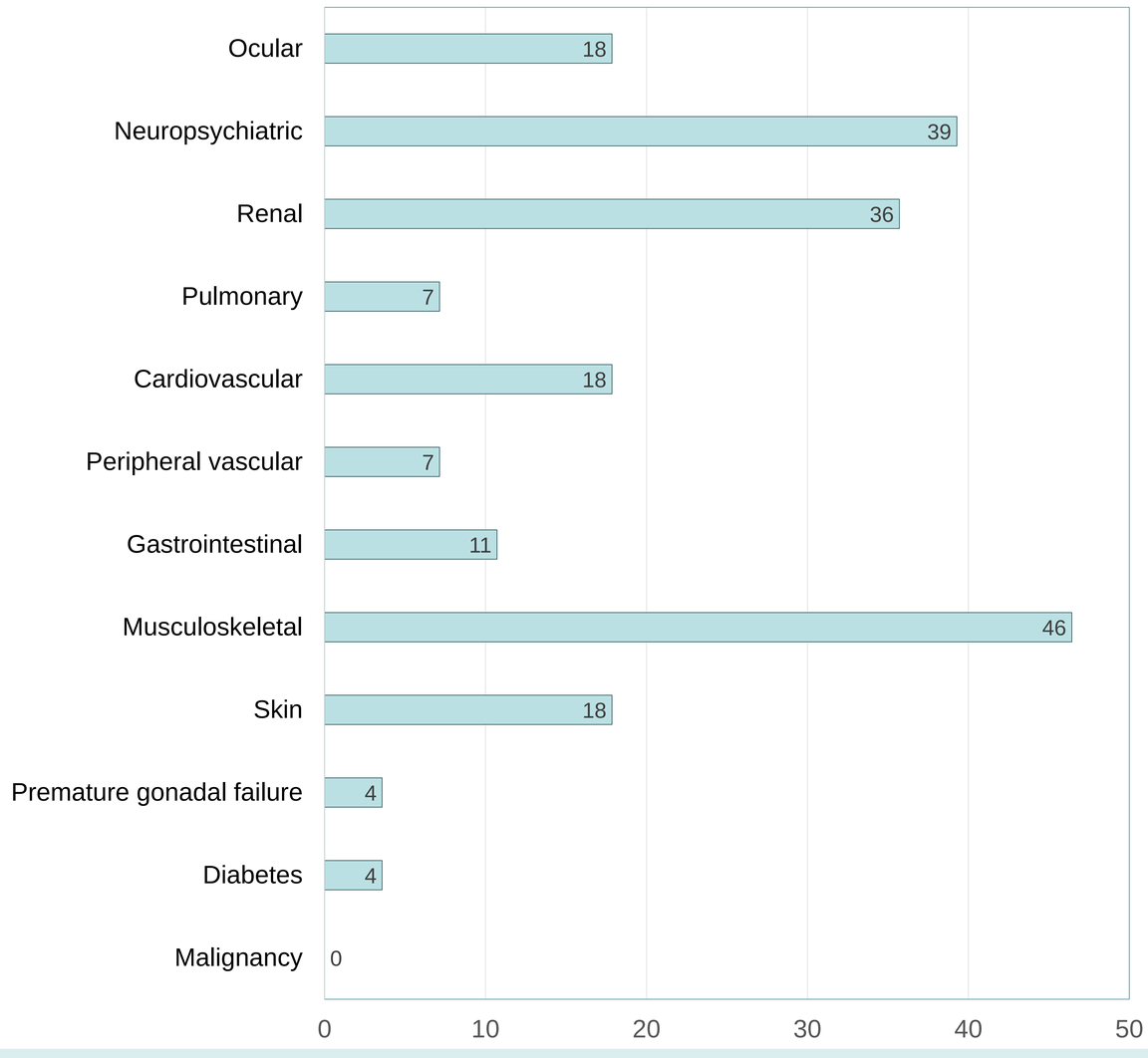
* Significantly lower than normative data
\+ Comparable to or significantly lower than cancer patients

Conclusion

These results underscore the severe disease course in cSLE:

- 62% of patients have a SLICC Damage Index ≥ 1.
- The majority of patients are still using prednisone.
- Many patients who had renal or CNS involvement also developed damage in that organ system.
- Health-related Quality of Life is impaired, and in some domains comparable to patients with cancer.

SDI - % damage per organ system



Methods

DESIGN: Cross-sectional study

STUDY POPULATION: Patients diagnosed with childhood-onset SLE (diagnosis <18 years) now older than 18 years.

OUTCOME: Ethnicity, age at diagnosis, ACR/SLICC classification criteria at diagnosis, clinical features over time, medication use over time, current disease activity, disease damage, quality of life.

Future Plans

- Increase patient numbers (total n ≥ 100)
- Include adult-onset SLE control group (total n ≥ 50)
- Complete data analysis of disease activity and damage
- Complete data analysis of burden of disease

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