

بسم الله الرحمن الرحيم





شبكة المعلومات الجامعية التوثيق الالكتروني والميكرو فيلم



جامعة عين شمس

التوثيق الإلكتروني والميكرو فيلم

قسم

نقسم بالله العظيم أن المادة التي تم توثيقها وتسجيلها
على هذه الأقراص المدمجة قد أعدت دون أية تغييرات



يجب أن

تحتفظ هذه الأقراص المدمجة بعيدا عن الغبار





Health-Related Quality of Life Assessment in Pediatric Systemic Lupus Erythematosus

Thesis

*Submitted for Partial Fulfillment of Master's Degree in
Pediatrics*

By

Mohamed Tarek Ramadan Al-Ashkar

M.B., B.Ch; Ain Shams University (2015)

Under supervision of

Prof. Dr. Elham Mohammad Hossny

Professor of Pediatrics

Faculty of Medicine - Ain Shams University

Prof. Dr. Azza Mohamed Youssef

Professor of Pediatrics

Faculty of Medicine - Ain Shams University

Dr. Nesrine Mohamed Radwan

Lecturer of Pediatrics

Faculty of Medicine - Ain Shams University

*Faculty of Medicine
Ain Shams University*

2021

بِسْمِ اللَّهِ الرَّحْمَنِ الرَّحِيمِ

قَالَ

سَبِّحْ اِنَّكَ لَا تَعْلَمُ لَنَا
اِلَّا مَا عَلَّمْتَنَا اِنَّكَ اَنْتَ
الْعَلِيمُ الْعَظِيمُ

صدق الله العظيم

سورة البقرة الآية: ٣٢

Acknowledgment

*First and foremost, I feel always indebted to **ALLAH**,
the Most Kind and Most Merciful.*

*I would like to express my respectful thanks and profound
gratitude to **Prof. Dr. Elham Mohammad Hossny**,
Professor of Pediatrics, - Ain Shams University for her keen
guidance, kind supervision, valuable advice and continuous
encouragement.*

*I am also delighted to express my deepest gratitude and
thanks to **Prof. Dr. Azza Mohamed Youssef**, Professor
of Pediatrics - Ain Shams University, for her kind care,
continuous supervision, valuable instructions, constant help and
great assistance throughout this work.*

*I am deeply thankful to **Dr. Nesrine Mohamed
Radwan**, Lecturer of Pediatrics - Ain Shams University, for
her great help, active participation and guidance.*

*Last, but by no means least, I wish to express my
appreciation to the patients and their families for their kind
cooperation and to my family for support and understanding.*

Mohamed Tarek

List of Contents

| Title | Page No. |
|---|----------|
| List of Abbreviations..... | i |
| List of Tables | i |
| List of Figures | iii |
| Introduction and Aim of the Work..... | 1 |
| Review of Literature | |
| Pediatric Systemic Lupus Erythematosus | 3 |
| Health-Related Quality of Life in Pediatric Systemic Lupus Erythematosus | 13 |
| Patients and Methods..... | 20 |
| Results | 25 |
| Discussion | 47 |
| Recommendations | 54 |
| Summary | 55 |
| References | 57 |
| Appendix..... | 81 |
| Arabic Summary | — |

List of Abbreviations

| Abb. | Full term |
|--------------|--|
| ANA | Antinuclear antibody |
| AZA..... | Azathioprine |
| CBC | Complete blood count |
| CHAQ | The Childhood Health Assessment Questionnaire |
| CMP..... | Complete metabolic panel |
| CRP..... | C-reactive protein |
| CYC | Cyclophosphamide |
| dsDNA..... | Double-stranded DNA |
| ESR..... | Erythrocyte sedimentation rate |
| EULAR | European League Against Rheumatism |
| GCs..... | Glucocorticoids |
| HAQ..... | Health Assessment Questionnaire |
| HCQ | Hydroxychloroquine |
| HRQOL | Health- related quality of life |
| IQR | Interquartile range |
| MCTD | Mixed connective tissue disease |
| MMF | Mycophenolate mofetil |
| PedsQL | Pediatric Quality of Life Inventory |
| PJP | Pneumocystis jirovecii pneumonia |
| PRO | Patient-reported outcome |
| pSLE..... | Pediatric SLE |
| QOL..... | Quality of life |
| RNP | Ribonuclear protein |
| SF-36 | Short Form-36 |
| SLE..... | Systemic lupus erythematosus |

List of Abbreviations Cont...

| Abb. | Full term |
|--------------|--|
| SLEDAI | Systemic Lupus Erythematosus Disease Activity Index |
| SLEQOL | SLE-specific Quality of Life questionnaire |
| SLICC | Systemic Lupus International Collaborating Clinics Classification Criteria |
| SMILEY | Simple Measure of the Impact of Lupus Erythematosus in Youngsters© |

List of Tables

| Table No. | Title | Page No. |
|--------------------|--|----------|
| Table (1): | EULAR/ACR Clinical Domains and Criteria for SLE (Aringer et al., 2019): | 9 |
| Table (2): | EULAR/ACR Immunologic Domains and Criteria for SLE (Aringer et al., 2019): | 9 |
| Table (3): | Demographic data for the studied group | 25 |
| Table (4): | Distribution of residency and Socioeconomic variables | 26 |
| Table (5): | Disease status in the studied sample | 27 |
| Table (6): | Number and percentage of patients with organ affection..... | 27 |
| Table (7): | Number of different organs affected in the studied group..... | 28 |
| Table (8): | Distribution of different drugs used for treatment for whole studied group..... | 28 |
| Table (9): | Distribution of immunosuppressive medication..... | 29 |
| Table (10): | SLEQOL SCORE for the studied group..... | 30 |
| Table (11): | Correlation of total score according to demographic data | 31 |
| Table (12): | Variation of SLEQOL scores according to residential classification. | 32 |
| Table (13): | Variation of SLEQOL scores according to the school grade | 33 |
| Table (14): | Variation of SLEQOL scores according to disease status | 34 |
| Table (15): | Variation of SLEQOL scores according to of lupus carditis | 37 |
| Table (16): | Variation of SLEQOL scores according to lupus cerebritis. | 38 |

List of Tables Cont...

| Table No. | Title | Page No. |
|--------------------|--|----------|
| Table (17): | Variation of SLEQOL scores according to lupus Arthritis | 39 |
| Table (18): | Variation of SLEQOL scores according to Cyclophosphamide Therapy..... | 39 |
| Table (19): | Variation of SLEQOL scores according to mycophenolate therapy | 40 |
| Table (20): | Comparison between cyclophosphamide and mycophenolate in SLEQOL scores | 41 |
| Table (21): | Correlation between SMILEY score and demographic data | 42 |
| Table (22): | Variation of SMILEY score according to residential classification | 42 |
| Table (23): | Variation of the SMILEY score according to school grade..... | 43 |
| Table (24): | Variation of SMILEY scores according to disease activity..... | 43 |
| Table (25): | Variation of SMILEY scores according to the class of lupus nephritis. | 45 |
| Table (26): | Variation of SMILEY scores according to clinical and lab data..... | 45 |

List of Figures

| Fig. No. | Title | Page No. |
|--------------------|---|----------|
| Figure (1): | Loss of self-tolerance causing the production of autoantibodies in a genetically susceptible individual causing clinical SLE..... | 4 |
| Figure (2): | Demonstrating the residency distribution among our studied patients | 26 |
| Figure (3): | Therapeutic modalities in the studied sample | 29 |
| Figure (4): | Variation of total score according to number of organs affected | 35 |
| Figure (5): | Comparison between disease status using SLEDAI score (activity and remission) according to total score of SLEQOL. | 35 |
| Figure (6): | Variation of SLEQOL score according to the class of lupus nephritis. | 36 |
| Figure (7): | Comparison between disease status using SLEDAI score (activity and remission) according to SMILEY score. | 44 |
| Figure (8): | Influence of immunosuppressives used on the SMILEY score | 46 |
| Figure (9): | Correlation of SMILEY score according to the Total score of SLEQOL | 46 |

INTRODUCTION

Pediatric systemic lupus erythematosus (pSLE) is an autoimmune disease which predominantly affects females (Nazri et al., 2018). The incidence of pSLE varies between 0.3 and 0.9/100,000 per year. Children tend to have a more severe disease than adults at the onset (Aygün et al., 2019).

HRQOL is defined as the perception that individuals have of their position in life, in the context of the culture and system of values in which they live and in relation to their objectives, expectations, standards, and concerns. HRQOL can change according to the environment and the experiences, as well as in response to certain diseases (Didsbury et al., 2016).

Different questionnaires have been developed to assess HRQOL. Validated Arabic version of SLEQOL is a developed generic instrument, based on the adult version, which provides the opportunity to address a child's HRQOL, regardless of the disease (Ravens-Sieberer et al., 2010a; Ravens-Sieberer et al., 2010b).

AIM OF THE WORK

Our objective is to assess the health-related quality of life HRQOL of patients with systemic lupus erythematosus in the pediatric age group using validated Arabic version of SLEQOL as a step to implement this service in the protocol of care for SLE in the Pediatric Allergy and Immunology Unit of Ain Shams University.

PEDIATRIC SYSTEMIC LUPUS ERYTHEMATOSUS

Definition:

Systemic lupus erythematosus (SLE) is a chronic, multi-system, autoimmune disease of unknown etiology with a heterogeneous range of clinical and serological manifestations. It affects multiple organ, and its hallmark is the production of auto-antibodies (Guzman and Hui-Yuen, 2020). Childhood-onset systemic lupus erythematosus (cSLE) is occurring in patients less than 18years (Silva et al., 2012).

Incidence of SLE:

The incidence of cSLE is about 0.3-0.9 per 100 000 children per year, and the prevalence is about 3.3-24 per 100 000 children (Macedo et al., 2016). cSLE is more common in African Americans, Asians, Hispanics, and Native Americans compared to whites (Hiraki et al., 2009).

The median age at presentation is around 11-12 years, with cSLE rarely reported under the age of 5 years. cSLE has a strong female preponderance; the female to male ratio is at 4:3 and 4:1 for disease onset in the first and second decades of life, respectively (Levy and Kamphuis, 2012).