Health Related Quality of Life of Pediatric Patients with Systemic Lupus Erythematosus (SLE)

Essay

Submitted for partial fulfillment of Masters Degree
in Pediatrics

By

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M.B.B.Ch, 2010

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2017

سورة البقرة الآية: ٣٢

Acknowledgments

First and forever, thanks to ***Allah****, Almighty for giving me the strength and faith to complete my thesis and for everything else.*

I would like to express my deepest gratitude of thanks and respect to ***Dr. Shereen Saad El-Sayed,*** *Professor of Pediatrics, Faculty of Medicine – Ain Shams University, for her motherly encouragement, helpful supervision and continuous guidance during execution of this work. I really have the honor to complete this work under her supervision.*

I am also expressing my sincere appreciation, and deepest gratitude to ***Dr. Hanan Mohamed Ezz El-din Azzam,*** *Professor of Psychiatry, Faculty of Medicine – Ain Shams University, for her valuable support and advices, so to her I am deeply indebted*

Last but not least, I can't forget to thank all members of my Family, especially my ***Parents*** *and my* ***Husband*** *for their care and support in every step of my life.*

Hoda Ismail Hussein Ismail

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List of Abbreviations

Abbr.	Title
AAP	: American Academy of Pediatrics.
ACR	: American College of Rheumatology.
ACS	: Acute Confusional State.
ADHD	: Attention deficit and hyperactive disorder.
AHA	: Autoimmune hemolytic anemia.
AIH	: autoimmune hepatitis.
AIMS	: Arthritis Impact Measurement Scales.
AIMS2-SF	: Arthritis Impact Measurement Scales- Short Form.
ANA	: Antinuclear Antibody.
ANCA	: Antineutrophil cytoplasmic antibodies.
Anti- SSA/Ro	: Anti-Sjögren's-syndrome-related antigen A.
Anti- SSB/La	: Anti-Sjögren's-syndrome-related antigen B.
Anti-RNP	: Antiribonucleoprotein.
Anti-Sm	: Anti smith antibodies.
APA	: Antiphospholipid antibody.
APL	: Antiphospholipid.
APPLE	: Atherosclerosis Prevention in Pediatric LupusErythematosus.
APS	: Antiphospholipid syndrome.
ASK	: Activities Scale for Kids.
a-SLE	: Adult onset systemic lupus erythematosus.
AV Block	: Atrioventricular block.
AZA	: Azathioprine.
BAL	: Bronchoalveolar lavage.
BBB	: Blood brain barrier.
Bcl2	: B-cell lymphoma 2.
BILAG	: British Isles Lupus Assessment Group.
BLK	: B lymphocyte kinase.
BLyS	: B-lymphocyte stimulator.

BRAA : Brain reactive autoantibodies.
C3 : Complement 3.
C4 : Complement 4.
CAD : Coronary artery disease.
CAPS : Catastrophic antiphospholipid syndrome.
CBC : Complete blood count.
CBT : Cognitive-behavioural therapy.
CD-40 : Cluster of differentiation- 40.
CH50 : Hemolytic complement 50.
CHAQ : Child Health Assessment Questionnaire.
CHAQ-DI : Child Health Assessment Questionnaire Damage Index.
CHB : Complete heart block.
CIMT : Carotid intimal medial thickness.
CKD : Chronic kidney disease.
CK-MB : Creatine kinase myocardial band.
CM : Cryptococcal Meningitis.
CMV : Cytomegalovirus.
CNS : Central nervous system.
CRP : C-Reactive Protein.
CSA : Cyclosporine.
CSF : Cerebrospinal fluid.
CT : ComputedTomography.
CVA : Cerebrovascular accident.
CYC : Cyclophosphamide.
DAIs : Disease activity indices.
DCs : Dendritic cells.
DDT : Dichlorodiphenyltrichloroethane.
DIC : Disseminated intravascular coagulation.
DLCO2 : Diffusing capacity of lungs for carbon dioxide.
DLE : Drug- induced lupus.
DM : Dermatomyositis.
DMARDS : Disease-modifying antirheumatic drugs.

DNA : Deoxyribonucleic acid.
dsDNA : Double Stranded- DNA.
EBV : Epstein–Barr virus.
ECG : Electrocardiography.
ECLAM : European Consensus Lupus Activity Measurement.
EEG : Electroencephalogram.
ESKD : End-stage kidney disease.
ESR : Erythrocyte Sedimentation Rate.
EULAR : European League against Rheumatism.
FCGR2B : Fc Fragment Of IgG Receptor IIb.
FDA : Food and Drug Adminstration.
GCSF : Granulocyte colony-stimulating factor.
Geri-AIMS : Arthritis Impact Measurement Scales- Geriatric Form.
GFR : Glomerular filtration rate.
GI : Gastrointestinal.
GWAS : Genome-wide association studies.
HAQ : Health assessment questionnaire.
HERVs : Human endogenous retroviruses.
HIV : Humman Immunodeficiency virus.
HLA : Human leukocyte antigen.
HLADRB1 : HLA class II beta chain gene.
HRQOL : Health-related quality of life.
HUS : Hemolytic uremic syndrome.
IAS : Intra-articular steroids.
ICF : International Classification of Functioning, Disability, and Health.
ICs : Immune complexes.
IFN : Interferon.
Ig A : Immunoglobulin A.
IgG : Immunoglobulin G.
IL : Interleukin.
IRF5 : Interferon regulatory factor 5.
IV : Intravenous.

IVIG : Intravenous immunoglobulins.
JAQQ : Juvenile Arthritis Quality of Life Questionnaire.
JDM : Juvenile Dermatomyositis.
JIA : Juvenile Idiopathic Arthritis.
JSLE : juvenile systemic lupus erythematosus.
LAC : Lupus anticoagulant.
LDL : Low-density lipoprotein.
LE Cells : Lupus erythematosus cells.
LFTs : Liver function tests.
LMV : Lupus mesenteric vasculitis.
LN : Lupus nephritis.
LSE : Libman–Sacks endocarditis.
LUMINA : Lupus in Minority Populations, Nature versus Nurture.
LUNAR : Lupus Nephritis Assessment with Rituximab
LV : Lupus vascuitis.
MAP-2 : Microtubule-associated protein 2.
MAS : Macrophage activation syndrome.
MCID : Minimal clinically important difference.
MCP-1 : Monocyte chemotactic protein–1.
MI : Myocardial infarction.
mI : Myoinositol.
MMF : Mycophenolate mofetil
MRA : Magnetic resonance angiography.
MRI : Magnetic resonance imaging.
MS : Multiple sclerosis.
MTX : Methotrexate.
NAA : N-acetylaspartate.
NETs : Neutrophil extracellular traps.
NF- κB : Nuclear factor-κB.
NLE : Neonatal lupus erythematosus.
NMDA : N-methyl-d-aspartate receptor.
NMO : Neuromyelitis optica.

NNV : Noninflammatory, necrotizing lupus vasculopathy.
NP : Neuropsychiatric.
NPSLE : Neuropsychiatric systemic lupus erythematosus.
NR2 : *N*-methyl-d-aspartate (NMDA) receptor.
NSAIDS : Non-steroidal anti-inflammatory drugs.
OCD : Obsessive–compulsive disorder.
ON : Optic neuritis.
PAH : Pulmonary alveolar hemorrhage.
PCD1 : Programmed cell death 1.
PCR : Polymerase chain reaction.
PedsQL : Pediatric Quality of Life Inventory.
PET : Positron emission tomography.
PLE : Lupus-related protein-losing enteropathy.
PNS : Peripheral nervous system.
PRES : Posterior reversible encephalopathy syndrome.
PRLS : Reversible posterior leukoencephalopathy syndrome.
PRQL : Pediatric Rheumatology Quality of Life.
PSLE : Pediatric Sysytemic lupus erythematosus.
QOL : Quality of Life.
QoMLQ : Quality of My Life Questionnaire.
RA : Rheumatoid Arthritis.
RBC/hpf : Red blood cell count per high power field.
RBC : Red blood cell.
RCPCH : Royal College of Pediatrics and Child Health.
RF : Rheumatoid Factor.
RNP : Ribonuclear protein.
RVLs : Renal vascular lesions.
SDI : Systemic Lupus International Collaborating Clinics/American College of Rheumatology (SLICC/ACR) Damage index.
SELENA-SLEDAI: Safety of Estrogens in Lupus Erythematosus National Assessment- Systemic Lupus Erythematosus Disease Activity Index.

SLAM : Systemic Lupus Activity Measure.
SLE : Systemic Lupus Erythematosus.
SLEDAI : Systemic Lupus Erythematosus Disease Activity Index.
SLICC : Systemic Lupus International Collaborating Clinics.
SMILEY : Simple Measure of the Impact of Lupus Erythematous in Youngsters.
SPECT : Single-photon emission computed tomography.
SR : Suicide risk.
STAT4 : Signal transducer and activator of transcription 4.
SVT : Supraventricular tachycardia.
TAC : Tacrolimus.
TB : Tuberculosis.
TEE : Transesophageal echocardiography.
TEs : Thromboembolic events.
TI : Tubulointerstitial.
TIN : Tubulointerstitial nephritis.
TLR : Toll-like receptors.
TMA : Thrombotic microangiopathy.
TNF : Tumor necrosis factor.
TNF-a : Tumor necrosis factor-α.
TNFAIP3 : TNF Alpha Induced Protein 3.
TNFSF13 : Tumor necrosis factor ligand superfamily member 13.
TNIP1 : TNFAIP3 Interacting Protein 1.
TTE : Transthoracic echocardiography.
TTP : Thrombotic thrombocytopenic purpura.
Up/c : UrineProtein/Creatinine(ratio).
UVB : Ultraviolet B.
VAS : Visual Analogue Scale.
VIDs : Vascular immune complex deposits.
VZV : Varicella Zoster virus.
WBC : White blood cell.
WHO : World Health Organization.

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Abstract

Background: Systemic Lupus Erythematosus (SLE) is an autoimmune rheumatic disease that is characterized by the production of autoantibodies which leads to immune complex deposition, inflammation and permanent organ damage. Hormonal, environmental and genetic factors play a contributory role in the development of SLE. **Aim of the Work:** To describe the psychological problems and factors affecting quality of life of patients with pediatric Systemic Lupus Erythematosus (SLE). **Conclusion:** The assessment of patients with lupus should include the determination of disease activity, chronic damage resulting from lupus activity or its treatment, health-related quality of life (HRQOL), and economic impact.

Key words: pediatric systemic lupus erythematosus (SLE), autoimmune rheumatic disease, autoantibodies, HRQOL, neuropsychiatric lupus

Introduction

Systemic lupus erythematosus (SLE) is a rheumatic disease characterized by autoantibodies directed against self-antigens, immune complex formation, and immune dysregulation, resulting in damage to essentially any organ. The disease can affect, for example, the kidneys, skin, blood cells, and nervous system. The natural history of SLE is unpredictable; patients may present with many years of symptoms or with acute, life-threatening disease ***(Klein-Gitelman, 2013).*** People with lupus can have times of very active disease, called a flare, and times where the disease is mostly quiet, called remission ***(Tucker et al., 2012).***

It occurs from infancy to old age, with peak occurrence between ages 15 and 40. Females are affected far more than males (6-10:1). Blacks (and possibly Hispanics, Asians, and Native Americans) are affected more than whites ***(Rus et al., 2014).*** Rheumatologic disorders in pediatric patients are typically more severe clinically than the same disorders in adults, and may have significant psychiatric and medical consequences ***(Brunner et al., 2011).***

Central nervous system involvement occurs in more than half of children and adolescents with SLE, usually early in the course of the disease, and typically includes both neurologic and psychiatric symptoms ***(Silber et al., 2008).***

Cognitive, mood, and psychotic symptoms in CNS lupus may be related to an underlying vasculitis, or may reflect the impact of anti-phospholipid, anti-neuronal, anti-receptor antibodies ***(Steinlin et al., 2008).***

While there are numerous studies of psychiatric problems in adult patients with SLE, and some describing mood, cognitive, and psychotic problems in pediatric SLE patients, there are no outcome studies of the long term consequences of pediatric psychosocial dysfunction ***(Turkel et al., 2001).*** SLE is associated with both primary and secondary effects on psychosocial functioning. The prevalence rate of depressive symptoms with SLE is 30%. Depression may be related to the effect of pain and fatigue on mood symptoms ***(Kawakatsu and Wada, 2008).*** Psychosis, depression, anxiety, cognitive deficits and emotional distress are frequently seen in patients with SLE. Depression and other neuropsychiatric symptoms in patients with SLE are associated with increased risk for suicidal behavior ***(Denberg et al., 1997).*** Emotional disturbances and problems with social functioning, personal discomfort in social situations, and depressed mood are more frequent in SLE patients with skin and joint abnormalities, confirming that psychosocial dysfunction may not only be a reflection of direct CNS involvement ***(Monastero et al., 2001).***

Quality of life can be defined as subjective perception of health in physical, emotional, mental, social and functional domains. It is represented in terms of well being as well as functioning and should be identified via patients' self-reporting. Even though defining quality of life as a concept for children, especially with regard to developmental stages, is a challenge, measures to assess quality of life in children from self-reporting as well as external reporting (parents) are emerging ***(Bullinger et al., 2002).*** However, not only is documenting the quality of life of patients and family important, but also the unravelling of the determinants or factors influencing the quality of life. Here psychological constructs, such as coping and adaptation, health locus of control and health beliefs, as well as social support and social networks have been identified as major factors influencing patient-perceived quality of life ***(Thompson and Gustafson, 1996).***

There are many challenges in coping with and adapting to life with a chronic disease, and increased survival cannot be assumed to be associated with increased quality of life. A recent systematic review shows there is wide variation in outcomes depending on the definitions and measurements used to estimate the prevalence of chronic health conditions, making the impact of disability on children's health and social functioning difficult to assess, and different authors have called for an international consensus about the