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Impact of Medical Counseling Program on Quality of Life in Children with Neuromuscular Diseases and their Families

Thesis submitted by

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Ain Shams University 2017

Acknowledgment

I would like to express my endless gratefulness to gracious \boldsymbol{God} for helping me to achieve this work.

It was a great honor to accomplish this research under the supervision of Prof. *Ahmed Mohamed Osman El-Kahky*, Professor in Medical Department-Institute of Postgraduate Childhood Studies- Ain Shams University, for his guidance, support, valuable directions and continuous encouragement.

I wish to express my greatest appreciation for Prof. *Samia Samy Aziz*, Professor in Medical Department-Institute of Postgraduate Childhood Studies- Ain Shams University, for her great support and kind and careful assistance and valuable knowledge during this work.

Finally, I would like to thank all my professors, my seniors, my colleagues and my family especially my wife for helping me directly and indirectly to carry on this research.

Hanna Saber Ibrahim

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Abstract

Introduction: Neuromuscular diseases affect any part of the nerve and muscle.

They may be genetically determined, congenital or acquired .These disorders include motor neuron diseases such as spinal muscular atrophy, peripheral neuropathy, neuromuscular junction diseases and muscle affection. Unfortunately, most neuromuscular diseases (NMDs) are incurable diseases. However, an effective rehabilitation program can help maintain a patient's quality of life (QOL), as well as maximize the patient's physical and psychosocial functions. An effective rehabilitation program can also minimize secondary medical co morbidity, prevent or limit physical deformity, and allow the patient to integrate into the society.

Objectives: Our study was done to improve the quality of life of patients with neuromuscular diseases and their families.

Methods: This was a prospective, interventional and uncontrolled study conducted at Abo Reesh teaching hospital between January 2015 - December 2015. The study was carried on infants, children and adolescents suffering from neuromuscular diseases and they were attending neurology clinic throughout one year. Sixty patients were selected from outpatient clinic with well-known diagnosis of NMD variant who were grouped according to their age (3-5) per group mothers also grouped (6-10) per group. The study was achieved through application of medical counseling program through 24



sessions once per week lasting for 6 months. Pre-and post program an individualized neuromuscular quality of life questionnaire was applied to assess the effectiveness of the program.

Results: Patients reported significantly lower QOL across all QOL domains, physical and psychosocial. With advancing age, patients reported decreased physical functioning and daily activities. While psychosocial functioning was impaired in, a significant number of patients (60%), older patients did not tend to perceive lower psychosocial QOL despite their increased physical limitations.

Recommendations: We recommend the following: all treatment measures either medical or surgical should be directed to increase quality of life better than treatment of the cause of alleviating the symptoms, measurement of treatment success is beyond the primary goal as modern directions in medicine targeting the whole individual not only the system affected, Psychological dimension is an important part of the disease process and treatment as well, physician should not omit this option during treatment planning.

Key words: Quality of life – Neuromuscular dis.-Med .counselling

List of abbreviations

ADL	activities of daily living
ALS	amyotrophic lateral sclerosis
AONs	Antisense Oligonucleotides
BDI	Beck depression index
BMD	Becker MD
CMD	Congenital MD
CMT	Charcot-Marie-Tooth disease
_	
CNS	Central nervous system
CSF	Cerebrospinal fluid
DMD	Duchene MD
DMD	Distal MD
DM1	Myotonic dystrophy type 1
DM2	Myotonic dystrophy type 2
DNA	Deoxyribonucleic Acid
ECG	Electrocardiography
EMG	electromyography
EDMD	Emery-Dreifuss MD
ERT	Enzyme replacement therapy
FDA	Food and Drug Administration
FEES	flexible endoscopic evaluation of swallowing
FRS	Functional Rating Scale
FSHD	Facioscapulohumeral dystrophy
FVC	Forced vital capacity

\square List of Abbreviations \angle

GABA	Gamma-aminobutyric acid
LGMD	Limb-girdle MD
MD	muscular dystrophy
MG	Myasthenia gravis
MS	Muscular sclerosis
NMD	Neuromuscular disorder
NSAIDs	Nonsteroidal anti-inflammatory drugs
OPMD	Oculopharyngeal MD
QOL	Quality of life
SMA	Spinal muscular atrophy
SMEP	strictly observed exercise program
SOD	Superoxide dismutase
TCA	tricyclic antidepressants
TMP	Timed Motor Performance

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Introduction

The term of neuromuscular diseases, (NMD) encompasses wide varieties of neuromuscular disease of different etiologies. The impact of having any variety of NMD is great in general. In the past, scientists thought that presence of multiple methods to diagnosis and new modalities of treatment was adequate in any national health trust (*Moyle et al.*, 2015).

Recently, progress of any disease or admission of any therapy should be linked to Quality of Life (QOL) research to obtain their impact on patients' life (*van Dulmen et al.*, 2015).

Quality of life research is approached on either healthcare system or patient-focused approach. Every classification has its own advantages or drawbacks (*van Dulmen et al.*, 2015).

Application of QOL research should be based on meticulous design, involved large scale of individuals and disease-specific if possible (*Schipper & Levitt*, 1985).

Several studies take the NMD to target their research of QOL Knowing patients' satisfactions and their feedback are small areas in QOL research (*Brack et al.*, 2005).

In Egypt, studying of NMD-QOL will be advantageous to health care systems whatever approach used.

Aim of the Study

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The aim of the study is to improve quality of life of patients with neuromuscular diseases and their families