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Assessment of behavioral and psychological disturbance in children and Adolescent patients with Beta Thalassemia Major

Thesis

Submitted for Partial Fulfillment of Master Degree in Pediatrics

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First, thanks are all to Allah for Blessing this work until it has reached its end, as a part of his generous help throughout our life.

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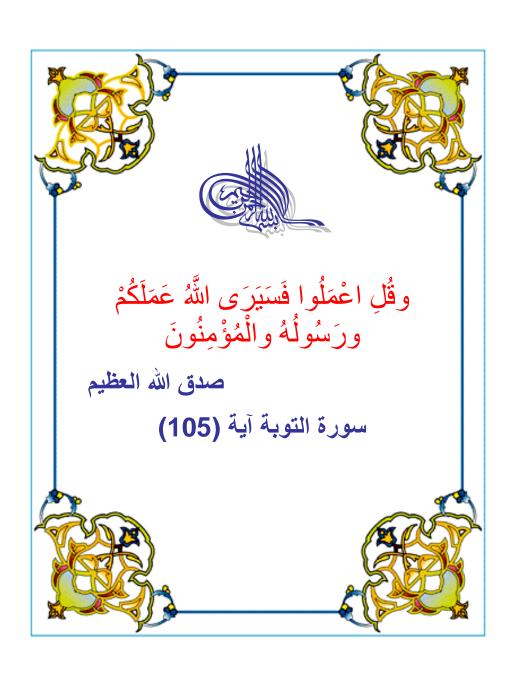
I am deeply grateful to My Mother and Fatherwho directed and encouraged me during the preparation of this work.

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List of abbreviations

γ : Gamma

α : Alpha

β : Beta

ε : Epsilon

Hb : Hemoglobin

IV : Intravenous

SC : Subcutaneous

TLC: Total leucocyte count

Plt : Platelet

T. bil : Total bilirubin

D. bil : Direct bilirubin

EF : Ejection fraction

SD : Standard deviation

LVEF : Left ventricular ejection fraction

DFO: Desferoxamine

RDW: Red cell distribution width

rHuEPO: Recombinant human erythropoietin

RBC: Red blood cell

NTBI : Non transferrin Bound plasma iron

T2 MRI : Magnetic resonance imaging T2-star

WHO: World health organization

MDD : Major depressive disorder

PBRS: Pediatric behavior rating scale

PBRS-P: Pediatric behavior rating scale parents

PBRS-T : Pediatric behavior rating scale teachers

STFR : Serum transferrin receptors

HU: Hydroxyurea

A : Alanine

G: Glycine

TH : Thalassemia

TM : Thalassemia Major

TI : Thalassemia intermedia

LCI : Labile cellular iron

PH : Pulmonary hypertension

ACTH : Adrenocorticotrophic hormone

TRH: Thyrotropin releasing hormone

ICT: Iron chelators therapy

IOC : Iron overload cardiomyopathy

OPSI : Over whelming post splenectomy infection

HAMP: Hepcidin antimicrobial peptide gene

L1 : Deferiprone

GH : Growth hormone

T3: Tri-iodothyronine

T4 : Tetra-iodothyronine

TSH: Thyroid stimulating hormone

LH : Luteinizing hormone

FSH : Follicular stimulating hormone

GnRH : Gonadotropin releasing hormone

ECG: Electrocardiogram

IGF-1: Insulin growth factor 1

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Assessment of behavioral and psychological disturbance in children and Adolescent patients with Beta Thalassemia Major

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Objectives: to study the behavioral and psychological disturbances in children and adolescent patients with beta thalassemia major. Methods: A cross sectional case- control study was conducted on 50 children, and adolescents with beta thalassemia major (30patients without complications and 20 patient with complications). They were recruited from hematology Clinic, Children Hospital, Ain Shams University Cairo, Egypt during the period from January 2013 to January 2014. Their age ranged between 6-18 years with a mean of +8.56 years. They included 27 (54%) females and 23 (46%) males. The control group included 50 healthy ages and sex matched healthy subjects. All patientswere subjected to clinical assessment and hematological assessment, behavioral and psychological assessment using a questionnaire: PBRS-P; appropriate for use in children and adolescents aged 6 to 18 years. **Results:** compared to controls all patients had statistically significant behavioral disturbances namely atypical behavior (P<0.001), irritability (P<0.001), grandiosity (P<0.001), aggressive behavior (P<0.001), affect disorder (P<0.001), hyper activity (p<0.001), inattention (p<0.001), disturbed social interaction (P<0.001) when compared to controls. On comparing patients without complications and patient with complications as regardedbehavioral disturbances no significant difference was found in atypical behavior (P=0.15), irritability (P=0.14), grandiosity (P=0.90), affect disorder (p=0.76) and hyper activity (P=0.054). Aggressive behavior, inattention and disturbed social interaction were significantly more frequent in complicated group (p= 0.005, p=0.035&p=0.004 respectively).

Conclusions: behavioral and psychological problems are common in patients with beta thalassemia major either with complications or without complications.