PLASMA LIPIDS AND LIPOPROTEINS PATTERN IN B-THALASSEMIA MAJOR

Thesis

Submitted for the partial fulfilment

of the M.D. Degree in Paediatrics.

By

FATMA EL ZAHRAA ABDEL BASSET MOHAMED

M.B.,B.Ch.,Ain Shams University, 1980,

M.Sc. of Paediatrics, Ain Shams University, 1985

Supervised by

Prof. AHMED SAMY KHALIFA

Prof. of Paediatrics.

Ain Shams University

Prof. KOTB AHMED TOLBA

Prof. of Paediatrics

Ain Shams University

Prof. FADILA HASSAN SABRY

Prof. of Clinical Pathology

Ain Shams University

1990

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TO MY HUSBAND

AND MY LOVELY TWIN

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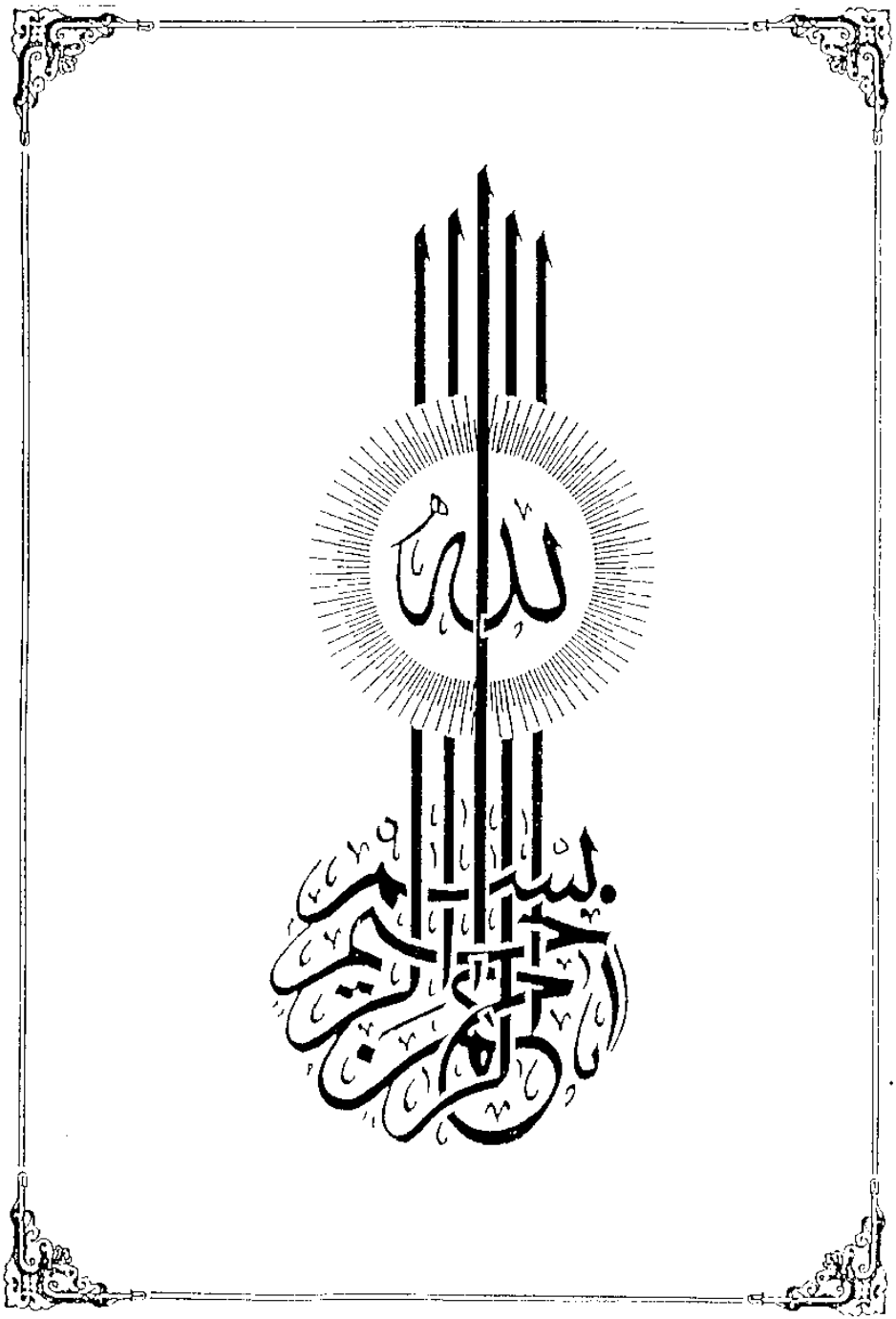

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LIST OF ABBREVIATIONS

CE:	Cholesterol ester(s).
PG:	Prostaglandin(s).
FFA:	Free FAtty Acid(s).
CM:	Chylomicron(s).
VLDL:	Very Low Density Lipoprotein(s).
LDL:	Low Density Lipoprotein(s).
HDL:	High Density Lipoprotein(s).
α:	Alpha.
Pre-β:	Pre-beta.
β:	Beta.
IDL:	Intermediate Density Lipoprotein(s).
ApoA-I:	Apoprotein A-I.
LCAT:	Lecithin-Cholesterol Acyl Transferase.
FABP:	Fatty Acid Binding Protein(s).
ACAT:	Acyl CoA Cholesterol Transferase.
SER:	Smooth Endoplasmic Reticulum.
RER:	Rough Endoplasmic Reticulum.
LPL:	Lipoprotein Lipase.
TG:	Triglyceride(s).
HMG Co reductase:	Hydorxy Methyl Glyteryl Co enzyme A Reductase.
LP-X:	Lipoprotein X.
Hb:	Hemoglobin.
GIT:	Gastro-Intestinal Tract.
HBV:	Hepatitis B Virus.
DTPA:	Diethylene Triamine Penta-Acetic Acid.
SQID:	Superconduction Quantum Interference Device.
DXS:	Diagnostic X-ray Spectrometery.
γ:	Gamma.
HAITF:	Host-Associated Iron Transfer Factor.
H2O2:	Hydrogen Peroxide.
RE:	Reticulo-Endothelial.
NANB:	Non A Non B.
HBsAg:	Hepatitis B Surface Antigen.
Anti-HBs:	Hepatitis B surface antibody.
Anti-HBc:	Hepatitis B Core Antibody.
HAV:	Hepatitis A Virus.
C.H.D.:	Coronary Heart Disease.
C:	Cholesterol.
HDL-C:	High Density Lipoprotein Cholesterol.

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LDL-C:	Low Density Lipoprotein Cholesterol.
H-TGL:	Hepatic Triglyceride Lipase.
T_3:	Tri-iodothyronine
GK	Glucokinase
G-1-P-DH	Glucose-1-phosphate dehydrogenase.
NAD	Nicotinamide adenine dinucleotide.
INT	Iodonitrotetrazolium,violet.
DAP	Dihydroxyacetone phosphate
G-1-P	Glycerol-1-phosphate.

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INTRODUCTION AND AIM OF THE WORK

Beta thalassemia is the commonest chronic hemolytic anemia in Egypt (Sabry,1973). The ineffective erythropoiesis, hemolysis and anemia are the basic pathologic processes in this disease. The children with B-thalassemia major are kept alive by repeated blood transfusion but with unfortunate consequences of massive accumulation of iron in different organs including the liver (Propper et al.,1980).

Thalassemic liver disease is not only an outcome of hemosiderosis, but also of repeated attacks of post-transfusional hepatitis (Masera et al.,1976).

Gradually fibrosis and eventually cirrhosis develop in these patients (Okon et al.,1976).

The liver plays a central role in the metabolism of lipids and lipoproteins,apart from its role in facilitating digestion and absorption of lipids by the production of bile, the liver has active enzyme system for both synthesis and catabolism of lipoproteins (McIntyre,1978).

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The aim of this work is to study the plasma lipids and lipoproteins pattern in patients with β-thalassemia major and to correlate this pattern with the liver functions and iron overload in these patients.

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CHEMISTRY OF LIPIDS

The lipids are a heterogenous group of compounds related to the fatty acids.

Lipids are important dietary constituents being an efficient source of energy both directly, and potentially, when stored in adipose tissues. They serve as a thermal insulator in the subcutaneous tissues, as a support around certain organs and as an electrical insulator for propagation of the depolarization waves along peripheral myelinated nerves.

Combinations of fat and protein, the lipoproteins, are important cellular constituents, existing both in the cell membrane and mitochondria and serving also as the means of transport of lipids in the blood.

Lipids have been classified into:

I-Simple lipids:

Those are esters of fatty acids with various alcohols. They include:

1-Fats:

Esters of fatty acids with glycerol.

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