Electro Cardiographic Assessment in Patients with Transfusion Dependant Beta Thalassemia Major Using Stress ECG and 24hrs ECG Monitoring

Thesis

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بسم الله الرحمن الرحيم

قَالُوا سُبْحَانَكَ لَا عِلْمَ لَنَا إِلَّا مَا عَلَّمْتَنَا
إِنَّكَ أَنتَ الْعَلِيمُ الْحَكِيمُ

صدق الله العظيم

البقرة الآية ٣٢

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استخدام جهاز الهولتر ورسم القلب بالمجهود فى مرضى أنيميا البحر المتوسط المعتمدون فى علاجهم على نقل الدم بصفة منتظمة

رسالة

توطئة للحصول على درجة الماجستير

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List of Abbreviations

Abb.	Mean
β	Beta
AHSP	Alpha Hb stabilizing protein
BM	Bone marrow
CMR	Cardiac magnetic resonance
CVS	Chorionic villous sampling
DBP	Diastolic blood pressure
DFO	Desferoxamine
DFT	Desferrithiocin
DM	Diabetes mellitus
EDV	End diastolic volume
EF	Ejection fraction
ESV	End systolic volume
FS	Fractional segment
HBED	Hydroxy benzyle ethylene diamine-diacetic acid
HCV	Hepatitis C virus
HDL	High density lipoprotein
HIC	Hepatic iron concentration
IFN	Interferon
IGF-1	Insulin like growth factor-1
IVSD	Inter ventricular septal diameter
LAD	Left atrial diameter
LDL	Low density lipoprotein

Abb.	Mean
LIC	Liver iron concentration
LPI	Labile plasma iron
LV	Left ventricle
LVEF	Left ventricle ejection fraction
LVIDd	Left ventricular intra diastolic diameter
LVPWd	Left ventricular posterior wall diameter
METs	Metabolic equivalent tasks
NTBI	Non transferrin bound iron
PH	Pulmonary hypertension
PVC	Premature ventricular contraction
PVR	Pulmonary vascular resistance
RDW	Red Cell distribution width
SBP	Systolic blood pressure
SVT	Supra ventricular tachycardia
TI	Thalassemia intermedia
TM	Thalassemia major

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INTRODUCTION

Thalassemia is a genetic disorders in globin chain production. In individuals with β-thalassemia, there is either a complete absence of β-globin production (β°-thalassemia) or a partial reduction (β+- thalassemia ***(Debaun and Vichinisky, 2007).***

Beta thalassemia usually become symptomatic as severe hemolytic anemia during 2nd 6 month of life, regular blood transfusion are necessary in these patients to prevent cardiac decompensation caused by anemia ***(Eldor et al., 2002).***

Conventional management of β-thalassaemia major requires regular blood transfusions. This leads to excess iron accumulation, initially in the reticuloendothelial system and subsequently in all parenchymal organs, mainly heart, pituitary gland, pancreas and gonads, resulting in serious and sometimes fatal clinical complications ***(Christoforidis et al., 2006)***.

Transfusions are given on regular basis to maintain hemoglobin level above 10g / dl, which prevents progressive marrow expansion and cosmetic problems associated with facial bone changes and minimize cardiac dilation and osteoporosis ***(Wonke et al., 2001).***

The major cause of morbidity and mortality in patients with beta thalassemia major is iron overload associated with chronic blood transfusion therapy, which can lead to iron deposition and damage to the heart, liver and endocrine organs and other organ failure. Iron toxicity is the leading cause of death in beta thalassemia major patients ***(Wood et al., 2005).***

Vascular dysfunction with increased arterial stiffness and endothelial dysfunction has been demonstrated in patients with beta-thalassemia major ***(Cheung et al., 2002).*** Endothelial dysfunction as well as arterial stiffening is an important precursors for atherosclerosis ***(Davignon and Ganz, 2004),*** studies have also suggested a link between iron load and risk of atherosclerosis ***(Timothy et al., 2007).***

Thalassemic (TM) patients are subjected to oxidative tissue injury because of continuous blood transfusions. It has been documented that circulating LDL from TM patients show marked oxidative modification that could represent an event leading to atherogenesis ***(Brizzi et al., 2002)***.

In recent years increasing evidence suggests that the oxidative modification of low-density lipoprotein (LDL) is the key step in the sequence of events leading to atherogenesis-related vascular alterations, so modified LDL