



ROLE OF CARDIAC MRI (1.5T) IN THALASSAEMIA

Essay

Submitted for fulfillment of Master degree
in Radiodiagnosis

By

Noha Ali Gad Hassan

M.B.B, Ch.2007
Ain Shams University

Supervised By

Dr. Ahmed Samir Ibrahim

Professor of Radiodiagnosis
Faculty of Medicine – Ain Shams University

Dr. Mary Yaftah Tadros

Lecturer of Radiodiagnosis
Faculty of Medicine – Ain Shams University

*Faculty of Medicine
Ain Shams University*
2015-2016



Acknowledgments

*First and foremost, I feel always indebted to **Allah**, the Most Beneficent and Merciful.*

*I wish to express my deepest gratitude and thanks to **Dr. Ahmed Samir Ibrahim**, Professor of Radiodiagnosis, Faculty of Medicine – Ain Shams University, for his constructive criticism, unlimited help and giving me the privilege to work under his supervision.*

*My most sincere gratitude is also extended to **Dr. Mary Yaftah Tadros**, Lecturer of Radiodiagnosis, Faculty of Medicine – Ain Shams University, for her enthusiastic help, continuous supervision, guidance and support throughout this work.*

*Last but not least, I can't forget to thank all members of my **Family**, for pushing me forward in every step in the journey of my life.*

Candidate

 **Noha Ali Gad**

List of Contents

<i>Subject</i>	<i>Page No.</i>
List of Abbreviations.....	i
List of Tables.....	iv
List of Figures	v
Introduction	1
Aim of the Work.....	4
Chapter (1): Anatomy of the Heart	5
Chapter (2): Thalassemia	21
Chapter (3): Techniques of Cardiac MRI.....	34
Chapter (4): Thalassemic Manifestations in Cardiac MRI and Illustrative Cases	74
Summary	98
References	102
Arabic Summary	—

List of Abbreviations

<i>Abbr.</i>	<i>Full-term</i>
AIVG	: Anterior interventricular groove
Ao	: Aorta
APM	: Anterior papillary muscle
AV	: Aortic valve
AV	: Atrioventricular
BB	: Black blood
BH	: Breath-hold scan
CHF	: Congestive heart failure
CMR	: Cardiac Magnetic resonance
CS	: Coronary sinus
CT	: Crista terminalis
D	: Diaphragm,
EPI	: Echo-planar imaging
EV	: Eustachian valve
FO	: Fossa ovalis
FSE	: Fast spin-echo
Gad	: Gadolinium
GRE	: Gradient-echo
IAS	: Interatrial septum,
IOC	: Iron overload cardiomyopathy
IVC	: Inferior vena cava
IVS	: Interventricular septum
LA	: Left atrium
LAAP	: Left atrial appendage
LAD	: Left anterior descending artery
LAO	: Left Anterior oblique
LCA	: Left coronary artery
LCCA	: Left Common carotid artery

LCX	: Left circumflex artery
LIC	: Liver iron content
LIV	: Left innominate vein,
LL	: Left lung
LMB	: Left mainstem bronchus,
LPA	: Left pulmonary artery
LV	: Left ventricle
LVOT	: Left ventricular outflow tract
MB	: Moderator band
MIC	: Myocardial iron concentration
MIP	: Maximum intensity projection
MR	: Magnetic resonance
mTTE	: Multiecho turbo field echo
MV	: Mitral valve
P4CH	: 4 chamber view
PA	: Pulmonary artery
PHT	: Pulmonary hypertension
PMVL	: Posterior mitral valve leaflet
PPM	: Posterior papillary muscle
PUV	: Pulmonary valve
PV	: Pulmonary vein
RA	: Right atrium,
RAAP	: Right atrial appendage
RCA	: Right coronary artery
RL	: Right lung
RMB	: Right mainstem bronchus
ROI	: Regions of interest
RPA	: Right pulmonary artery
RV	: Right ventricle,
RVOT	: Right ventricular outflow tract
SA	: Short-axis
SPAMM	: Spatial modulation of magnetization

SSFP	: Steady state free precession
SSh	: Single shot dual inversion
SVC	: Superior vena cava,
SVC	: Superior vena cava
T	: Trachea
TDT	: Thalassaemia dependent transfusion
TEs	: Echo times
TFE	: Turbo field echo
TSE-MRI	: Turbospin echo MRI
TV	: Tricuspid valve
WB	: White blood
β-TM	: β -thalassemia major

List of Tables

<i>Table No.</i>	<i>Title</i>	<i>Page No.</i>
Table (1):	Causes of death in patients with β – Thalassemia	33
Table (2):	Optimal Planes for Imaging Cardiac Structures and Chambers	49
Table (3):	Parameters for CMR T2* imaging	67
Table (4):	Correlation between iron overload detected by magnetic resonance imaging (ms) and tissue (mg Fe/g dry weight)	92

List of Figures

<i>Figure No.</i>	<i>Title</i>	<i>Page No.</i>
Figure (1):	Position of the heart in the thorax. From left to right, transverse, coronal, and sagittal images using turbospin echo MRI (TSE-MRI) technique.....	5
Figure (2):	External morphology of the heart	6
Figure (3):	Morphological right atrium.....	7
Figure (4):	Axial and sagittal TSE images.	8
Figure (5):	Right axial GRE, left coronal TSE images: <i>raap</i> (right atrial appendage).....	8
Figure (6):	Axial TSE image shows left atrial appendage (<i>laap</i>) and pulmonary vein (<i>pv</i>)	9
Figure (7):	GRE horizontal long axis image. <i>Fo</i> : fossa ovalis.....	10
Figure (8):	The morphologic right ventricle	11
Figure (9):	Horizontal long axis GRE image.....	12
Figure (10):	Short axis GRE view shows the tricuspid valve leaflets	13
Figure (11):	Axial GRE image shows the moderator band	14
Figure (12):	Right ventricle inflow and outflow tract GRE image.....	14
Figure (13):	The morphological left ventricle	15

Figure (14):	GRE left ventricular outflow tract view shows the components of the left ventricle.....	16
Figure (15):	Short axis GRE image shows the mitral valve leaflets and commissures	17
Figure (16):	GRE short axis view shows the papillary muscles.....	17
Figure (17):	Essential characteristics of the morphological right and left ventricles.....	19
Figure (18):	GRE images.....	20
Figure (19):	Effect of excess production of free α globin chains	25
Figure (20):	Pathophysiology of pulmonary hypertension in hemoglobinopathies	31
Figure (21):	Representative black blood images of the heart in the short-axis.....	37
Figure (22):	Axial (A) and sagittal (B) half-Fourier acquisition single-shot turbo spin-echo images	38
Figure (23):	Creation of tag lines of the myocardium, using radial tagging technique	40
Figure (24):	Example of short-axis Spatial modulation of magnetization (SPAMM) tagging	40
Figure (25):	Example of Cardiac Magnetic Resonance Tagging.	41
Figure (26):	Phase-contrast images of a normal subject. A and B, Transverse aortic (A) and pulmonary artery (B) phase-contrast images	42

Figure (27):	Four-chamber (A) and short-axis (B) cine images using SSFP	43
Figure (28):	a–c. Axial, Sagittal and coronal images for the heart	47
Figure (29):	Schematic shows orientation of major body planes with respect to patient and their corresponding appearance on bright blood imaging sequences	47
Figure (30):	Schematic shows orientation of major cardiac planes with respect to heart and their corresponding appearance on bright blood sequences	49
Figure (31):	Normal cardiac MRI anatomy shown in healthy subject.	50
Figure (32):	The 2-chamber scout is planned from the axial scout (<i>Left</i>) planning image; (<i>right</i>) resulting image	53
Figure (33):	The 4-chamber scout is planned from the 2-chamber scout (<i>Left</i>) planning image; (<i>right</i>) resulting image	54
Figure (34):	The short-axis scout is planned from the 4-chamber scout (<i>Left</i>) planning image; (<i>right</i>) resulting Image	54
Figure (35):	Alignment of SA stacks for analysis of ventricular volumes on the horizontal long-axis.....	55
Figure (36):	The short-axis (SA) cine stack is acquired from the base to the apex (not all views are shown)	55

Figure (37):	The 4-chamber cine is planned from the short-axis cine (<i>Left</i>) planning image; (<i>right</i>) resulting image	56
Figure (38):	The 3-chamber cine is planned from the short-axis cine (<i>Left</i>) planning image; (<i>right</i>) resulting image	57
Figure (39):	The 2-chamber cine is planned from the short-axis and 4-chamber cines (<i>Left</i>) planning image; (<i>middle</i>) planning image; (<i>right</i>) resulting image	57
Figure (40):	Myocardial biopsy showing individual myocyte hypertrophy with multiple deposits of brown granular material within the cytoplasm of the myocyte confirming heavy myocardial iron deposition (left image).....	59
Figure (41):	T2 images of the heart and liver in a normal volunteer and in a patient with thalassemia.....	61
Figure (42):	Short axis T2 image of the heart showing one large region of interest placed in the left ventricular septum for T2 evaluation	62
Figure (43):	A typical image shows the full-thickness ROI in the interventricular septum for T2* measurement. ROI, region of interest.....	62
Figure (44):	Decay curves from the liver in two patients with `thalassemia, one patient with high liver iron content (red decay curve) and one with lesser liver iron content (green decay curve).	64

Figure (45):	A typical case showing T2* measurement from the bright-blood and black-blood techniques.	65
Figure (46):	Myocardial iron overload.	66
Figure (47):	T2 iron content color maps of the liver and heart in a normal volunteer and in a patient with β -thalassemia.	68
Figure (48):	Heart plot chart	70
Figure (49):	Liver plot chart.....	71
Figure (50):	Example of ex vivo cardiac T2* scans.	75
Figure (51):	MRI of myocardium and liver of a thalassemic patient with high (top) and low (bottom) iron deposition.	77
Figure (52):	Cardiac MRI in patients with thalassemia major. A. Severe liver and heart iron. B. Severe liver iron and normal heart iron	78
Figure (53):	A 16-year-old male with β -thalassaemia major. a T2 map of the short axis of the heart shows low values of the left ventricle and the septum	79
Figure (54):	Cardiomyopathy in β -thalassemia.....	80
Figure (55):	A: short-axis, gradient-echo imaging sequence shows myocardial signal before and after treatment.....	81
Figure (56):	A (diastole) and B (systole): cine magnetic resonance sequence in four-chamberview.....	82
Figure (57):	A TM patient of 5.5 years old with detectable cardiac iron overload.....	83

Figure (58):	Patient referred for cardiac MRI to investigate suspected pericarditis.	84
Figure (59):	Pericardial thickening in constrictive pericarditis (arrows).....	84
Figure (60):	Loculated pericardial effusion (arrows) demonstrated with cine CMR images in orthogonal planes.....	85
Figure (61):	Coronal single-slice (MR) gradient echo images of heart with (ROI) in the anterior (A), apical (B), and inferior (C) segments of left ventricle (LV).....	86
Figure (62):	Cardiac magnetic resonance imaging of a patient with pulmonary hypertension.	88
Figure (63):	Pulmonary hypertension (PHT): (SSFP) cine MRI in the chamber view in diastole and systole, axial (MIP) pulmonary of MR-angiography, and delayed myocardial enhancement (inversion recovery sequence after contrast agent administration). Short axis view	88
Figure (64):	Left ventricular (LV) short axis using gradient-echo sequence with multiple echo times (TEs) for evaluation of T2*.....	89
Figure (65):	Comparison of two patients with different iron loading profiles and ventricular function.	90
Figure (66):	Iron chelation therapy for patients with thalassemia with cardiac iron overload.	94

Figure (67): A 35 year old male with beta-thalassemia major, regularly transfused since the age of 30 months, started chelation treatment with subcutaneous desferrioxamine at the age of 4 years... ..	95
Figure (68): Cardiac and liver T2* iron load measurements.	96
Figure (69): MR gradient echo images of differential tissue iron clearance before and during intravenous chelation therapy	97

Introduction

Thalassemia is the commonest single gene disorder worldwide, with approximately 94 million heterozygotes for beta thalassemia and 60,000 homozygotes born each year (*Weatherall et al., 1996*).

Although survival is improving in cohorts of patients in whom deferoxamine was introduced at a younger age, myocardial siderosis resulting in heart failure remains the major cause of death (50–70%) in thalassaemia major patients. This occurs at a strikingly young age with between 15–50% dying by the age of 35 years (*Modell et al., 2000*).

Some thalassemia centers have used liver iron concentration by biopsy as their gold standard for chelation titration (*Olivieri, 2001*). The liver is the dominant iron storage organ and liver iron concentration correlates closely with the total iron balance (*Angelucci et al., 2000*). Elevated liver iron also prospectively predicts poor endocrine and cardiovascular outcomes in patients with thalassemia major (*Telfer et al., 2000*).

Unfortunately, liver biopsy is invasive, expensive and subject to sampling error (*Butensky et al., 2005*). Although the risk of ultrasound-guided liver biopsy is relatively low, hemorrhage requiring prolonged hospitalization occurs in