

Institute of postgraduate childhood studies Department of childhood medical studies

Role of Echocardiography in Congenital Heart Diseases in Neonatal Intensive Care Unit

Thesis

Submitted for Partial Fulfillment of Master degree in Childhood Studies

(Department of Medical studies - Child Health and Nutrition)

Presented By

Reem Mamdouh Soliman Mahmoud Khattab

M.B.B.CH-Ain Shams University

Supervisors

Dr. Nayera Ismail Attia

Professor of Pediatrics

Institute of Post-Graduate Childhood Studies

Medical Studies Department - Ain Shams University

Dr. Hebatalla Mohamed Attia

Assistant professor of Cardiology
Faculty of Medicine - Ain Shams University

2012

بسم الله الرحمن الرحيم " قالُوا سُبْحَانَكَ لا عِلْمَ لَنَا إلاّ مَا عَلَمْتَنَا اللهُ عَلَمْتَنَا إِنَّكَ أَنْتَ الْعَلِيمُ الْحَكِيمُ "

صدق الله العظيم

سورة البقرة - الآية (32)

Dedication

Dedicated with all affection to my mother, **Dr.Zeinab Elnaggar**, my brother **Eng.Amir Khattab**, my soulmate & husband **Dr.Ramy Mostafa**, my sister **Reham Eldeeb** and to all my friends

especially **Sara Abdallah**, **Reem Zakaria**, **Ahmed Ramzy**, **Ghada Ghait**.

Dedicated also to Elzeitoon Medical Region & Saray Elkoba Medical Center especially **Dr.Suzan Abdelazeem**, **Dr.Safaa Hussein** and **Dr.Mohamed Elkashef**.

Dedicated to Atfal Masr Hospital, Emergency Department especially **Dr.Mohamed Taha**, **all nurses & workers** and all my collegues in the hospital especially **Miada Said**, **Mai Ghazy**, **Ahmed hashim**, **Ahmed Elkhawaga**, **Amira Ahmed**.

** to my grandmother & father's soul...

Reem Khattab
2012

Acknowledgement

First and foremost, thanks are all to ALLAH

I would like to express my deep gratefulness & appreciation to **Dr**. **Nayera Ismail Attia**, Professor of Pediatrics, Institute of Post Graduate

Childhood Studies, Medical Studies Department, Ain Shams University for

providing me with precious guidance and meticulous supervision which is

beyond acknowledgement.

I would like to express my great indebtation to **Dr. Hebatalla Mohamed Attia**, assistant professor of Cardiology, Faculty of Medicine, Ain Shams

University for her help & support and useful comments.

I wish to express my sincere thanks & indebtation to **Dr. Hala Gaber EI Rabei**, Fellow in Neonatal Intensive Care Unit, Gynecology and Obstetrics

Hospital, in Shams University for her encouragement and help throughout this study especially in performing echocardiography.

Last but not least, I would like to express my sincere thanks to every person in the NICU obstetrics & gynecology hospital, Ain Shams University including the neonates & their families for their great help.

Reem Khattab
2012

Abstract:

Background: Congenital heart defects are among the most common major congenital anomalies, and they occur worldwide with an incidence of about 8–12/1,000 live births. Many infants die without the diagnosis of complex CHDs, especially in developing countries. Echocardiography is used postnatally in high-risk infants for the diagnosis or exclusion of congenital heart defects and for assessment of cardiovascular function

Aim: to detect the prevalence of simple and complex congenital heart diseases among neonates admitted to the neonatal intensive care unit of Obstetrics and Gynecology Hospital, Ain Shams University over five years and evaluate and assess the clinical indications of echocardiography and its nature in relation to echo findings in the neonatal intensive care unit.

Methods: Echocardiography assessment was performed according to symptoms and signs in a neonate suspected to have congenital heart disease. It comprised 446 neonates admitted to a neonatology unit over five years subdivided to three groups according to the results of echocardiography as regards absence of congenital heart disease (group 1) or presence of simple (group 2) or complex congenital heart disease (group 3).

Results: on studying the distribution of Timing of Echocardiography in all neonates, echocardiography was mostly done in the first week of life (73.8% of cases) especially group two neonate having simple CHD (55.4%). Also, the highest indication for doing echocardiography was the presence of a **murmur** (39.5%), the presence of **non cardiac congenital anomalies** (22.6%) or **bad medical maternal history** (17%) and **cyanosis** (7.8%) with high statistical difference in group three neonates who had complex congenital heart diseases (P<0.01). As regards the most common echocardiographic finding in neonates with simple CHD, there was increased cases had PFO+PDA (37.3%), atrial septal aneurysm + PDA (15%), PFO alone (7.9%) and for complex CHD, there were increased cases of D-TGA (26.9%), Fallot's tetralogy (19%), and Hypoplastic left heart syndrome (14.3%).

Conclusion: Cardiac echocardiogram in neonates suspected having congenital heart diseases shows that prevalence of CHD (simple and complex) was 90.3 %.

Keywords

Echocardiography – congenital heart - neonatal intensive care unit

Contents

Title	Page
List of abbreviations	i
List of figures	iii
List of tables	V
Introduction	1
Aim of the study	5
Review of the literature	7
Chapter 1: Embryology of the cardiovascular system	8
Chapter 2 : Congenital heart disease (CHD) & its classification	25
Chapter 3: Etiology of CONGENITAL HEART DEFECTS	37
Chapter 4: Assessment of a child with suspected congenital heart	
diseases	
Chapter 5 : Echocardiography	55
Patients & Methods	82
Results	88
Discussion	108
Summary	119
Conclusion	125
Recommendation	127
References	129
Arabic summary	147

List of Abbreviations

AP	aorticpulmonary
AS	Aortic stenosis
ASD	Atrial septal defect
AV	atrioventricular
CAVC	complete atrioventricular canal defect
CCHDs	critical congenital heart defects
CHD	Congenital heart diseases
CHF	Congestive heart failure
CoA	Coarctation of aorta
DCM	dilated cardiomyopathy
DNA	Deoxyribose nucleic acid
DORV	Double-outlet right ventricle
D-TGA	D-Transposition of great arteries
ECG	electrocardiogram
EF	ejection fraction
HF	heart failure
HCM	hypertrophic cardiomyopathy
HLHS	Hypoplastic Left Heart Syndrome
HOCM	Hypertrophic obstructive cardiomyopathy
IVC	Inferior vena cava
LV	Left ventricle
LVED	left ventricular end-diastolic dimension
LVES	left ventricular end-systolic dimension
LVEDV	left ventricular end-diastolic volume
LVESV	left ventricular end-systolic volume
LVPW	left ventricular posterior wall
NICU	Neonatal intensive care unit
PA	Pulmonary atresia
PA-IVS	pulmonary atresia with intact interventricular septum
PDA	Patent ductus arteriosus
PFO	Patent foramen ovale
PPHN	Persistent Pulmonary Hypertension of the Newborn
PS	Pulmonary stenosis
PVR	pulmonary vascular resistance

RD	Respiratory distress
RV	Right ventricle
SA	sino-atrial
SF	shortening fraction
SV	single ventricle
SVC	superior vena cava
TA	Tricuspid atresia
TAPVR	Total anomalous pulmonary venous return
TEE	Tranesophageal echocardiography
TGA	Transposition of great arteries
ToF	tetralogy of Fallot
VSD	Ventricular septal defect

List of figures

FIG.NO.	Name	Page
1	Transverse section of embryo after removal of amnion showing the	9
	position of cardiogenic field and pericardial cavity	
2	The effects of rapid growth of brain on position of the heart	9
3	Formation of cardiac loop and its bending inside the pericardial	10
	cavity	
4	Left and front view of heart showing division of bulbous cordis	11
	into truncus arteriosus, conus-cordis, and trabeculated portion of	
	right ventricle	
5	Formation of the atrial septum	12
6	Formation of the septum in the atrioventricular canal	13
7	Formation of atrioventricular valves (AV) and chordae tendinae.	14
8	Formation of muscular part of interventricular septum	15
9	Development of conotruncal swellings and closure of	16
	interventricular foramen	
10	Aortic arch development	17
11	Fetal circulation before birth	20
12	Human circulation after birth	24
13	evaluation of undiagnosed cardiac murmurs	46
14	The differential diagnosis of cardiac exam findings in acyanotic	51
	neonates	
15	The differential diagnosis of cardiac exam findings in cyanotic	52
	neonates	
16	The differential diagnosis of chest radiographic and	53
	electrocardiographic findings in acyanotic neonates.	
17	The differential diagnosis of chest radiographic and	54
	electrocardiographic findings in cyanotic neonates	
18	Gross anatomy of the heart	59
19	thoracic imaging landmarks	60
20	various transducer locations in echocardiography	62
21	subcostal view	64
22	Apical two chambers	64
23	Apical four chamber view	65
24	Left parasternal long axis	66
25	Short axis view aortic valve	66
26	Short axis view left ventricle	67
27	suprasternal short –axis view & suprasternal aortic (Ao) arch	68
	(long-axis) view	
28	The left ventricle	72
29	A dilated, hypokinteic right ventricle (RV) is demonstrated using	76
	three dimensional echocardiography	
30	Ballard score	85

FIG.NO.	Name	Page
31	Frequency distribution of echocardiographic findings among all	91
	neonates	
32	Frequency distribution of Timing of Echocardiography in all groups	92
33	Frequency distribution of birth weight for all patients	93
34	Frequency Distribution of different diagnoses among all cases:	94
35	Comparison between both groups as regards indications of admission to NICU	95
36	Comparison between the three groups as regards associated maternal & obstetrical conditions among all neonates:	97
37	Frequency distribution of Echocardiographic Indications among neonates under study	98
38	Echocardiographic findings among group 3 (complex congenital heart defects)	103
39	Neonatal echocardiography of neonate had D-TGA	106
40	Neonatal echocardiography of neonate had Fallot tetralogy	106
41	Neonatal echocardiography of neonate had PFO	107

List of Tables

Table	Name	Page
no.		
1	Relative Frequency of Major Congenital Heart Lesions	27
2	Chromosomal anomalies associated with heart defects	37
3	Prevalence of Associated Congenital Heart Defects in Patients with	38
4	Other System Malformations	20
4	TERATOGENS ASSOCIATED WITH HEART DEFECTS	39
5	INDICATIONS FOR FETAL ECHOCARDIOGRAM	40
6	Segmental approach to defining cardiac anatomy by echocardiography	70
7	Descriptive data of all patients	90
8	Frequency distribution of Timing of Echocardiography in all groups	91
9	Comparison between the three groups as regards weight	92
10	Frequency distribution of clinical findings among all neonates on admission:	93
11	Frequency Distribution of different diagnoses among all neonates	94
12	Comparison between the three groups as regards indications of admission to NICU	95
13	Comparison between the three groups as regards associated maternal & obstetrical conditions among all neonates	96
14	clinical indications of echocardiography among all cases	97
15	Comparison between all groups as regards indications for echocardiography	99
16	Comparison between term and preterm neonates as regards indications for Echocardiography	100
17	Echocardiographic findings among group 2 (simple congenital heart defects)	101
18	Echocardiographic findings among group 3 (complex congenital heart defects):	102
19	Frequency d2istribution of echocardiographic findings among term and preterm neonates	104
20	Prevalence of echocardiographic findings among cases with bad maternal & obstetric history:	105



Introduction

Congenital heart diseases (CHD) are relatively common with a prevalence ranging from 3.7 to 17.5 per 1000 live births (**Bolisetty**, **2004**).

It can be defined as a structural abnormality of the heart or intrathoracic great vessels which is actually or potentially of functional significance. It represents a spectrum of conditions, from those that may be fatal in the neonatal period, to those with which a normal lifespan would be expected (Clarke, 2005).

Most deaths from congenital heart defects occur in the first year of life and these are most likely to be related to extra-cardiac anomalies, cardiovascular collapse during the changes from fetal to newborn physiology, heart failure (Bache, 2002).

The relationship between congenital heart disease, malnutrition, and growth retardation is well documented. Infants with congenital heart disease are prone to malnutrition for several reasons including decreased energy intake, increased energy requirements, or both. Different types of cardiac malformations can affect nutrition and growth to varying degrees (Gilger, 1990). For these defects, timely recognition in the newborn period is vital to prevent death or cardiovascular collapse with its attendant morbidity (Hall, 2003).

Current guidance recommends a routine clinical examination for all newborns and again at 6–8 weeks of age (Hall, 2003).

In the neonatal period a diagnosis of CHD may be considered for two reasons: (1) a heart murmur or other cardiovascular abnormality identified in an asymptomatic infant or (2) the development of symptoms and signs that could be attributable to congenital heart diseases (Clarke, 2005).

In one population-based study, more than half of babies with undiagnosed congenital heart defects were missed by routine neonatal examination and more than one-third by 6 weeks (**Wren**, 1999). One strategy to avoid this is to advance the time of diagnosis from postnatal to fetal life (antenatal screening) by ultrasound in the early 1980's which gives the parents an opportunity for information and counseling with options for a planned delivery and intervention or termination of pregnancy (**Bricker**, 2000).

Although lesions diagnosed antenatally many are by ultrasonography, serious and potentially lethal critical congenital heart defects (CCHDs) may not be apparent on prenatal ultrasound, on subsequent physical examination after birth, or on follow-up after discharge. When detected early, CCHDs are either cured or ameliorated by surgery in the vast majority of cases; therefore, a universal screening test for CCHD would be beneficial if it were demonstrated to have acceptable sensitivity and specificity and to offer information that could not be provided by routine examination and observation (Richmond, 1999).

Echocardiography has become the most important non-invasive tool in the diagnosis and management of cardiac diseases. Two dimensional echocardiography provides a full anatomical evaluation in most congenital heart defects. Physiologic data on the direction of blood flow can be obtained with the use of pulsed wave, and color flow Doppler (Daniel, 2006).

Prenatal or fetal echocardiography can diagnose congenital heart diseases by 18 weeks of gestation and this prenatal diagnosis allows for delivery in a well equipped place for such conditions where many congenital heart defects are surgically repaired based on the echocardiogram with no need for cardiac catheterization (**Daniel**, 2006).

It must be noted also that initial evaluation of a neonate with suspected congenital heart disease includes four-extremity blood pressure, chest x-ray, electrocardiogram and hyperoxia test (which is perhaps the most sensitive and specific tool in the initial evaluation) (Stephanie,2008).

The ability to identify affected newborn infants, when totally asymptomatic, and institute programs and treatments that prevent serious morbidity and mortality is a great privilege for the pediatrician (Wren, 2000).