Incidence of congenital heart disease among patients Referred to Echocardiography Unit at Cairo University Children Hospital (CUCH)

Thesis

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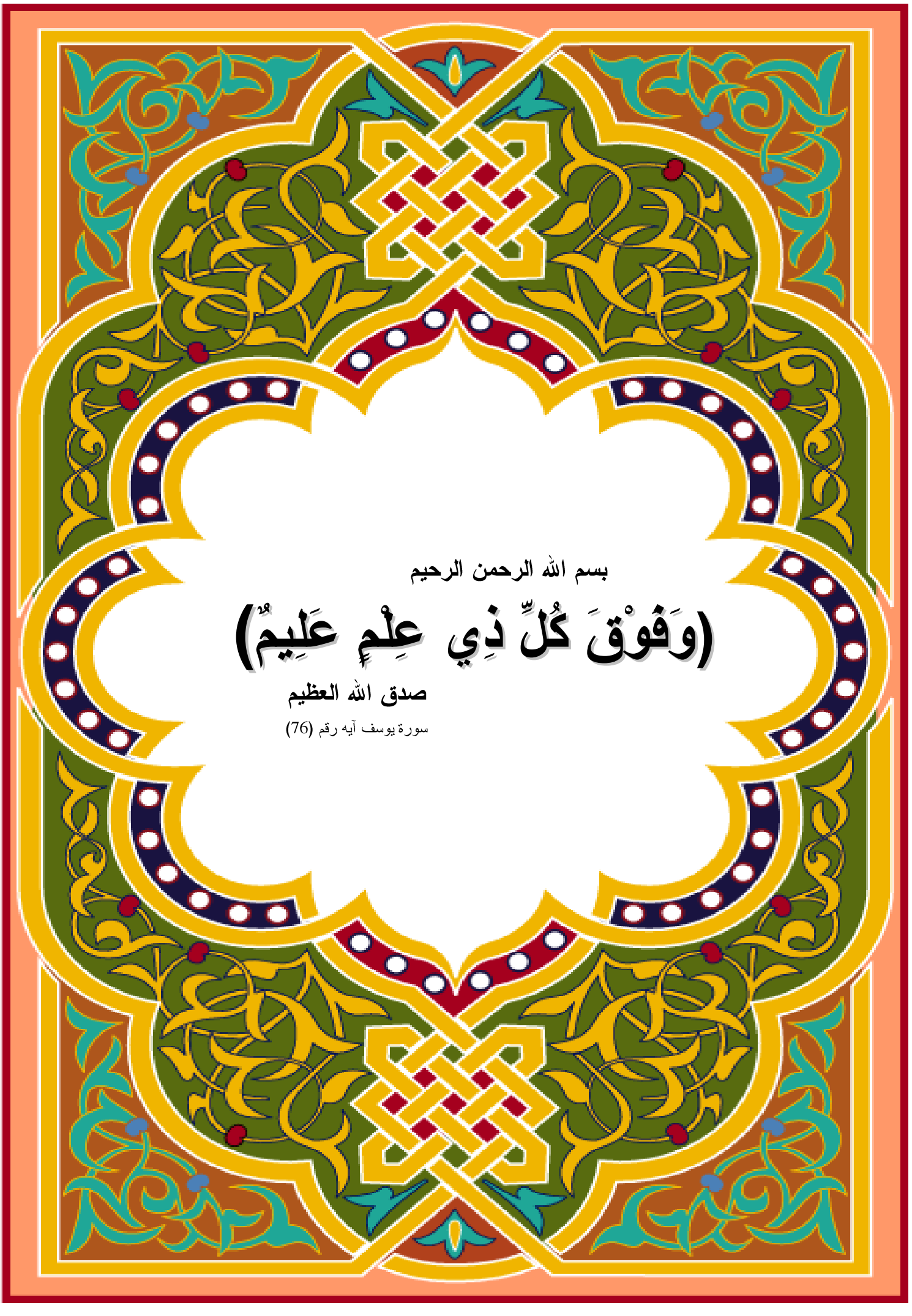

بسم الله الرحمن الرحيم

(وَفَوْقَ كُلِّ ذِي عِلْمٍ عَلِيمٌ)

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Abstract

Key Words: congenital heart disease– echocardiography

Observational study was conducted to determine the Incidence and relative frequency of congenital heart diseasediagnosed by echocardiography Unit in pediatric Hospital, Cairo University Children Hospital (CUCH) over a period of 1 year (1 January 2013-31 December 2013).

Congenital heart diseases were the most frequent cardiac abnormalities (41.9%).

Majority of the cases (69.7%) were Acyanotic, remaining(30.3%) were cyanotic. Among the Acyanotic VSD (14.87 %*) was the commonest 2nd was ASD (13.98 %*) while in acyanoticgroup D-TGA (6.65 %*) was the commonest followed by TOF (6.43 %*).

Male preponderance,the most clinical presentation was Breathlessness and poor weight gain,majority of case was sever CHD are important observation made in the study.

*. %: % of CHD.

CONTENTS

List of Abbreviations

ACHD	Acyanotic congenital heart disease
ASD	Atrial septal defect
APV	Absent pulmonary valve
AS	Aortic stenosis
AVSD	Atrioventricular septal defect
AVC	Atrioventricular Canal
BAV	Bicuspid aortic valve
BCAV	Bicuspid aortic valve
BT shunt	Blalock–Taussig shunt
CAD	Coronary artery disease
CCHD	Cyanotic congenital heart disease
ccTGA	Congenitally corrected TGA
CHD	Congenital heart disease
COA	Coarctation of aorta
CW	Continuous wave
DORV	Double outlet right ventricle
D-TGA	Dextro-Transposition of the great arteries
ECHDO	European congenital heart disease organization
EA	Ebestien anomaly
***E/A* ratio**	The ratio of the early (E) to late (A) ventricular filling velocities
ED	End diastolic
ESPAP	Estimated systolic pulmonary artery pressure
EF	Ejection fraction
ET	Ejection time
FISH	Fluorescence in situ hybridization
ES	End systolic
FS	Fractional shortening
HLHS	Hypoplastic left heart syndrome
HR	Heart rate
ICT	Isovolumic contraction time
IRT	Isovolumic relaxation time
IVS	Inter ventricular septum
LA	Left atrium
LVEDD	Left ventricle end-diastolic diameter
LVEF	Left ventricular ejection fraction
LVESD	Left ventricle end-systolic diameter
LVIDd	Left ventricle internal diameter in diastole
LVIDs	Left ventricle internal diameter in systole
LV	Left ventricle
LVPW	Left ventricle posterior wall
LVOT	Left ventricular outlet
MDM	Maternal diabetes mellitus
MMHG	MilliMeters of Mercury
MPI	Myocardial performance index
MV	Mitral valve
NYHA	New York Heart Association Classification
PA	Pulmonary artery

PA	Pulmonary atresia
PAPVD	Partial anomalous pulmonary venous drainage
PBF	Pulmonary blood flow
PBS	Pulmonary branch stenosis
PDA	Patent ductus arteriosus
PFO	Patent foramen ovale
PGE_1	Prostaglandin E1
PH	Pulmonary hypertension
PPHN	Persistent pulmonary hypertension
PRF	Pulse repetition frequency
PS	Pulmonary stenosis
PTA	Persistent truncus arteriosus
PW	Pulsed wave
Qp:Qs	Pulmonary-Systemic Flow Ratio
RV	Right ventricular
RVOT	Right ventricular outflow tract
RHD	Rheumatic heart disease
TA	Tricusped atresia
TAPVR	Total anomalies pulmonary venous return
TCAV	Tricuspid aortic valve
TGA	Transposition of great arteries
TOGV	Transposition of the great vessels
TOF	Tetralogy of Fallot
TV	Tricuspid valve
TYPE 1a	Normally related great vessels with pulmonary atresia
TYPE 1b	Normally related great vessels with pulmonary stenosis
TYPE 2a	D-transposed of the great arteries with pulmonary atresia
TYPE 2b	D-transposed of the great arteries with pulmonary stenosis
TYPE 3a	L-transposed of the great arteries with pulmonary atresia
TYPE 1c	Normally related great vessels
TYPE 3c	L-transposed of the great arteries
UCAV	Unicuspid aortic valve
VPS	Valvular pulmonary stenosis
VSD	Ventricular septal defect
SAM	Subaortic membrane
SV	Single ventricle
SVD	Single ventricle defect

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INTRODUCTION

The normal heart is like a three store building (atria, ventricles, and great arteries): the atria are the platform, the ventricles are the first floor, and the great arteries are the second floor, connected to each other through valve orifices and totally separated by septa. The segments are disposed in such a way as to allow deoxygenated venous blood to go to the lungs through the pulmonary artery, and oxygenated venous blood to go to the systemic organs through the aorta. Small and great circulations are in sequence, with no communication with each other apart from the lung capillary network***(Thiene and Frescura, 2010)***.

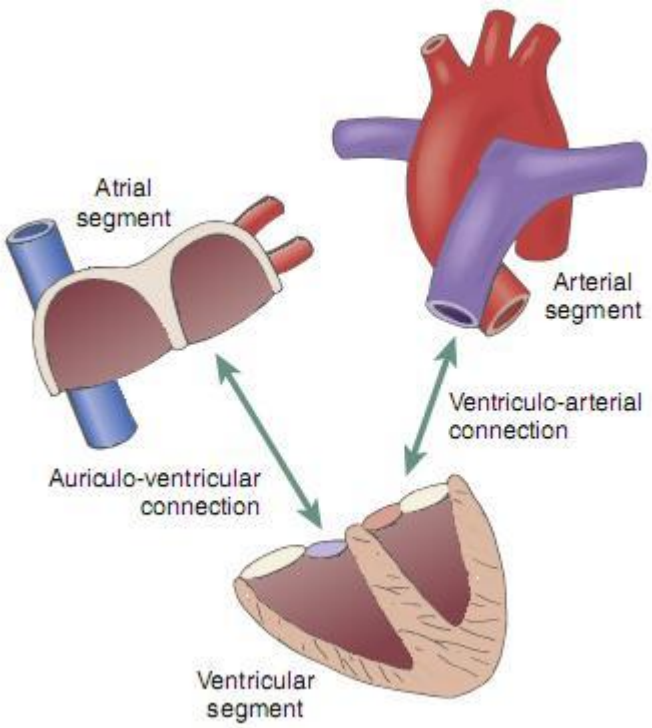

Figure {1}: Schema of segmental approach: heart is divided into three segments (atria, ventricles, and atreial trunks) and two junctions (atrio-ventricular and ventriculo-arterial junctions) ***(Bettex&Chassot, 2011).***

Congenital is derived from the Latin con, together, and genitus, born. However the implication that CHD simply means "present at birth" require elaboration. The natural history begins before birth. The majority of congenital anomalies of the heart are present six week after conception, and most anomalies compatible with six months of intrauterine life permit live offspring at term. A given malformation may exist in relative harmony with the fetal circulation only to be modified considerably, at least physiologically, by the dramatic circulatory adjustments at birth. Weeks, months or years may elapse before the anomaly reveals itself as the "typical" clinical picture. Pathophysiological and structural changes continue or, conversely, the malformation may "vanish" ***(Perloff, 1998).***

Congenital heart disease (CHD) is a complex problem which can affect the patient, the family, the society and the country. CHD describes a wide range of conditions resulting from abnormalities of the heart structure and/or function that is present from birth. The severities of conditions are classified as complex, moderate, and simple***(Marelli et al, 2007)***. CHD is the most common birth defect, with approximately 9 in 1,000 new borns being affected***(Van der Linde et al, 2011).***Although CHD is the most common survivable birth defect, the etiology of most CHD remains unclear***(Yuan et al, 2013)***.It represents one of the most common causes of death of children at different ages starting from abortion of the fetus, stillbirth, death of the patient at birth, during infancy, during early childhood, during late childhood and may at adulthood***(Ferencz et al, 2008)***.

From a surgeon's perspective, CHD consists of shunts, obstructions, valvular regurgitation, or combinations of these. Operative interventions are ingenious and numerous, and they encompass complete or partial repairs, single and multistage palliative procedures, and cardiac transplantation***(Edwards, 2010)***.

Cardiac imaging plays an important role in establish the diagnosis, interventional management;follow up after palliative or corrective surgery. Various imaging modalities for the diagnosis of CHD progress from plain chest radiograph, 2-dimensional echocardiography and conventional cardiac catheterization to noninvasive advanced cardiac imaging, 3- and 4- dimensional echocardiography, transesophagealechocardiography, cardiac MRI (cMRI), andmultidetector CT (MDCT). Echocardiography remains a first line noninvasive imaging tool for establishing the diagnosis and follow up in most patients. However, echocardiography has inherent limitations, including a limited acoustic window. This is particularly the case with postsurgical sternal wires and mediastinal scar tissue, and extracardiac vascular structures. Echocardiography is also an operator dependent imaging tool with poorer spatial resolution than CT or CT angiography(CTA)***(Bailliard et al,2008)***.

CHD was considered a predominantly pediatric condition, as patients with complex forms seldom survived beyond childhood. However, patient care markedly improved, resulting in a growing and aging population. Indeed, adults now outnumber children with both severe and other types of CHD***(Marelli et al, 2007)***.

Ultimately, the goals of pediatric research and clinical care are to maximize health and minimize symptomatology, disability, and dysfunction that may impact the lives of children with acute and chronic disease processes. Over the last several decades, new surgical techniques and advances in cardiopulmonary bypass, intensive care, interventional cardiac catheterization, non-invasive imaging, and medical therapies have significantly lowered neonatal mortality rates for children with the most complex congenital heart disease (CHD) (e.g. hypoplastic left heart disease) to less than 10%***(Tweddell et al, 2002).***

CARDIOVASCULAR EMBRYOLOGY

Commitment to the cardiogenic cell lineage occurs early in development soon after gastrulation (the embryonic state following the blastula), approximately 48h following fertilization ***(Khan, 2006)***.

Heart is formed in a fashion where components are added in a sequence to an initial primary structure, the cardiac crescent. The first step in the heart development is the formation of two heart fields of precardiac mesoderm, which are situated on opposite sides of the primitive foregut situated at the embryonic midline and contain endocardial as well as myocardial precursor cells. The first or the posterior heart field contributes to the bulk of the left ventricle and the secondary or the anterior heart field forms the right ventricle, outflow tracts, sinus venosus and atrial chambers ***(Bruneau, 2008)***.The first evident assembly of the mesoderm-derived myocardial (or cardiogenic) plate is seen at 18 days, followed by initiation of heart tube formation at 22 days***(Mitchell et al, 2007)***which connects at the cephalic end with the developing aortic arch system, and caudally with the vitelline and umbilical venous systems ***(Reller et al, 1991)***.