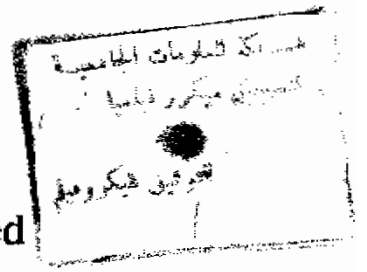
ENDOCARDIAL CUSHION DEFECTS

Thesis

Submitted in partial fulfilment of
Master Degree in Cardiology



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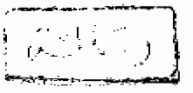

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بسم الله الرحمن الرحيم

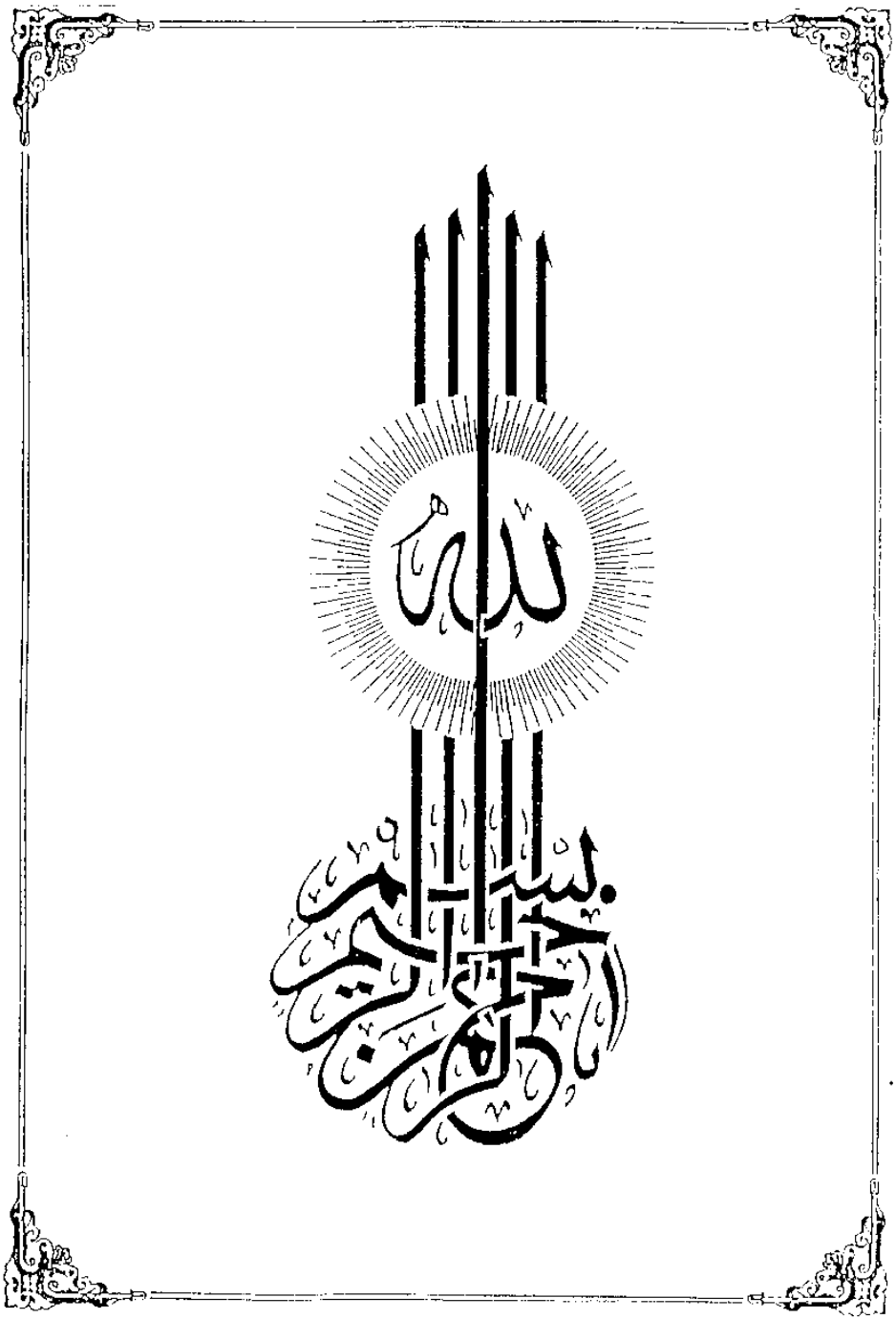
اقْرَأْ بِاسْمِ رَبِّكَ الَّذِي خَلَقَ

خَلَقَ الْإِنسَانَ مِنْ عَلَقٍ

اقْرَأْ وَرَبُّكَ الْأَكْرَمُ الَّذِي عَلَّمَ بِالْقَلَمِ

عَلَّمَ الْإِنسَانَ مَا لَمْ يَعْلَمْ

العلق ١-٥



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CONTENTS

INTRODUCTION

Endocardial cushion defects are wide spectrum cardiac anomalies which include ostium primum atrial septal defect with cleft mitral with or without tricuspid septal leaflet cleft, ventricular septal defect and defects in atrioventricular valves. It was found that about 3% of infants and children with congenital heart disease have endocardial cushion defect 60 - 70% are of the partial (incomplete) type, and 25 - 36% are of the complete type.

A female to male ratio is approximately 1.3: 1.

Over half of the patients with the complete type have associated Down's syndrome. In most cases, it is a serious disease and highly fatal if left untreated.

If the complete type is not complicated by other major defects, death often occurs before 15 years of age (avergae at age of 2 years). But if other major defects are present, death occurs earlier within the first year of life.

AIM OF THE WORK

The aim of this work is to study the variability inthe clinical presentation of endocardial cushion defects in relation to the anatomical and haemodynamic findings.

Cases diagnosed as endocardial cushion defects are proved by cardiac catheterization in Ain Shams University Hospitals during the last five years will be studied.

For each patient, we will study the following parameters:

1- History and clinical examination.

2- 12 leads surface electrocardiogram.

3- Chest X-ray.

4- Haemodynamic findings.

5- Echocardiographic and echodoppler.

6- Angiographic findings.

Finally, the review will throw light on the prognosis and possible lines of treatment.

Fig. (1): Photographs of the right and left sides of a normal child's heart to show the components of the mitral (a) tricuspid (b) valves and the distribution of the endocardial cushion tissue in them. The solid dark area in (b) is the membranous septum (Alomeri, 1965).

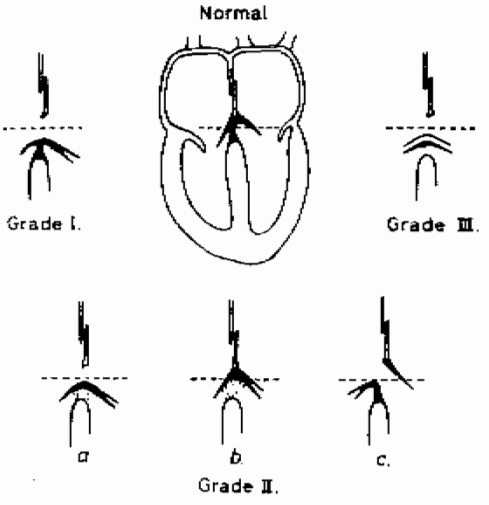

Figure (2): Diagramatic sections in the frontal plane to show the normal sites of fusion of the endocarial cushion tissue compared with the situation in the three grades of this type of defect. The endocardial cushion tissue is drawn in solid black. In the centre is the normally formed heart showing fusion of the endocardial cushions with the atrial and ventricular septa above and below. The horizontal dotted line (drawn at the same level in each diagram) shows the normal level of attachment of the atrioventricular (AV) valves. Note that the mitral valve (on the right of the diagram) is normally higher than the tricuspid valves. The region of the atrio-ventricular canal alone is shown enlarged in the other diagrams. The ostium primum defect with its cleft and inferiorly displaced mitral valve is shown in the upper left-hand corner. The transitional defects are depicated below. Grade IIa shows an ostium primum defect where there is also abnormal attachment of the chordae to the top of the ventricular septum (the tricuspid valve may also be cleft or otherwise deficient). Grade IIb is entirely an interventricular communication, the isolated ventricular defect of AV canal type or "cushion VSD". Abnormal chordae are shown and the mitral valve may be clefted (the tricuspid valve may or may not be similarly abnormal). Grade IIc shows the stuations in which the defect is in that part of the membranous septum that separates the left ventricle from the right atrium. The mitral valve is normal, but the tricuspid valve is abnormal and often cleft. In the upper right hand corner is shown the complete AV canal in which all 4 cardiac chambers are in communication with each other and the single AV valve straddles the defect, but is not tethered to the top of the ventricular septum (Alomeri, 1965).

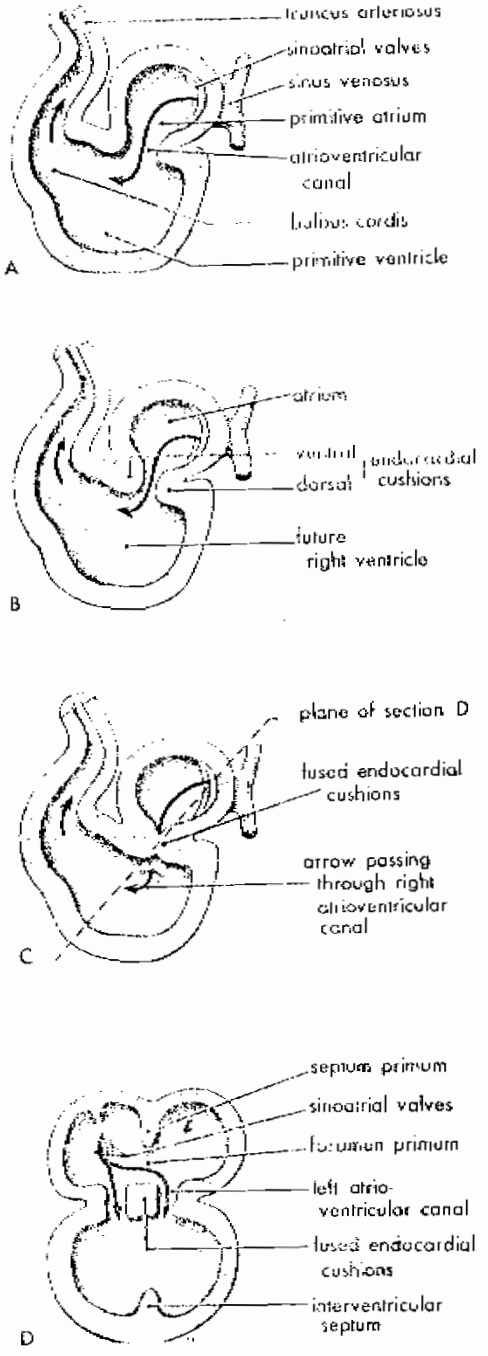

Fig. (3): From (A) to (C), sketches of sagittal sections of the heart during the 4th and 5th weeks, illustrating division of the atrioventricular canal. (D), Coronal section of the heart at the plane shown in (C). Note that the interatrial and interventricular septa have also started to develop (Eidmiller, 1988).

On the left side, the left portions of the anterior and posterior cushions Join to form the septal leaflet of the mitral valve with subsequent rotation of the ventricles, this cusp assumes a relatively anterior position. The formation of the tricuspid valve is more complex. The anterior and right lateral cushion origin, the lateral cushion itself contribute only minimally to the anterior leaflet and the medial portion of this leaflet, as will as the medial papillary muscle and chordae tendineae are derived from the conotrunkal septum. The right side of the posterior endocardial-cushion contributes to the medial tricuspid valve leaflet and also completes the closure of the most posterior of the membranous ventricular septum. The lower edge of the septum primum, which separates the atria, fuses with the centre of the anterior and posterior endocardial cushions, and as they grow toward each other and fuse, the orifice at the lower margin of the septum primum is sealed. As this process of closure of the interatrial septum proceeds, a series of formation develop in the central portion of the septum primum to maintain communication between the atria (Hurst, 1990).

Partitioning of the primitive atrium:

The primitive atrium is divided into right and left atria by the formation and subsequent modification and fusion of two septa, the septum primum and the septum

secundum. The septum primum also unites with the fused endocardial cushions.

Septum primum:

A thin, crescent-shaped membrane, grows toward the fusing endocardial cushions from the dorsocranial wall, or roof, of the primitive atrium. As this curtain-like septum grows, a large opening, the foramen primum, forms between its crescentic free edge and the endocardial cushions. The foramen primum becomes progressively smaller and disappears when the septum primum fuses with the fused endocardial cushions (atrioventricular septum).

Septum Secundum.

Before the foramen primum is obliterated, perforations appear in the dorsal part of the septum primum that coalse to form another opening, the foramen secundum. Concurrently, the free edge of the septum primum fuses with the left side of the fused endocardial cushions, obliterating the foramen primum. Toward the end of the fifth week, another crescentic membrane, the septum secundum, grows from the ventro-cranial wall of the atrium, immediately to the right of the septum primum. As this thick septum grows, it gradually overlaps the foramen secundum in the septum primum. The septum secundum forms and incomplete partition, forming an oval opening called the foramen ovale.

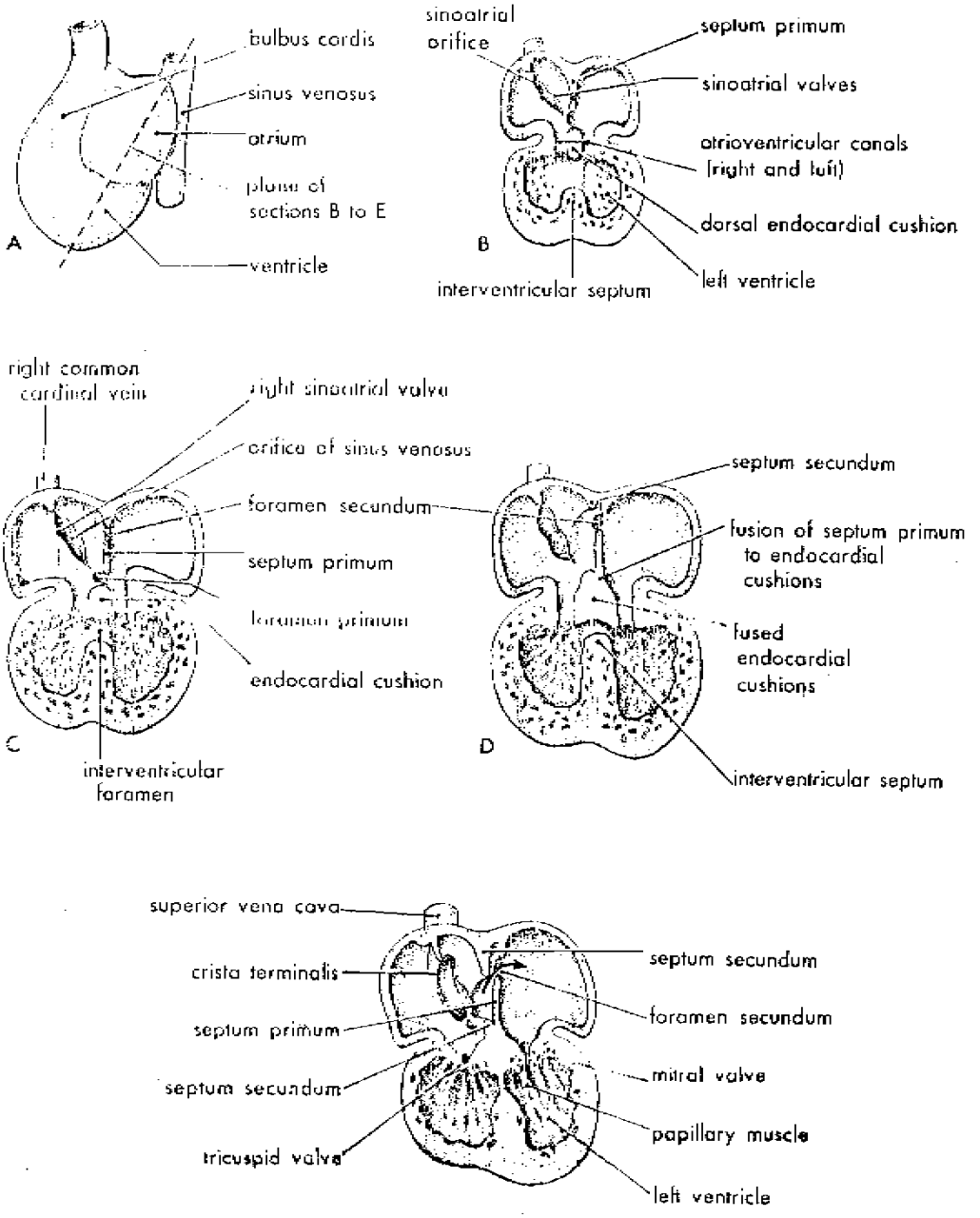

Fig. (4): Drawing of the developing heart, showing partitioning of the atrioventricular canal, primitive atrium and ventricle. A, sketch showing the plane of the coronal sections, (B), during the 4th week (about 28 days), showing the early appearance of the septum primum, interventricular septum, and dorsal endocardial cushion. (C), Section of the heart (about 32 days), showing perforations in the dorsal part of the septum. (D), section of the heart (about 35 days), showing the foramen secundum, (E), about eight weeks, showing the heart after it is partitioned into 4 chambers (Eidmiller, 1988).

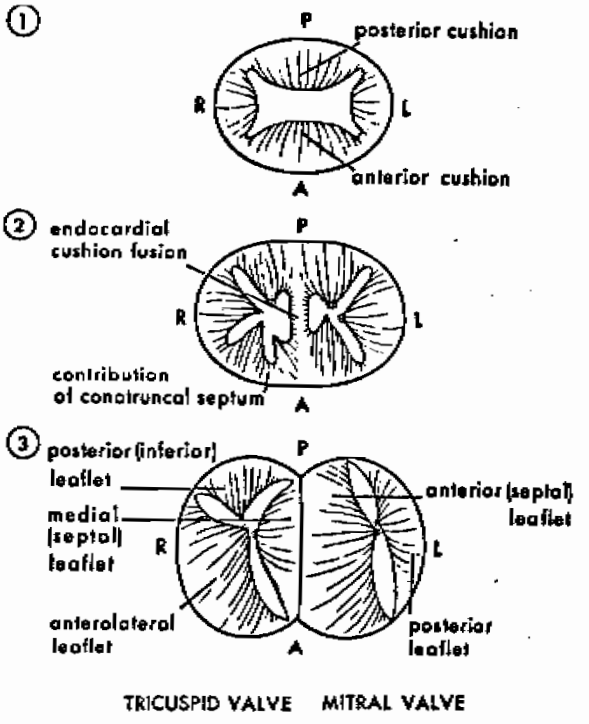

Figure (5): Diagram showing stages in the development of tricuspid and mitral valves from primitive endocardial cushions (Rudolph, 1974).

tissue from the right side of the fused endocardial cushions.

This tissue merges with the aortico-pulmonary septum and the thick muscular part of the interventricular septum. After closure of the interventricular foramen, the pulmonary trunk is in communication with the right ventricle, and the aorta communicates with the left ventricle (Eidemiller et al., 1988).

Anatomic features of the mitral valve:

In the normal mitral valve, the width of the base of the anterior leaflet is less than half of the posterior laflet by a ratio of about 1: 2 (Duplessis and Marchand, 1964). But this ratio can be disturbed where the width of the anterior leaflet at its base being equal to or greater than the corresponding part of the posterior leaflet. The leaflets of the mitral valve and the papillary muscles will be either normally oriented or rotated clockwise. This rotation appeared to have resulted from the abnormally posterior position of the anterolateral papillary muscle.

The cleft of the anterior mitral leaflet was usually shaped like an inverted "V" with the apex pointing towards the valve ring. But sometimes may be rectangular in shape.