SURGICAL MANAGEMENT OF AV CANAL MALFORMATIONS

ESSAY

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By

Mohamed Aziz Mohamed Ezzat

M.B.,B.Ch.

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SUPERVISED BY

Prof. Dr. Mohamed El-Fiky

Professor of Cardio-Thoracic Surgery

Ain Shams University

FACULTY OF MEDICINE

AIN SHAMS UNIVERSITY

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بسم الله الرحمن الرحيم

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INTRODUCTION

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AV canal malformations is not an uncommon congenital heart disease.

The spectrum of cases is wide including different anatomical variants.

It may coexist with other cardiovascular anomalies which modify the natural life history and the prognosis of the surgically treated patients. Also, it may coexist with other generalised congenital anaomalies as Down's syndrome or splenic anomalies.

The aim of this study is to identify these anatomical variants and to explain their embryologic basis, methods of diagnosis and different surgical techniques.

EMBRYOLOGY

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The vascular system makes its appearance in embryos of about three weeks age as scattered masses of cells called Angiogenic cells or blood islands. These cells form the primitive heart tube.

The wall of this tube consists of three layers, an external layer formed of one or two cell layers thick called external myocardial mantle, an internal layer formed of a single layer of endothelial cells. These two layers are separated from each other by a relatively thick acellular third layer called cardiac jelly, from which the cushion tissues develop.

This tube is called the bulbo-ventricular tube as it will form the embryonic ventricle and the bulbus cordis.

The growing bulbo-ventricular tube bends to the right and anteriorly forming the bulbo ventricular loop. The deepening concavity on the left side of the bulbo ventricular loop is called the bulbo ventricular or cono ventricular groove or sulcus.

The atrial portion of the heart dilates to form a large common atrium. The growth of the common atrium takes place in a dorso-cephalad direction, dorsal to the bulbo ventricular tube. The atrioventricular junction shifts cephalad with the growth of the atrium and is now called atrioventricular canal or AV canal (Hurst et al, 1982).

the myocardial and endocardial tubes of the bulbo ventricular loop differ in their segmentation. The myocardial tube consists only of two gross segments, the ventricle and the bulbus cordis.

The endocardial tube is convuluted in such a way that it forms five straight segments (limbs) and in between them four curves or bends. The proximal segment of the endocardial tube starting at the venous end is the AV canal. It runs from a cephalic posterior to a caudal anterior position. Next comes the first curve or proximal bend (pre-ampulla) in which the endocardial tube turns 90° to the right to form the proximal transverse limb or interventricular foramen (IVF). In the second curve, it turns 90° cephalically to form the ascending limb. Then it turns 90° medially to form the distal transverse limb. In the fourth curve it turns 90° backwards to form the terminal limb.

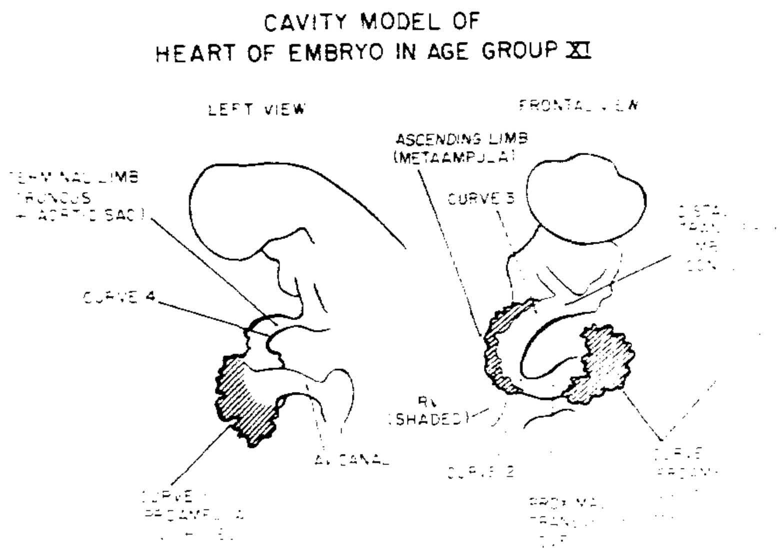

Fig. [illegible] : Endocardial heart tube segmentation
Goor and Lillehei, 197[illegible] .

The myocardial ventricle includes the proximal bend of the endocardium and part of the proximal transverse limb, while the bulbus cordis engulfs the remainder of the endocardial tube (Goor and Lillehei, 1975).

Atrio-Ventricular Cushions:

During AV canal expansion, the cardiac jelly becomes cellular, and is molded into 2 major, anterior and posterior and 2 small lateral atrio-ventricular cushions. The anterior cushion is attached to the roof of the AV canal and the posterior one is attached to the floor. As a result of protrusion of the cushions into the lumen of the AV canal, the flow is divided into two streams, mitral and tricuspid.

The ventricular aspect of these 2 cushions is concave, so that their lateral ends protrude into the ventricle like horns known as the tubercles. Hence, there are 2 right and 2 left tubercles, superior and inferior on each side. Fusion of the anterior and posterior AV cushions results in the formation of the septum of the AV canal. Also as a result of fusion of these cushions, the primary IVF acquires a dorsal wall which is the right end of the fused AV cushions (Goor and Lillehei, 1975).

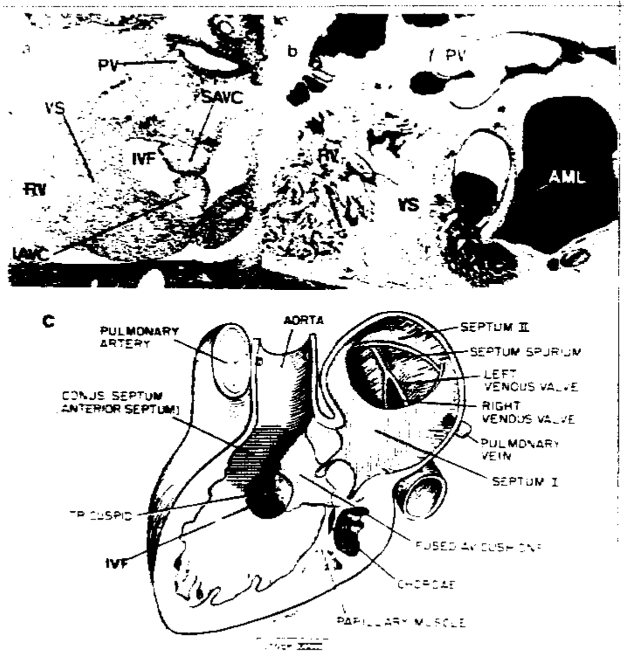

Fig.(2): AV canal partitioning, anterior mitral leaflet formation and semilunar cusp formation. A) Sagittal microsection of human embryonic heart passing through AV cushions and ventricular septum. Superior and inferior AV cushions (SAVC, IAVC) approach each other but are not fused. Pulmonary truncus (valve, PV) is still filled with truncal mesenchymal ridges. B) Sagittal microsection of human embryonic heart projected plane is same as in A. Anterior and posterior AV cushions are fused forming anterior mitral leaflet (AML). Pulmonary truncal mesenchymal ridges are hollowed out with shape of semilunar cusps and related sinuses of valsalva. C) Left ventricular view diagram of developmental stage between A and B. Right extreme end of fused AV cushions provides a dorsal wall for IVF. (Goor and Lillehei, 1975).

The atrioventricular endocardial cushions contribute to the formation of the atrioventricular septum, the membranoùs interventricular septum, and the AV valves. They also aid in clossure of the ostium primum (Van Mierop et al, 1962).

Development of the Ventricular Septum:

During the 5th week, the right and left definitive ventricles are indicated as slight projections on the external surface of the primitive common ventricle. It is uncertain whether the right ventricle is a derivative of the common ventricle or of the caudal end of the primitive bulbus or of both. In either events, the appearance of a crescentic ridge in the inside of the heart indicates the separation between the two ventricles and as the heart enlarges, this ridge deepens to form the muscular septum. This septum has a free sickle-shaped margin. The dorsal and ventral horns of the septum meet and fuse with the corresponding endocardial cushions of the AV canal near their right extremities.

The conotruncus moves from its initial far lateral position to a more medial location to lie in a mid sagittal plane in a depression of the atrial roof between the primitive right and left atria.

Also the AV canal moves from left to right to lie in a mid sagittal plane, so that, the right extremity of the canal comes to lie caudal to the orifice of the truncus.

Partitioning, rotation and absorption of the conotruncus occurs resulting in fusion of the septum aorto pulmonale and the trunkal septum. Also, fusion results with the ventricular septum through the right extremity of the AV cushions (Warwick and Williams 1973).

In the complete form of the endocardial cushion defect, the interventricular communication lies between the summit of the abnormal ventricular septum and the common AV valve. However, the major abnormality is a deficiency of muscular tissue subjacent to atrioventricular valves, although the membranous septum can be deficient as well. The size of the interventricular communication vries form a large to a small space between atrioventricular valve tissue and the upper margin of the ventricular septum. The peculiar scooped-out appearance of the ventricular septum reflects a deficiency of the upper part of the muscular septum. When viewed from the right ventricle, the defect is located inferior and posterior to the crista supra ventricularis and is frequently covered by the septal leaflet of the tricuspid valve. When viewed from the left ventricle, it is situated in the outflow tract of the left ventricle (Perloff, 1978).

Development of the Septum Primum:

The roof of the common atrium presents a depression in relation to the truncus. This depression deepens and divides the common atrium into right and left atria, and is now called septum primum. Internally, it has a free sickle shaped margin, the superior limit of which extends on the roof as far as the superior AV cushion, while, the inferior limit extends on the floor as far as the inferior AV cushion. Proliferation of the AV cushions brings about clossure of the ostium primum (Goor and Lillehei, 1975).

Development of the AV valves:

In principle, each atrioventricular valve leaflet is derived from two components, each component contributes half a leaflet. Two neighbouring masses of endocardial cushion tissues become separated from the ventricular wall by a process of undermining except only their basilar and apical portions which remain attached to the heart wall, that is on the atrioventricular ring and condensations of ventricular