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Value of Transesophageal Echocardiography in Congenital Abnormalities of the Atria and Venous Atrial Connections

Thesis

Submitted in partial fulfilment for MD in Cardiology

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List of abbreviations

CHD	Congenital heart disease
ASD	Atrial septal defect
OSASD	Ostium secundum atrial septal defect
OPASD	Ostium primum atrial septal defect
SVASD	Sinus venosus atrial septal defect
PAPVC	Partial anomalous pulmonary venous connection
TAPVC	Total anomalies pulmonary venous connection
PFO	Patent foramen ovale
ASA	Atrial septal aneurysm
SVC	Superior vena cava
IVC	Inferior vena cava
RUPV	Right upper pulmonary vein
LUPV	Left upper pulmonary vein
RLPV	Right lower pulmonary vein
AVSD	Atrioventricular septal defect
PLSVC	Persistent left superior vena cava
TTE	Transthoracic echocardiography
TEE	Transesophageal echocardiography
2D	Two-dimensional
3D	Three-dimensional
MRI	Magnetic resonance imaging
PAP	Pulmonary artery pressure
MRA	Magnetic resonance angiography

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Introduction and aim of the work

Review of the literature

Embryology of the atria and venous atrial connections: -

The cardiovascular system is the first system to function in the embryo; blood begins to circulate by the end of the third week. This rapid development is essential because the growing embryo needs an efficient method of acquiring nutrition and disposing waste products **(Moore et al, 1979).**

Normal Development: -

The heart first takes form by the fusion of two endothelial tubes that are formed in the splanchnic mesoderm. The splanchnic mesoderm surrounding these tubes differentiates into the myocardium and epicardium. The endocardium of the fully formed tube corresponds to the primitive endothelial cardiac tube, from this endothelial tube develop also the cardiac valves and septae.

At 21 days of gestation fusion of the right and left cardiac tubes occurs to form a single cardiac tube and then 2 processes take place: -

1) The tube develops two constrictions that demarcate several cardiac segments from caudal to cranial these are:
   - The sinus venosus, which receives the umbilical, vitteline and common cardinal vein.
   - The atrium, the ventricle, the bulbus cordis, the aortic sac and aortic arch.
2) The cardiac tube elongates because it is fixed at both sinus venosus and aortic ends and it bends upon itself as it elongates forming S shape configuration.

The bulbus cordis and the ventricle are displaced anteriorly and to the right while the atria and sinus venosus are displaced posteriorly to lie behind the ventricles, subsequently the ventricles migrate to the left and the bulk of the cardiac mass lies in the left hemithorax **(Amplatz, 1986).**

Fate of the sinus venosus and formation of the adult right atrium: -

Initially the sinus venous opens in the primitive atrium and its right and left horns are of same size. By the end of the fourth week the right horn becomes larger than the left because of progressive increase of blood flow in the right precardinal venous system through two left to right shunts. The first left to right shunt results from transformation of vitteline and umbilical veins while the second one comes from communication between the anterior cardinal veins by oblique anastomosis.

In summary the definite right atrium is formed of two parts:

1) The smooth part of the wall of the right atrium, which is called the sinus venarum, is derived from the sinus venosus and its right horn.
2) The rough part of the right atrium is derived from the primitive atrium (this part has conical pouch called the auricle and trabeculated appearance) **(Williams et al., 1995).**

Formation of vena cavae: -

The vena cavae are derived from the cardinal system of veins, which is completely intraembryonic, and give rise to all systemic veins. The left and right precardinal veins drain the cephalic region of the embryo and they are connected with the corresponding postcardinal veins to form the right and left common cardinal veins, which enter the sinus venosus.

Drainage of both sides of the head and neck is directed to the right precardinal veins and common cardinal veins, which form the superior vena cava. The inferior vena cava is formed from the following developmental segments from caudal to cranial these are:

-Post-renal part from the right sacrocardinal vein.
-Renal and prerenal parts from the right subcardinal vein.
-Hepatic part from the right hepatic vein (**Rahilly et al., 1995**).

Chiari network is a remnant of the valve of the inferior vena cava and the valve of coronary sinus; functionally this network has no

significance. Pulmonary embolism has been attributed to formation of thrombi in the network **(Schneider et al., 1995).**

Formation of the adult left atrium and pulmonary veins: -

The left atrium is formed of two parts:
1) The smooth part derived from the common pulmonary vein, which is developed as invagination in the posterior wall of the primitive atria in the sino atrial region.
2) The rough part of the left atrium is developed from the primitive atrium.

The pulmonary veins developed as a common vein which is gradually absorbed and incorporated in the wall of the left atrium and the proximal portion of its tributaries is also absorbed resulting in four pulmonary veins with separate openings in the wall of the left atrium **(Los, 1986).**

Atrial septation: -

The septum primum appears in the roof of the atrium as a single fold to the left of the ostium primum. The subseptal opening between the atria decreases as the septum primum grows and it becomes obliterated when the septum fuses with the atrioventricular canal. Immediately before this another opening called foramen secundum appears in the septum primum through this opening blood can pass from the right side of the heart to the left side and this is essential prenatal.

The septum secundum develops as a sagital fold to the right of the septum primum. Its free border remains as a sharp crescentic edge or limbus which bounds an interatrial opening termed foramen ovale situated between the septum primum and septum secundum. The highly oxygenated blood from the inferior vena cava is projected against the edge of septum primum and the septum primum form a valve for foramen ovale. Ensuring right to left passage only **(Rahilly et al., 1995)**.

Increased pressure in the left atrium postnatally closes the valve at

first functionally and then anatomically during the first two days of postnatal life however, in some people a small slit opening remains in the atrial septum as a part of foramen ovale (probe patency) but it is without functional significance **(Patten, 1931).**

Atrio ventricular canal: -

The atrio ventricular canal is at first connected to the future left ventricular portion of the cardiac tube then it becomes transformed more to the right in an obscure manner to become attached to both ventricles. Two areas of cardiac mesenchyme called endocardial cushion project in the canal and soon fuse with each other dividing the canal into two channels, one connecting the left atrium to the left ventricle and the other connecting the right atrium to the right ventricle. The septal surfaces of the endocardial cushion differentiate further to form the septal leaflet of the mitral and tricuspid valves **(Becker and Anderson, 1982).**

Recent studies in the embryology of the heart: -

Concepts of development must be consistent with the anatomy seen in the postnatal life. In a trial to correlate the embryological development of the atrial septum and atrioventricular junction with the morphology of these structures in the postnatal life, **Anderson et al., 1999,** reviewed old and new accounts of cardiac morphology with regard to the definite structure of the atrial septum and they found that:

1) The atrial septum is formed of two major parts, folds of atrial wall and true interatrial portions. The flap valve of foramen ovale and its inferior rim are the true septal structures while other parts of the septum particularly anterosuperior rim are infoldings enclosing extra cardiac fat.
2) The systemic veins enter the right side of the primary atrium whereas the pulmonary veins enter the left side of the primary atrium and once this achieved the primary septum together with the mesenchymal cap grows between the two venous systems.