Congenital Extra-cardiac Vascular Anomalies as Detected on Multislice Computed Tomography Angiography

Thesis

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in **Radio-diagnosis**

By

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INTRODUCTION

Congenital heart diseases (CHD) are the most frequent types of birth defects. For the moderate and severe forms, the incidence of CHD is approximately 6/1,000 live births ***(Salvador et al., 2014).*** Comprehensive anatomic assessment in complex CHD is crucial for adequate patient management. Trans-thoracic echocardiography (TTE) along with cardiac catheterization serve as the cornerstone modalities in complex CHD primary evaluation. A high proportion of extra-cardiac vascular and non- vascular malformations are peculiar for CHD. Those further common abnormalities may influence the precise planning of corrective or palliative surgical or non-surgical therapy. In patients with complex CHD, TTE with color Doppler provides excellent delineation of the intracardiac anomalies comprising hemodynamic evaluation as well. However, TTE is deficient in accurately characterizing extra cardiac thoracic structures as the aorta and the aortic arch branches, the pulmonary arteries and their branches, the pulmonary veins, or associated other vascular structures and airways ***(Shalaby et al ., 2016).***

Cardiac catheterization provides most of the ancillary requisite information, but it is invasive in

nature and therefore entails inherent complications aside from the exposure to ionizing radiation and iodinated contrast administration. Moreover, cardiac catheterization is not informative regarding associated airway pathology ***(Goitein et al., 2014).***

The capability of electrocardiography-gated computed tomography-angiography (ECG-gated CTA) to accurately volumetrically image the morphologic features of complex CHD has been well portrayed in adults and young patients ***(Paul et al., 2005).*** Moreover, retrospectively ECG-gated helical CT permits both morphologic and functional evaluation

of the heart as hemodynamic information, comprising extra-cardiac and intra-cardiac shunts as well as valvular diseases; however this in return for higher radiation dose compared to prospective ECG-gated sequential scans ***(Goo, 2010).***

It also enables the systematic evaluation of other thoracic structures like cardiovascular structures, the airways and the lungs by using maximum and minimum intensity projections to delineate the vascular and airway structures, respectively. Extra-cardiac great vessels can be evaluated along their length; thus augmenting the role of cardiac CT scans in children with congenital heart diseases ***(Shehata et al., 2017).***

Despite the poorer temporal resolution of cardiac CTA in comparison to echocardiography, yet the constellation of swift acquisition time, expanded anatomic coverage, high spatial resolution, multiplanar reformation and 3D capability coupled with a flexible ECG synchronization have ameliorated the image quality of cardiac CT scans and reduced the potential risks ***(Shehata et al., 2017).***

Over and above, Multidetector computer tomography (MDCT) can be performed with no necessity for sedation, images are taken in very short time, which makes it particularly indicated in young children. It is also feasible in patients with pacemakers, mechanical prosthesis, metallic conduits and coils, while MRI is contraindicated in those cases or shows metallic artifact ***(Ohnesorge et al., 2007).*** In view of the forementioned features, MDCT is considered an ideal non- invasive method for assessing pediatric patients with congenital extra-cardiac anomalies ***(Hellinger et al., 2011).***

Aim of the work

The aim of this study is to evaluate the advantage of recent advances of ECG gated MDCT in diagnosis of congenital heart disease in pediatrics and assessment of associated extra cardiac abnormalities within the great vessels in comparison with echocardiography findings.

Embryology of the heart and extra-cardiac vessels

The heart is considered the first fully developed and functioning organ during vertebrate development ***(Martinsen and Lohr, 2005).*** The vascular system is apparent by the middle of the 3rd week, when the embryo can't maintain its nutritional needs solely by diffusion. Cardiac progenitor cells subsist in the epiblast, directly lateral to the primitive streak. From there they migrate through the streak. Cells intended to form cranial cardiac segments entailing the outflow tract, migrate first, whereas cells constructing more caudal portions composed of right ventricle, left ventricle, and finally sinus venosus, respectively, migrate in sequential order. The cells advance toward the cranium and position themselves rostral to the buccopharyngeal membrane in the splanchnic layer of the lateral plate mesoderm ***(Fig. 2.1) (Sadler, 2003).***

Late in the presomite stage of development, these cells are induced by the underneath pharyngeal endoderm formulating cardiac myoblasts ***(Sadler, 2003).*** Moreover; Blood islands appear in this mesoderm forming blood vessels and cells via vasculogenesis process**.** Thereafter, the islands unite ending in endothelial-lined horseshoe-shaped tube wrapped up by myoblasts forming what is known as cardiogenic field ***(Larsen, 2001).***

Other blood islands reside near the midline of the embryonic shield in parallel fashion forming a a pair of longitudinal vessels, dorsal aorta ***(Sadler, 2003).***

I. Develoment of the heart chambers

1. Formation and positioning of the developing heart tubes

Primarily, the central cardiogenic area as well as the future pericardial cavity are frontal to the buccopharyngeal membrane and neural plate ***(Fig. 2.2 A).*** Escorted by neural tube closure and ensuing formation of the brain vesicles, the central nervous system (C.N.S) grows up cephalad so promptly that it expands over the central cardiogenic area ***(Fig.2.2).*** Resultant from swift brain growth and embryonic vertical folding, the **buccopharyngeal membrane** is drawn forward, whilst the pericardial cavity steps to the cervical region and down to the thorax thereafter ***(Fig. 2.2) (Martinsen and Lohr, 2005).***

At the moment the embryo folds cephalocaudally, it also bends laterally ***(Fig. 2.3).*** Thence, caudal regions of the paired cardiac primordia merge not counting their caudalmost ends. Concourrently, the crescent part of the horseshoe-shaped area widens to compose the future ventricular regions and outflow tract. Therefore, the

heart now is an expanded tube formed of an inner endothelial layer and an external myocardial layer. It welcomes venous blood at its caudal pole and pumps blood into dorsal aorta out of the 1st aortic arch at its cranial end ***(Figs. 2.4)*** ***(Sylva et al., 2013).***

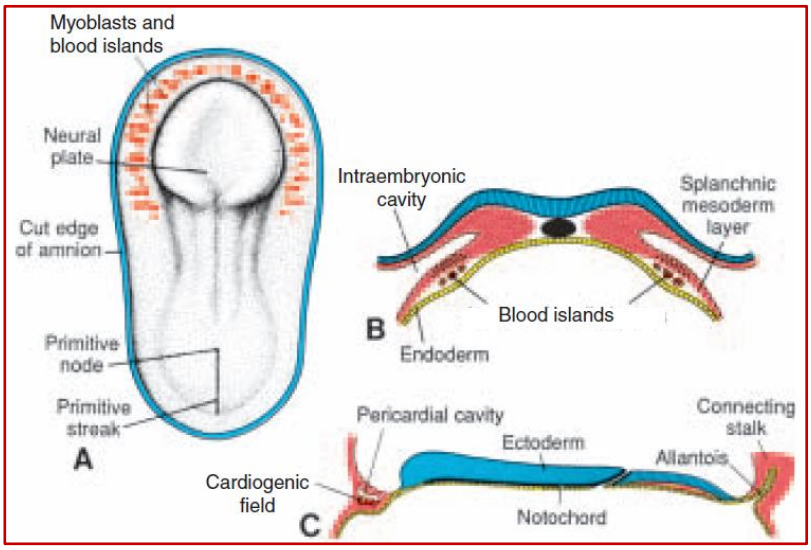

Figure 2.1 Dorsal view of a presomite embryo (approximately 18 days) after removal of the amnion. **(A).** Prospective myoblasts and hemangioblasts reside in the splanchnic mesoderm late of and in front of the neural plate on each side of the embryo. **(B)** Transverse section through a similar-staged embryo to show the position of the blood islands in the splanchnic mesoderm layer. **C.** Cephalocaudal section through a similarstaged embryo showing the position of the pericardial cavity and cardiogenic fields ***(Sadler, 2003).***

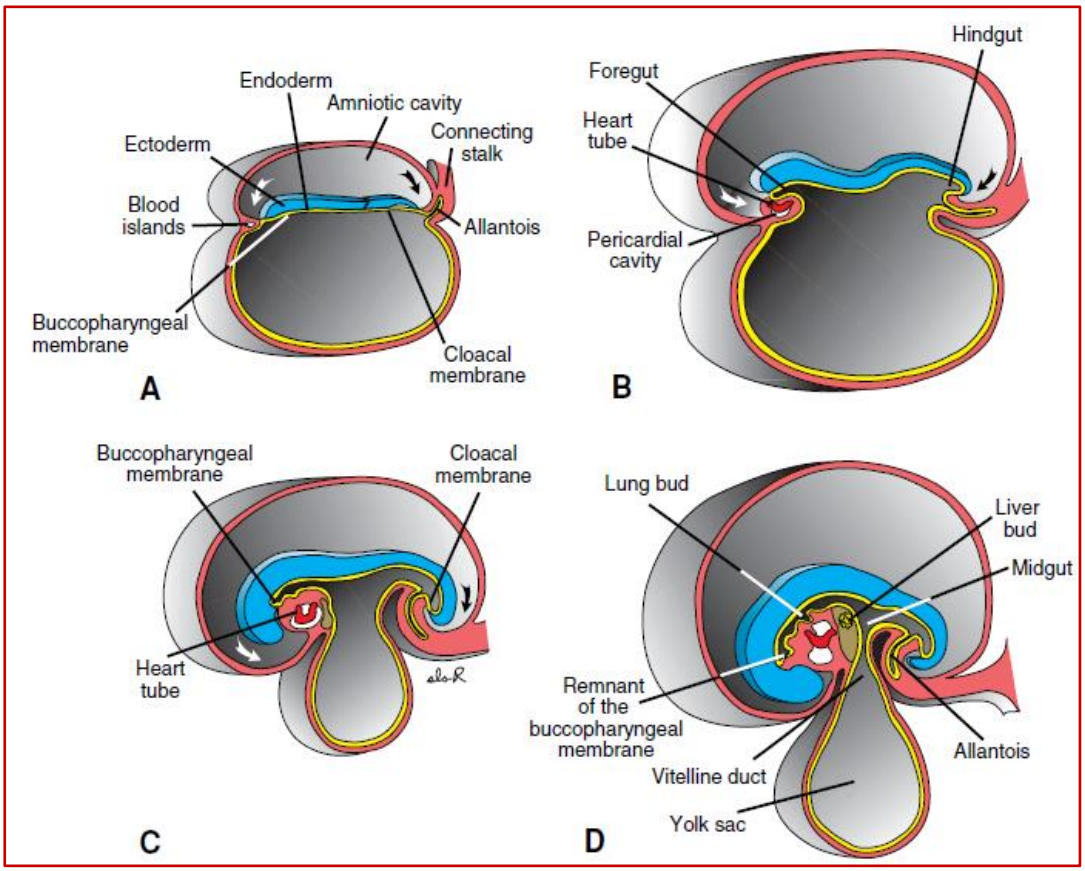

Figure 2.2 Figures showing effects of the rapid growth of the brain on positioning of the heart. Initially the cardiogenic area and the pericardial cavity are in front of the buccopharyngeal membrane. **(A)** 18 days. **(B)** 20 days. **(C)** 21 days. **(D)** 22 days ***(Sadler, 2003).***

The evolving heart tube bulges increasingly into pericardial cavity.

Primitively, the tube remains connected via fold of mesodermal tissue to the dorsal pericardial cavity, nominated the **dorsal mesocardium** ***(Figs. 2.3 and 2.4)*** which fades away later generating the **transverse pericardial sinus,** linking both sides of the pericardial cavity. The heart is now suspended in the cavity via blood vessels at both cranial and caudal ends. The ventral mesocardium is never existent ***(Fig. 2.4) (Sadler, 2003).***

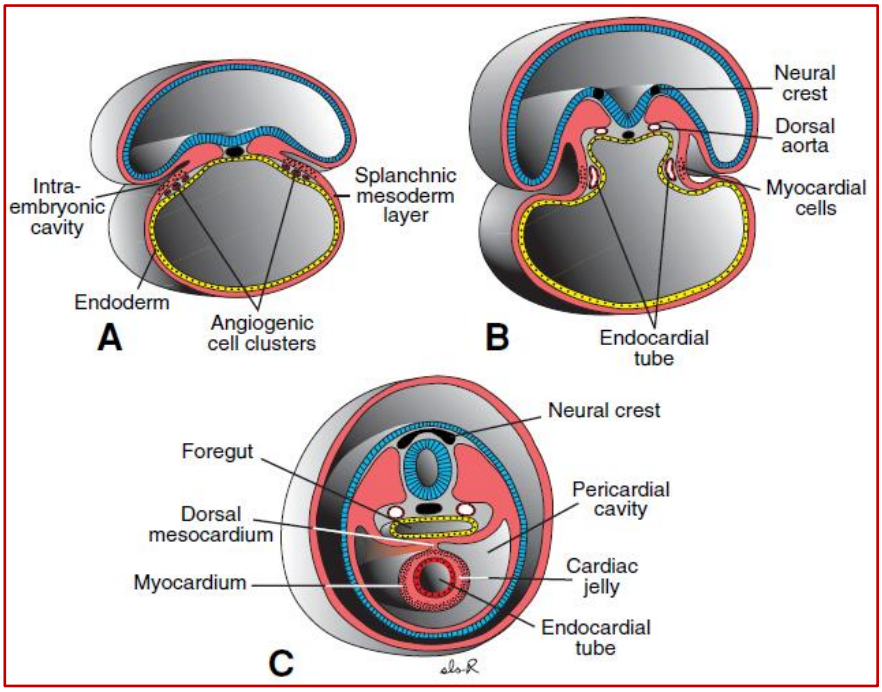

Figure 2.3 Transverse sections through embryos at different stages of development, showing formation of a single heart tube from paired primordia. **(A)** Early presomite embryo (17 days). **(B)** Late presomite embryo (18 days). **(C)** Eight-somite stage (22 days). Fusion occurs only in the caudal region of the horseshoe-shaped tube. The outflow tract and most of the ventricular region form by expansion and growth of the crescent portion of the horseshoe ***(Sadler, 2003).***

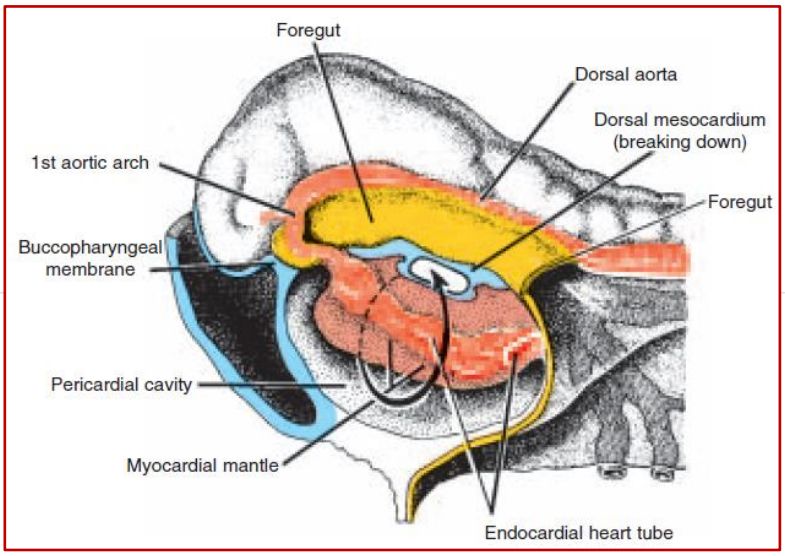

Figure 2.4 Cephalic end of an early somite embryo. The developing endocardial heart tube and its investing layer bulge into the pericardial cavity. The dorsal mesocardium is breaking down ***(Sadler, 2003).***

Throughout these events, the myocardium gets thicker while releasing a dense layer of extracellular matrix, differentiating it from the endothelial linning ***(Figs. 2.3 and 2.4).*** Moreover, mesothelial cells from sinus venosus province migrate over the heart forming the **epicardium.** So, the heart tube is constituted of three layers: (*a*) the **endocardium,** representing the innermost endothelial lining; (*b*) the **myocardium,** deemed as the middle muscular wall; and (*c*) the **epicardium** or **visceral pericardium,** wrapping-up the tube. This outermost layer is responsible for coronary arteries genesis ***(Sadler, 2003).***

2. Cardiac Looping

The heart tube continues to stretch and bend on day 23. The cephalic portion of the tube shifts ventrocaudally, and to the right, whilst the atrial (caudal) portion bends in the opposite direction generating the **cardiac loop** that is complete by day 28 ***(Fig 2.5).***

Synchronously, local expansions become apparent throughout the entire tube length. Paired **atrial portion**, composes a common atrium and get integrated into the pericardial cavity. The **atrioventricular (AV) junction** stays narrow while forming the **atrioventricular canal** ***(Fig. 2.6).***The **bulbus cordis** is narrow excluding its proximal third constituting the **trabeculated portion of the right ventricle (RV)** ***(Figs. 2.6).*** The midportion, the **conus cordis,** will be configured as the outflow tracts. The distal part of the bulbus, the **truncus arteriosus (TA),** will set up the roots and proximal parts of the great arteries; specifically the aorta and pulmonary artery ***(Fig. 2.6) (Larsen, 2001).***

The junction between the ventricle and the bulbus cordis marked exteriorly by the **bulboventricular sulcus** stays narrow to be designated as the **primary interventricular foramen** ***(Fig. 2.6) (Sadler, 2003).***

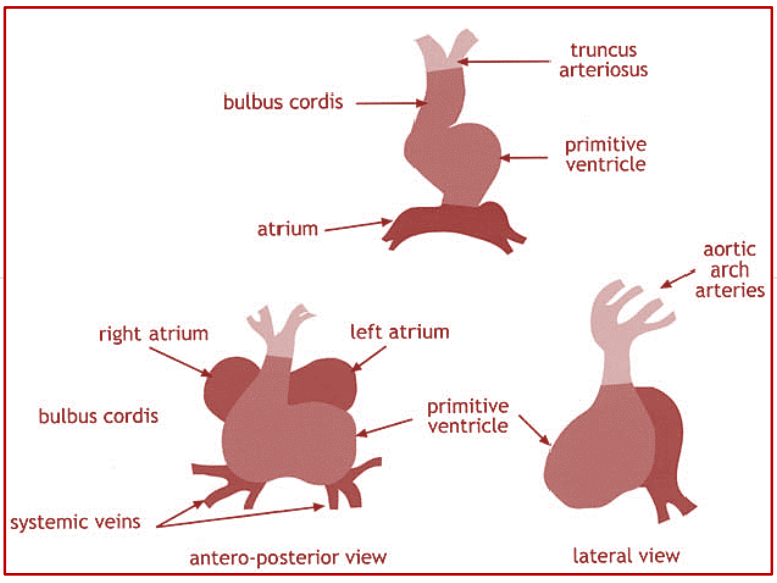

Fig. 2.5. Looping of the single endocardial heart tube transforms it into a complex fourchamber structure. Looping starts on day 23 of development, and the four-chambered heart is evident on day 28 ***(Sadler, 2003).***

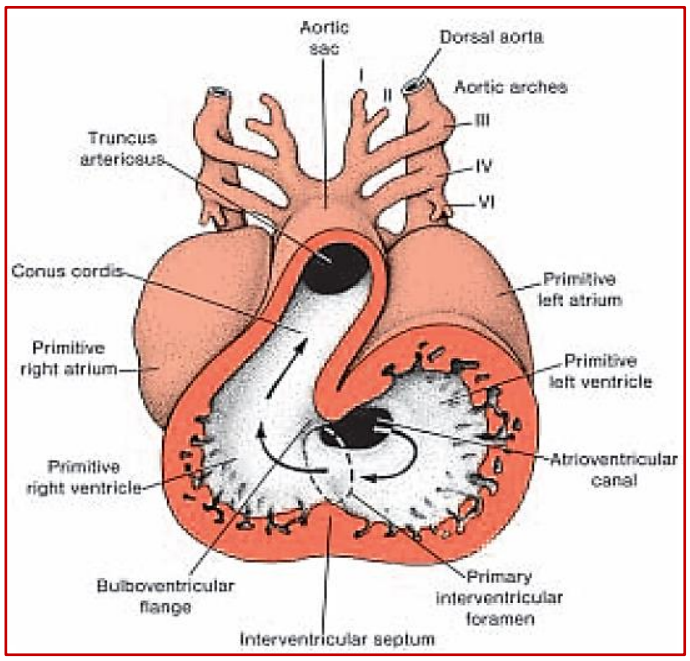

Figure 2.6 Frontal section through the heart of a 30-day embryo showing the primary interventricular foramen and entrance of the atrium into the primitive left ventricle. Note the bulboventricular flange. *Arrows,* direction of blood flow ***(Sadler, 2003).***

By the end of loop formation, the primitive trabeculae starts to appear in two well-delineated areas just proximal and distal to the **primary interventricular foramen *(Fig. 2.6).*** Tentatively; the bulbus stays smooth walled. The primitive trabeculated ventricle is labelled the **primitive left ventricle (LV).** Likewise, the trabeculated proximal third of the bulbus cordis may be denominated as the **primitive right ventricle (RV).** The conotruncal part principally situated on the right side of the pericardial cavity, shifts gradually to a more medial position ***(Sadler, 2003).***

3. Chambre septation

A. Partitioning the Atrio-Ventricular Canal

By the end of the 4th week, two mesenchymal **atrioventricular endocardial cushions,** pop-up at the upper and lower borders of the AV canal ***(Figs.2.7 and 2.8).*** Firstly, the AV canal points only to the primitive left ventricle, being seprabale from the bulbus cordis by **bulbo(cono) ventricular flange**. Thereafter, the AV canal grows to the right so that blood passing through AV orifice is directed to both primitive ventricles ***(Abdulla et al., 2004).***

Aside from upper and lower endocardial cushions, the two **lateral atrioventricular cushions** appear on the right and left borders of the canal ***(Figs. 2.7 and 2.8).*** Synchronously by the end of the 5th week, the superior and inferior cushions protrude further into the lumen and get fused, giving rise to distinct right and left AV

orifices ***(Abdulla et al., 2004).***

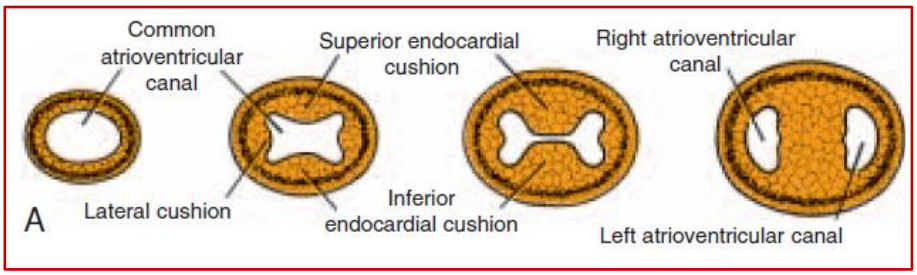

Figure 2.7. Formation of the septum in the atrioventricular canal. **A.** From left to right, days 23, 26, 31, and 35. The initial circular opening widens transversely ***(Sadler, 2003).***

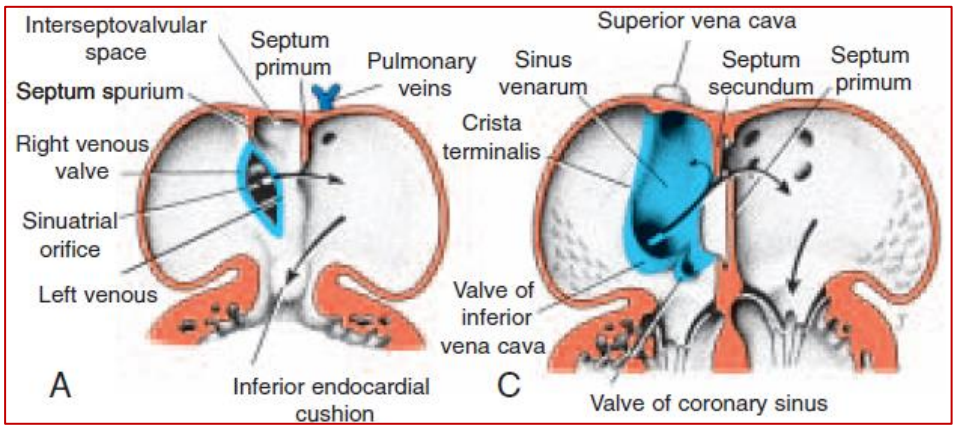

Figure 2.8. Frontal section through the heart of a day 35 embryo. At this stage of development blood from the atrial cavity enters the primitive left ventricle as well as the primitive right ventricle. Note development of the cushions in the atrioventricular canal. Cushions in the truncus and conus are also visible. *Ring,* primitive interventricular foramen. *Arrows,* blood flow ***(Sadler, 2003).***

B. Partitioning the Atria

By end of the 4th week, a sickle shaped crest called the septum primum, protrudes from the dorsal mesocardial mesenchyme intraluminally into the atrium. With its growth, the ostium primum - a foramen that permits right to left atrial blood shunting – gets shrinken. Nonetheless, programmed cell death close to the superior rim of the septum primum fashions, the denovo ostium secundum, that perserves shunting of the oxygenated blood. Afterwards, an incomplete, ridged septum secundum harboring foramen ovale adjacent to right atrial floor is formed next to septum primum; both merge with the septum intermedium of the AV cushions ***(Fig. 2.9, 2.10) (Martinsen and Lohr, 2005).***

Within the fully developed heart, the **trabeculated atrial appendage** represents the embryonic left atrium, whilst the smooth-walled part originates from the pulmonary veins ***(Fig. 2.11).*** Similarly on the right side, the embryonic right atrium gives origin to the trabeculated **right atrial appendage** while the smooth-walled **sinus venarum** stems from the right horn of the sinus venosus ***(Sadler, 2003).***