

Cor trilobulare biventriculare with left superior vena cava

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Cor trilobulare biventriculare is a rare congenital malformation of the heart in which there is a complete absence of the atrial septum. It is usually associated with other anomalies like complete atrioventricular canal defect, polysplenic syndrome, isolated dextrocardia, Ellis-van Creveld syndrome, or persistent left superior vena cava. We report a case of a stillborn male foetus of 35 weeks gestation with common atrium, complete atrioventricular canal defect, and persistent left superior vena cava. The possible embryological basis and clinical implication of this variation are discussed. (Folia Morphol 2011; 70, 2: 135–138)

Key words: Cor trilobulare biventriculare, persistent left superior vena cava, common atrium

INTRODUCTION

Cor trilobulare biventriculare is a rare congenital malformation of the heart in which there is a complete absence of the atrial septum. Atrial septal defects (ASD) occur in 4 of every one lakh people, with a gender predominance in females (F:M ratio of 2:1). The incidence of ASD is 6–10% of all cardiac anomalies.

The complete absence of the atrial septum is rare and is considered to be the least common variety of ASD. It is usually associated with other anomalies like complete atrioventricular (AV) canal defect, polysplenic syndrome, isolated dextrocardia, Ellis-van Creveld syndrome, or persistent left superior vena cava (SVC).

We describe the autopsy findings in a male foetus with common atrium associated with AV canal defect and persistent left SVC. Other associated malformations are seen.

CASE REPORT

A male foetus still born at 35 weeks gestation was taken from the Gynaecology Department of NRI Medical College, Chinakakani. It presented various

abnormal external features — broad chest, short neck, low-set ears, polydactyly, syndactyly, and bilateral cleft lip and palate (Fig. 1). On autopsy non-rotation of the gut was observed. The appendix was in the umbilical region.

A detailed study of the cardiovascular system revealed the following features (Fig. 2): The dimensions of the heart were 2.7 cm transversely and 2.4 cm vertically; right ventricular hypertrophy was observed; the SVC was present on the left side; inferior vena cava was normal in position; there was pulmonary trunk stenosis at the commencement; right pulmonary artery was enlarged, and the left pulmonary artery was narrow; the ascending aorta and the proximal part of the arch of the aorta were dilated; three branches arose from the arch of the aorta-bicarotid trunk, left subclavian artery, and retroesophageal right subclavian artery (Fig. 3).

A study of the interior of the heart revealed the following features (Fig. 4): common atrium was present. Inter-atrial septum was completely absent; the opening of the inferior vena cava was normal in position; persistent SVC opened into the roof of the



Figure 1. External features of the foetus.

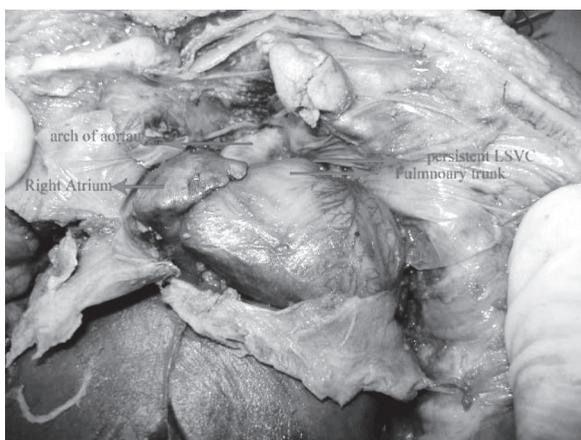


Figure 2. External features of the heart; LSVC — left superior vena cava.

common atrium on the left side; the coronary sinus was absent; the ascending aorta was dilated, dextroposed, and overrode the interventricular septum, which was defective (Figs. 5, 6); the aortic orifice

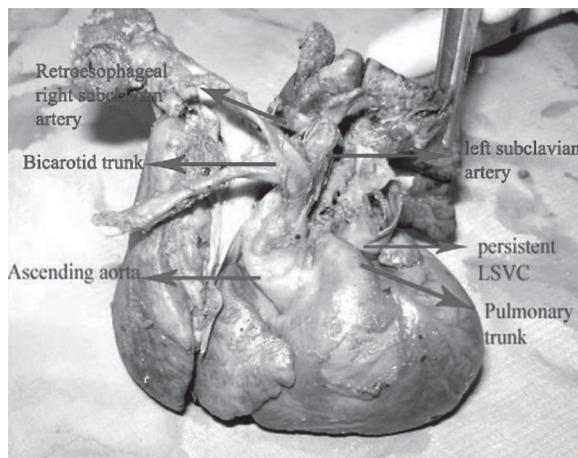


Figure 3. Branching pattern of aortic arch and left superior vena cava (LSVC).

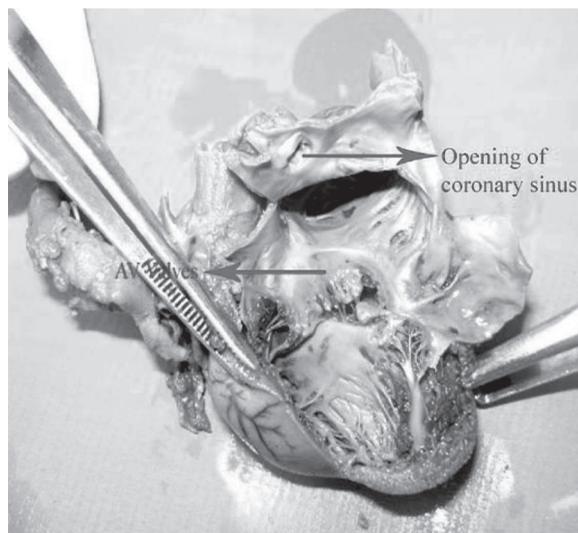


Figure 4. Internal features of the heart; AV — atrioventricular.

had three cusps — right and left anterior and one posterior; the right coronary ostium was located at the upper margin of the right anterior cusp; the left coronary ostium and ostium of the right conus artery were located in the left anterior aortic sinus; and the pulmonary orifice had three cusps which were rudimentary, appearing like a ring.

The common AV orifice had one anterior and one posterior cusp on the right side, and one anterior and one posterior cusp on the left side. The anterior cusp on the right side was continuous with the anterior cusp of left side over the defective interventricular septum (Fig. 4).

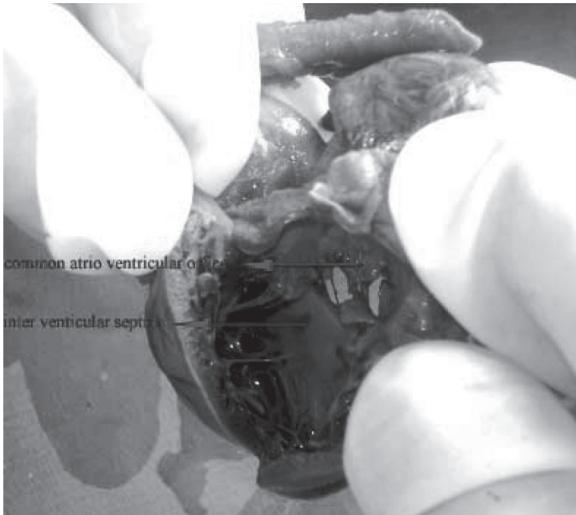


Figure 5. Defective interventricular septum.

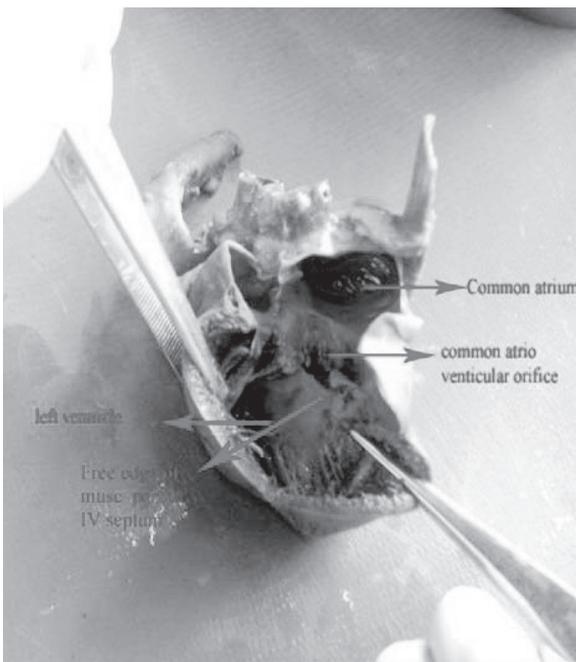


Figure 6. Interior of the heart; IV — interventricular.

DISCUSSION

Common atrium, a rare congenital malformation of the heart occasionally associated with abnormalities of venae cava and coronary sinus, is thought to be one form of endocardial cushion defect, and it was first described by Young and Robinson in 1907 [19]. Abbott [1] in her analysis of 1000 congenital heart diseases described this anomaly in 5 cases. Cunningham [4] reported a case with complete absence of the atrial sep-

tum together with communication between the mitral and tricuspid valves, an absence of the membranous part of the ventricular septum, and aneurismal dilatation of the pulmonary arteries. Campbell and Nissen [2] classified the anomaly into three groups: persistent ostium primum, a partial AV canal, and a complete AV canal. Ellis et al. [6] reported 5 cases of common atrium, and he considered that a complete absence of atrial septum is always associated with AV valve anomalies. Dubost and Blondeau [5] reported 2 cases, and Shaher and Johnson [16] reported 3 cases with common atrium. Giknis [7] wrote that 7 of 11 cases with Ellis-van Creveld syndrome have a single atrium. Rastelli et al. [15] noted the following features: complete absence of atrial septum or its representation by a small strand of tissue present in the cephalad wall of the common chamber; and the absence of interventricular communication and an accompanying cleft in the anterior leaflet of the mitral valve. Hung et al. [9] reported 14 cases of common atrium with persistent AV canal. Lu et al. [3] reported common atrium with common AV valve and ventricular defect in association with Holt-Oram syndrome.

Lewis et al. [12] wrote that a complete absence of the atrial septum may exist alone and unassociated with malformations of AV valves. Similar cases were reported by Watkins and Gross [18], Probyn-Williams [14], Cunningham [4], Dubost and Blondeau [5], and Munoz-Armas et al. [13].

Levy and his associates [11] recommended the term single atrium for a condition with complete absence of atrial septum, absence of malformations of AV valves, and absence of interventricular communication. The term common atrium is used for a condition with complete absence of atrial septum accompanied by malformations of AV valves, with or without interventricular communication.

Normal interatrial septum formation occurs between 20 to 34 days of intra uterine life. Growth arrest of septum primum and secundum results in an absence of atrial septum. On the 28th day the primitive atrium is dorsal to the primitive ventricle. The two chambers communicate through a narrow passage, the AV canal. On the 35th day, two thickenings (endocardial cushions) develop on the dorsal and ventral sides of the AV canal, which grow and fuse to form the septum intermedium. This septum divides the AV canal into the right and left AV orifices. The mesenchyme around each orifice proliferate to form valve leaflets. Small portion of AV valves also develop from endocardial cushions. The membranous part of the interventricular septum is developed from en-

docardial cushions. Endocardial cushion defects may lead to common AV orifice, AV valve malformations, and failure of development of the membranous part of the interventricular septum. Patients with common atrium show a decrease in exercise tolerance early in life, increased fatigability, shortness of breath, cyanosis, and heart failure, as described by Hirai et al. [8].

In the present case, common atrium is associated with persistent left SVC and completely unroofed coronary sinus. The presence of a persistent left SVC is the most common congenital anomaly of the systemic veins with a prevalence ranging from 0.3% to 2% in the general population and a higher prevalence of 4.4% among those with congenital heart disease. In 80% to 90% of cases, persistent left SVC drains into the coronary sinus and right atrium, and it is usually asymptomatic. Rarely, the persistent left SVC will drain into the left atrium through the unroofed coronary sinus, a direct connection to the left atrium. Unroofed coronary sinus defects comprise < 1% of all ASD. Left SVC is due to persistence of the caudal part of the left anterior cardinal vein, left common cardinal vein, and left horn of sinus venosus. When the wall between the coronary sinus tube and left atrium fails to form (which is caused by maldevelopment of the arteriosinus fold) the systemic venous blood drains directly into the left atrium. This defect is called unroofed coronary sinus. Unroofed coronary sinus was classified into four types by Kirklin and Barratt-Boyes [10]: type I — completely unroofed with left SVC; type II — completely unroofed without left SVC; type III — partially unroofed midportion; and type IV — partially unroofed terminal portion.

This anomaly can predispose to complications like systemic emboli, brain abscess, premature death, right heart failure, left ventricular dysfunction, tricuspid or mitral regurgitation, atrial flutter or fibrillation, and pulmonary and systemic hypertension as described by Raj and others [17].

CONCLUSIONS

Completely unroofed coronary sinus with a left SVC draining into common atrium is a rare congenital anomaly which accounts for less than 1% of all ASD and can predispose to a number of complications. Awareness of such anomalies can help in early diagnosis and surgical correction for improved outcome.

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