



COR BILOCULARE ASSOCIATED WITH COMPLEX MALFORMATIONS

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ABSTRACT

Cor biloculare is one of the rare congenital malformations of the heart in which the heart has only two chambers, an atrium and a ventricle. The common atrium communicates with the common ventricle through the common atrioventricular valve. This cyanotic congenital heart disease is associated with other malformations like dextrocardia, polysplenic or asplenic syndrome, anomalous pulmonary venous drainage and persistent left superior vena cava. The death of patients with cor biloculare is common in neonatal period and a very few can survive to adult life. We present autopsy findings in a three day old female newborn with cor biloculare associated with persistent left superior vena cava, anomalous pulmonary venous connection, and atretic aorta with patent ductus arteriosus continuation of aorta. The possible embryological basis of this complex malformation is discussed. Other associated malformations were micrognathia, cleft palate and left lower limb deformity.

INTRODUCTION

Bilocular heart is probably the rarest congenital heart malformation [1]. It is characterized by a single atrium communicating with a single ventricle through a common atrioventricular (AV) valve. Cor biloculare is one of the most primitive cardiac defects. It is usually associated with other anomalies like dextrocardia, situs inversus, abnormal pulmonary veins (PVs), truncus arteriosus and persistent left superior vena cava (PLSVC) [2,3].

We present the autopsy findings in a female newborn with cor biloculare associated with abnormal pulmonary venous connection, PLSVC and aortic atresia with patent ductus arteriosus continuation of aorta. Other associated malformations were micrognathia, cleft palate

and left leg deformity. The case reveals association of rare cardiac anomalies with complex somatic malformations hitherto undescribed.

CASE STUDY

A female infant born full term, died on third day of respiratory distress was taken from the Pediatric Department of this Medical College. On external examination, it presented various anomalies as micrognathia, long philtrum, hypertelorism, left eye corneal opacity and low-set ears (Fig. I). Left leg and foot were deformed and rotated posteriorly at knee joint (Fig. I). Intraoral examination revealed complete U shaped cleft palate (Fig. II).



Head to toe dissection was meticulously performed. On dissection, all the viscera including liver and spleen were normally situated. Dissection of thorax revealed multiple rare cardiac anomalies. A detailed study of the cardiovascular system revealed the following features (Fig. III,IV,V): the heart was normally placed with deep AV groove, double SVC with left SVC draining into atrium via dilated coronary sinus; IVC was normal in position; pulmonary trunk was dilated with patent ductus arteriosus extending directly into the descending aorta; hypoplastic ascending aorta and arch of aorta; three usual branches arose from the arch of the aorta; the PV from each lung joined into one vein, the veins of both sides further joined into a single common PV which entered the right side of atrium. The atrium has a normal right auricle and a narrow connection with the left auricle.

The careful examination of the interior of the heart revealed (Fig. VI,VII) cor biloculare- a common (single) atrium with complete absence of interatrial septum and a common (single) ventricle with complete absence of interventricular septum. The interior of common atrium (Fig. VI) revealed connection with two appendages – with the right auricle situated in right AV groove and with left auricle situated to the left of origin of great vessels by means of a narrow tube passing behind the great vessels. The openings of right SVC, IVC were in normal position.

The PLSVC drained through dilated coronary sinus. A single common PV opened in the common atrium near opening of right SVC.

The common atrium communicated with the common ventricle through a large oval shaped common (single) AV valve. The valve was a bicuspid with one right (anterior) and other left (posterior) cusps. A failed attempt to form a right AV valve is indicated by presence in the atrium of a tiny oval nodule attached by fine chordate to atrial wall.

The ventricular wall was hypertrophied. The interventricular septum was completely absent and there was no rudimentary chamber (Fig.VII). The chordae tendinae of anterior cusp of common AV valve were attached to a papillary muscle on anterior wall and chordae of posterior cusp were attached to two small papillary muscles on posterior (inferior) wall of ventricle. The origin of pulmonary trunk and pulmonary valve were normal. The origin of aorta was atretic. Endocardial fibrosis near the origin of aorta could be seen. The pulmonary trunk and patent ductus arteriosus were dilated. Ascending aorta and arch of aorta were hypoplastic (narrow tubular). The dilated patent ductus arteriosus continued as descending aorta. The proximal portion of aorta and its three usual branches might be receiving blood through regurgitation from ductus.

Figure 1. Whole body photograph of the case. Note 1- Micrognathia, 2-Long philtrum, 3-Low set ears, 4-Deformed left leg and foot.



Figure 2. Photograph after resection of mandible. Note 1- Nasal septum, 2-Complete (U-shaped) Cleft Palate (open mouth), 3-Tongue.



Figure 3. Photograph of Heart of newborn- Anterior view- Common/Single Atrium (A), Common/Single Ventricle (V), Hypoplastic aorta with its branches (Ao), Pulmonary Trunk (Pt), Left Auricle (LA). Note- Deep atrioventricular groove.

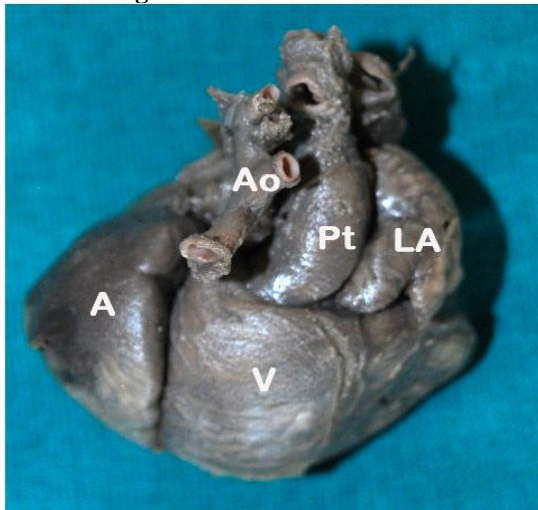


Figure 4. Photograph of Heart of the case- Right posterolateral view- Common/Single Atrium (A), Common/Single Ventricle (V), Right Superior Vena Cava (Rt. SVC), Inferior Vena Cava (IVC). Note: Persistent Left Superior Vena Cava (PLSVC) draining in coronary sinus (CS), Pulmonary Vein (PV) draining in common Atrium; Left Auricle (LA).

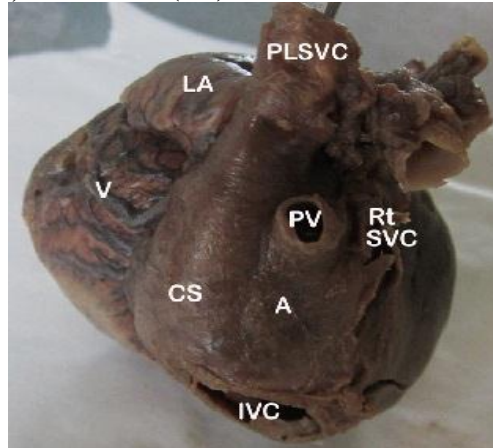


Figure 5. Photograph of superior view of heart. A- common atrium, V-Common ventricle, Ao- Hypoplastic aorta with its branches, Pt- Pulmonary Trunk, LA- Left Auricle, PLSVC-Persistent Left Superior Vena Cava, PV- Pulmonary vein.

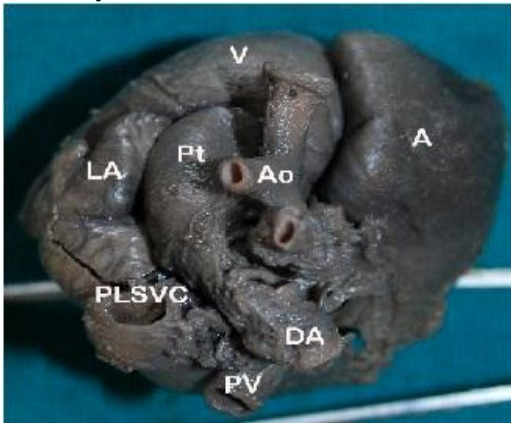


Figure 6. Photograph of interior of common atrium opened by vertical incision through SVC and IVC. RA- Right Auricle, CS- opening of Coronary Sinus, Conn to LA- Connection to Left Auricle, Single Atrioventricular (AV) valve. Note- Absence of interatrial septum

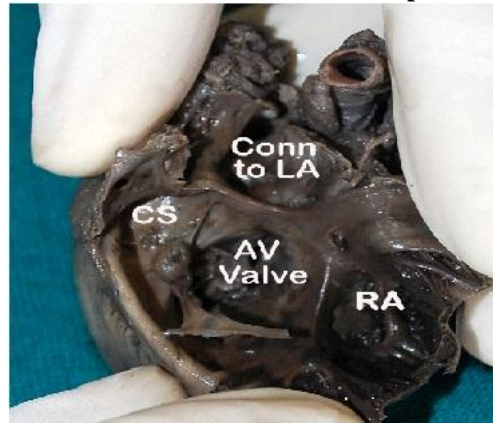
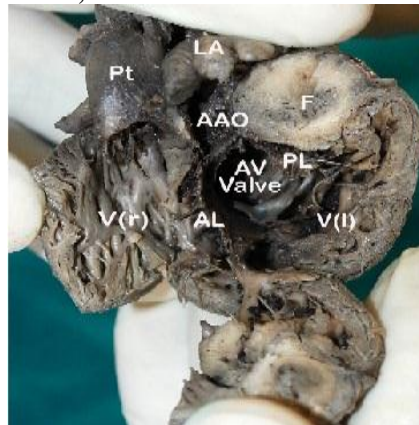


Figure 7. Photograph of interior of common ventricle (anterior wall detached). V(r) & V(l)- right & left portions of common ventricle, AAo- Atretic aortic orifice, Pt- Pulmonary Trunk, LA- Left Auricle. Atrioventricular (AV) valve with its anterior (AL) & posterior (PL) leaflets, F-endocardial fibrosis. Note-Absence of interventricular septum.



DISCUSSION

Cor biloculare (the two chambered heart) is probably the rarest of all congenital malformations of the heart [1,2]. Abott [4] observed only 14 cases of cor biloculare among 1000 cases of congenital heart disease. Cor biloculare represents one of the most primitive types of cardiac defect. Cor biloculare consists of a single atrium and a single ventricle (due to non development of interatrial and interventricular septa) communicating through the common AV valve. Brown [5] divided this rare anomaly into 3 groups- 1) with an undivided truncus arteriosus, 2) with normal division into aorta and pulmonary trunk and 3) incomplete forms with some septal formation but a persistent single AV valve. The present case appears to be of second type. Cor biloculare is a cyanotic congenital heart disease. Blood enters the common atrium from both systemic and PVs where it has variable mixing. Further mixing may occur as it enters the single ventricle through common AV orifice. Severity of cyanosis depends on diameters and pressures in aorta and pulmonary artery [6]. Minimal mixing of arterial and venous blood in the heart with little or no cyanosis has also been reported [2]. The absence of murmurs and thrills in these cases make the ante mortem diagnosis difficult [7].

Most babies with cor biloculare die shortly after birth. Abott [4] states the mean age of survival as 3 and 1/4 years, lonest survival in her series was 16 years. Longer survival in cor biloculare patients has been reported by Nelson and Wells, 1948 (27 years); Ogata H et al, 1989 (42 years) and Wang et al, 2014 (44 years) [6,8,9].

The atrium had two appendages in the present case of cor biloculare. Similar findings have been reported by Schechter and Meranze (1944) [10]. A single AV orifice is almost always present in cor biloculare. However, Taussing (1947) reported an unusual case with two AV orifices [11]. In the present case, in addition to a common AV valve, a failed attempt to form right AV valve can be seen in the form of a tiny oval nodule connected by fine chordae in the atrium. The common AV valve usually has three or four cusps but it is difficult to ensure their exact number [2]. Various authors have reported variable number of AV valve cusps in cor biloculare- four [12-14], three [10] or two cusps [8] (similar to our case).

Cor biloculare is almost always associated with other anomalies [2]. The common associated malformations are dextrocardia, situs inversus, polysplenia or asplenia, truncus arteriosus, abnormal pulmonary venous connection and PLSVC [2,3]. Heart and other viscera were normal in position and truncus arteriosus was normally divided in the present case.

Association of cor biloculare with anomalous pulmonary venous connection has been described [3,15]. A few authors have presented the associated anomalous pulmonary venous connection in the form of two PVs opening in common atrium [5,14]. However, in a present case, a single (common) PV (formed by union of right and left PVs) opened in the common atrium. A similar

pulmonary drainage by a common vein in the atrium of cor biloculare has been described by Kugel 1932 [12].

The systemic venous anomalies associated with cor biloculare have been reported in the form of absence of IVC [3,11], common opening of SVC and IVC in the atrium [14], presence of double SVC with the PLSVC draining in the atrium via coronary sinus [3,16]. The present case was also associated with double SVC with PLSVC draining via coronary sinus.

Many authors have described pulmonary valve abnormality and pulmonary artery stenosis or atresia in case of cor biloculare [3,6,11-14, 17]. However, present case had dilated pulmonary trunk with atresia of aortic orifice. The ascending aorta and arch of aorta were hypoplastic. The dilated patent ductus arteriosus was continuing with distal aorta. Similar cases have been reported by Schechter and Meranze (1944) and McCracken et al (1964) [10,16].

The term univentricular heart is now used to describe hearts with only one ventricle connected with atria (double inlet ventricle or a single AV connection) and those with one hypoplastic ventricle (includes pulmonary/aortic atresia or severe stenosis with hypoplastic ventricle). The term also includes the hearts not amenable to biventricular repair [18,19]. In view of such debates over terminology, a few authors have described the cases of incomplete/partial cor biloculare as univentricular heart associated with common atrium due to the presence of at least a partial interventricular septum or of another small rudimentary/accessory/non-functional ventricular chamber [20,21]. However, the present case is a true cor biloculare without any traces of interventricular septum or second rudimentary ventricular chamber.

The presence of micrognathia, complete cleft palate, glossoptosis (=Pierre Robin Sequence); long philtrum, low set ears, left eye corneal opacity and left leg and foot deformed and twisted posteriorly (no talipes equinovarus) in this case was suggestive of some complex syndrome like Stickler Syndrome, Velocardiofacial Syndrome, Treacher Collins Syndrome and TARP syndrome but all the findings could not fit into any of these condition. Also no genetic tests were performed to confirm or rule out any genetic syndrome.

Embryological Basis of Cor Biloculare and Associated Anomalies:

Normal interatrial septal formation occurs between 20-34 days of embryonic period. Developmental failure of septum primum and secundum leads to absence of the interatrial septum and thus common (single) atrium. On 35th day, two endocardial cushions develop one on each ventral and dorsal side of AV canal (the communication between primitive atrium and ventricle) which later fuse to divide the canal into right and left AV orifices. The membranous part of interventricular septum develops from endocardial cushions and muscular part from wall of



primitive ventricle. Endocardial cushion effect may result in common AV orifice, AV valve malformations and failure of development of interventricular septum [22]. Thus developmental failure of septum primum, secundum and endocardial cushions may lead to cor biloculare. Patients with cor biloculare shows cyanosis, dyspnea and decreased exercise tolerance [6].

Abnormalities of PVs are often found with common atrium (defective/absent atrial septum) [3]. Initially a single common PV develops around 24th embryonic day as an outgrowth from dorsal wall of still undivided atrium to the left of septum primum. A slight displacement of common PV with regard to the atrial septum or vice versa can result in connection of this PV with right atrium. When common PV grows, it divides into two parts and enters developing lung. First two divisions give rise to four PVs. As the atrium expands the primordial PV and its main branches are gradually absorbed into wall of atrium. As a result, four PVs are formed. The number of PVs opening into atrium can vary depending upon extent of incorporation of branches of common PV [23,24,25].

Abnormalities of systemic veins, especially PLSVC are also associated with common atrium [3]. The PLSVC is the most common congenital anomaly of the systemic veins (0.3% to 2% in the general population and 4.4% among congenital heart disease patients). In 80% to 90% cases, PLSVC drains into right atrium via coronary sinus. PLSVC occurs due to non-regression of left anterior cardinal and common cardinal vein. Normally the part of left anterior cardinal vein caudal to innominate vein regresses and forms the ligament of marshal. It may persist as left SVC draining into coronary sinus which gets dilated

(coronary sinus develops from left horn of sinus venosus which receives left common cardinal vein) [22]. The association of cor biloculare with PLSVC is due to maintenance of original more symmetrical arrangement on sinus venosus [3].

Large pulmonary artery and hypoplastic aorta with atresia of aortic orifice in the present case may be the result of defective trunco-conal septum which normally divides the truncus arteriosus into aorta and pulmonary trunk. Another possibility is the effect of endocardial fibrosis of ventricular wall near origin of aorta. To keep the blood supply to body, the ductus arteriosus remains patent communicating blood to distal aorta directly and to proximal aorta by regurgitation.

CONCLUSION

The present case of cor biloculare reports its association with complex cardiac, vascular and somatic malformations. Cor biloculare, a rare congenital heart anomaly has to be differentiated from Fallot's tetralogy, tricuspid atresia and complete AV canal defect [26]. The case emphasizes the importance of obstetric ultrasonography and fetal echocardiography screening for the early diagnosis of cor biloculare and other congenital heart diseases.

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None to declare

CONFLICT OF INTEREST

None to declare

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