

Pediatric Cardiology - Case Report

A Case Report of Common Atrium with Left Atrial Isomerism

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Background --- Common atrium (CA), is a rare congenital anomaly occasionally associated with abnormalities of the venae cavae and coronary sinus. This case report is being done to present a rare congenital heart disease which is common atrium with left isomerism and to be able to discuss the anatomical and embryological abnormality associated with atrioventricular septal defect (AVSD).

Case --- We present a case of a 5-year old female, who presented with murmur at birth, bradycardia and easy fatigability during early childhood. Work-ups revealed left atrial isomerism, interrupted inferior vena cava (IVC) and azygos draining to superior vena cava, common atrium, valvar pulmonary stenosis (PS) and small ventricular septal defect. She underwent mitral valve repair, creation of atrial partition, pulmonary valve commissurotomy, supravulvar pulmonary artery patching and permanent pacemaker insertion.

Conclusion --- Common atrium with left isomerism is a rare congenital defect associated with other malformations, frequently being less severe and more amenable to corrective surgery. *Phil Heart Center J 2012;16:63-67.*

Key Words: Left atrial isomerism ■ Common Atrium

Common atrium (CA), is a rare congenital anomaly occasionally associated with abnormalities of the venae cavae and coronary sinus. Levy and associates,¹ reported a case of complete absence of the atrial septum, indicating that this condition may exist alone as a specific entity, without an endocardial cushion defect. They recommended that the term *single atrium* should be used to denote the condition characterized by (1) complete absence of the atrial septum, (2) absence of malformation of the AV valves, and (3) absence of interventricular communication. They suggested that the term common atrium should be used to denote the condition of complete absence of the atrial septum, accompanied by malformation of the AV valves, with or without interventricular communication. In the presence of two ventricles, it is always associated with an AVSD. The pathologic spectrum of this lesion ranges from patients with coexistent primum and secundum ASDs to others who lack the entire septum except for a small muscular cord.

This case report is being done to present a rare congenital heart disease which is common atrium with left isomerism and to be able to discuss the anatomical and embryological abnormalities associated with AVSD.

CASE

RS, is a 5 year old female who was born to a 33 year old G7P6(6016) with noted decreased fetal heart rate by ultrasound at 8 months of gestation. She was delivered full term via caesarean section secondary to fetal distress (fetal bradycardia). There was an incidental finding of murmur at 2 weeks old. Initial chest xray showed normal vascular pattern with right ventricular prominence. (*Figure 1*). ECG result was left axis deviation with sinus rhythm with a rate of 107 bpm. (*Figure 2*). 2D echo showed presence of congenital heart disease, left isomerism common atrium, pulmonary stenosis, and patent ductus arteriosus. During regular check-up, the patient was noted

to be bradycardic although asymptomatic. At 4-5 years old, she has easy fatigability with sweating after 10 minutes walk. Pertinent physical examination revealed dynamic precordium, apex beat at the 5th ICS LMCL, no heave, no thrill, S1 normal S2 split, P2 soft, grade 3/6 systolic murmur in the left upper sternal border with dusky nail beds.

The patient underwent cardiac catheterization for pressure studies which revealed left atrial isomerism, interrupted IVC and azygos draining to SVC, common atrium, PS valvar, VSD small. Cardiac CT angiography done revealed interrupted lower portion of IVC, and anomalous vein that connects the left renal vein to the left hepatic vein that drains into the interrupted IVC, right sided polysplenic, bilobed lungs and a

heart with a single large atrium and prominent pulmonary artery. On the 5th hospital day, the patient underwent mitral valve repair, creation of atrial partition, pulmonic valve commissurotomy, supravulvar pulmonary artery patching and permanent pacemaker insertion. She was later discharged recovered and improved.

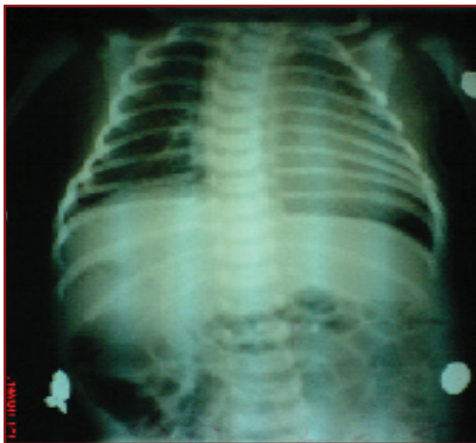


Figure 1a: Frontal chest radiograph of the patient



Figure 1b: Lateral chest radiograph of the patient

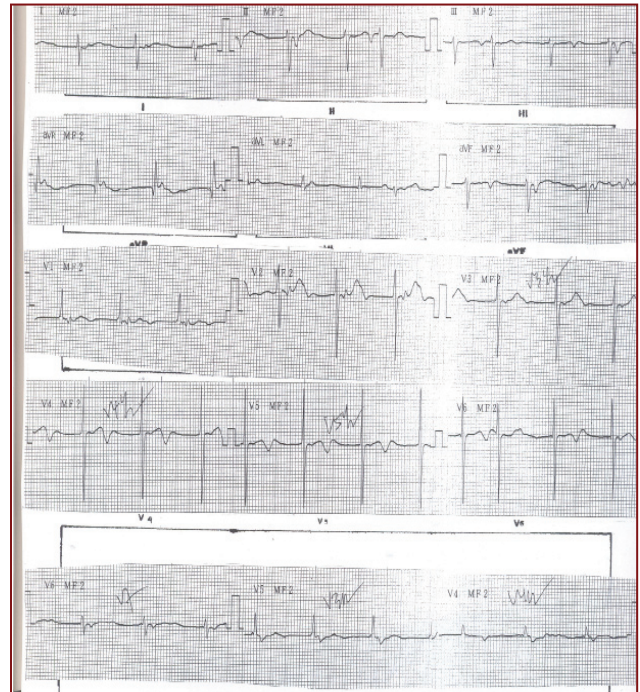


Figure 2a: 12 lead ECG of the patient taken at 1-month of age

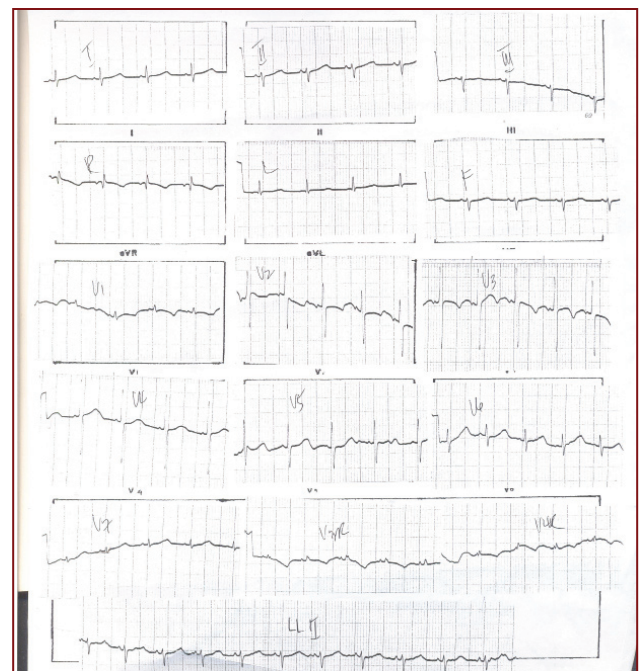


Figure 2b: 12 lead ECG of the patient taken at 5 years of age

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heart failure symptoms from left to right shunts. The absence of the IVC on lateral chest film as well as the continuation of the azygos vein in the frontal chest film with discordance of the apex and abdominal viscera should lead one to suspect this disease entity.

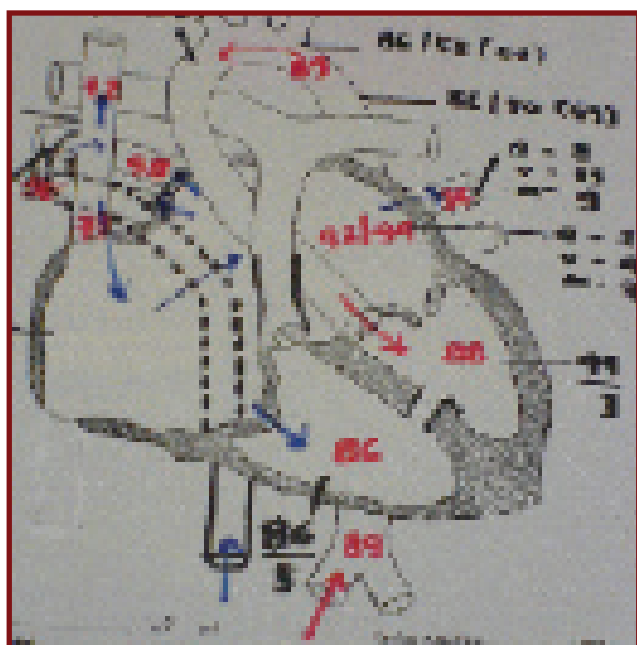


Figure 3: cardiac catheterization data of the patient

DISCUSSION

Epidemiology. US data showed a prevalence estimate of cardiovascular malformations in large cohort from 4-8 per 1000 births. Atrioventricular septal defect (AVSD) accounts for 5-8% of these defects.²

Pathophysiology. Disruption of the development of the primitive heart and venous connection at 20-30 days gestation, when the cardiac chambers are incompletely septated, is the underlying reason why there is a "preponderance of common atria, single ventricle, abnormal pulmonary venous connections, and conotruncal anomalies observed in heterotaxy syndrome."³ Hutchinson et al.⁴ explained that changes in the embryonic body curvature account for the spectrum of anatomy seen in the heterotaxy syndrome, which results primarily from a defect in lateralization, leading to "failure of normal asymmetric development."

Literature³ cited that in patients with the classic left isomerism, there is 'bilateral bilobed lungs, bilateral pulmonary atria, centrally located liver, a stomach in indeterminate position and multiple spleen' (*Polysplenia*). This condition is more common in females and more present with

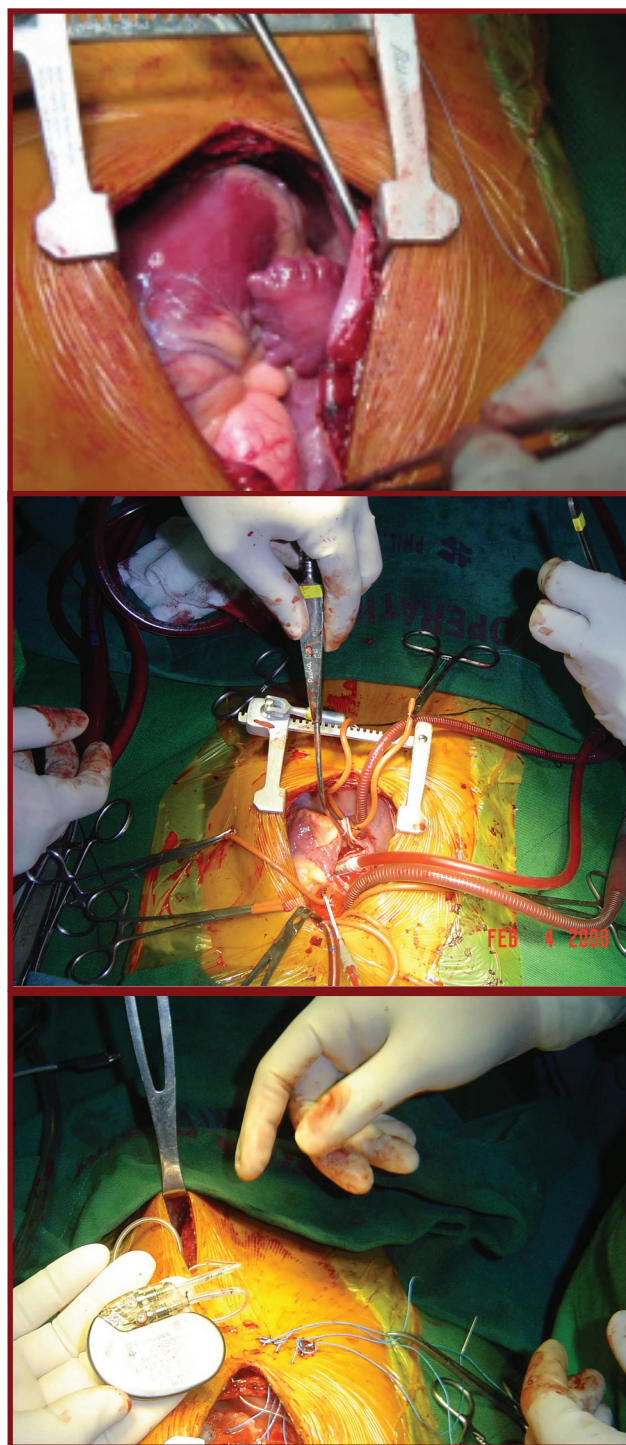


Figure 4: surgical repair of the patient

Common atrium involves the failure of development of the endocardial cushions that are responsible for the septation of the atrium. The common pathophysiology is a left to right shunting at the level of the atrium, ventricles or both.

Without the shunting at the level of ventricles, the hemodynamics is like that of ASD causing volume overload of the right atrium and ventricle.⁵

In patients with almost absent atrial septum or common atrium, there is mixing of the systemic and pulmonary venous returns due to absence of the septum and high incidence of anomalous venous connections, especially in those with isomerism of the atrial appendages. The blood from the left superior caval vein empties directly into the left upper corner of the atrium flowing into the coronary sinus and into the right atrium. The direction of the flow in the presence of a large ventricular component will depend on the resistance to the outflow from the two ventricles. Any obstruction to the right ventricular outflow tract or pulmonary vascular obstructive disease will favor right to left shunting.⁵

The size of the shunt through the atrial defect is ascertained by the size of the defect and the relative compliance of the two atria and ventricles. When the defect is extremely large, there is an obligatory mixing in a common or near common atrium, creating a component of right-to-left shunting. The degree of left-to-right shunting increases with age as the compliance of the right ventricle increased and the pulmonary vascular resistance decreases, resulting to a progressive enlargement of the right ventricle and engorgement of the pulmonary vasculature.²

Clinical Presentation and Work Ups. Most patients with common atrium present in infancy with symptoms of excess pulmonary blood flow, fatigue, tachypnea, and failure to thrive. However, if increased pulmonary vascular resistance develops, the left-to-right shunt decreases and the child will thrive. In general, these patients are

symptomatic earlier in life than patients with only a primum ASD.²

The precordium is hyperactive with a prominent right ventricular impulse. The second heart sound is widely split and fixed with respiration. The intensity of the pulmonary component of the second sound is proportionate to the severity of pulmonary hypertension. A crescendo-decrescendo systolic ejection murmur is present over the upper left sternal border and radiates to the axilla and to the back. A distinct holosystolic murmur of mitral valve regurgitation may be heard at the apex. A middiastolic murmur commonly is detected over the lower left sternal border resulting from an increase in right atrial to right ventricular blood flow. Patients with common atrium frequently have anomalies of cardiac and abdominal situs and asplenia.⁵

The radiographic and electrocardiographic characteristics of patients with common atrium are indistinguishable from those with other forms of AVSD. Chest roentgenography usually shows the prominent pulmonary artery segment and abnormally dense pulmonary vascular markings.²

Frontal chest radiograph of a patient with polysplenia shows the continuation of the azygos vein whereas the lateral chest film shows the absence of the inferior vena cava stripe.³

On electrocardiogram, superior orientation of the frontal QRS axis -30 to -135 (-90 to -120) with left axis deviation to the left or right upper quadrant may be seen together with right bundle branch block or right ventricular hypertrophy.⁶

On echocardiography, the subcostal four-chamber imaging plane is most suitable for accurate diagnosis to see the absence of atrial septum. A muscle bundle or band coursing through the atrium should not be interpreted as an atrial septum. The color flow recognizes the localization of the anomalous pulmonary venous connections and magnitude of associated AV valve incompetence.

Some authors advocate the use of three

dimensional (3D) echocardiography to provide excellent images of the AV valve morphology and relationships with the rest of the cardiac structures.⁷

Cardiac magnetic resonance imaging is now being utilized more often since it provides a more precise delineation of anatomy and evaluation of function with this noninvasive method as compared with either echocardiography or angiography alone.²

On cardiac catheterization and angiography, the hemodynamic diagnosis of common atrium depends on the demonstration of complete mixing of systemic and pulmonary venous blood. The oxygen saturations of pulmonary and systemic arterial blood are nearly identical. Pulmonary blood flow exceeds systemic flow, except in patients with severe pulmonary vascular obstructive disease. Right ventricular pressure is increased more often than in secundum ASD or partial AVSD. If definitive repair is delayed, significant pulmonary vascular obstructive disease may develop more easily than in patients with secundum ASD or partial AVSD. Typical gooseneck deformity can be seen in the left ventriculogram.⁶

Management. Prior to surgical repair, medical therapy usually is instituted when signs and symptoms of excess pulmonary blood flow and failure to thrive are present. Digoxin and diuretic therapy are traditional forms of therapy. Common atrium requires surgical repair, which should be performed early in life because the patient usually has symptoms and is at risk for developing pulmonary vascular obstructive disease.⁵

Management is primarily surgical and involves the following: patch closure of the atrial septal defect, mitral valve annuloplasty or cleft closure. Repair of other defects may be done during the same operation.² Repair is usually done electively in children aged 2-5 years of age. However, it is indicated earlier when the mitral regurgitation is significant.²

A special surgical consideration is avoidance of injury to the conduction system, for there are no marks by which the location of the AV node and his bundle area can be visually confirmed.⁸

Left isomerism is associated with much longer survival, its constellation of associated malformations frequently being less severe and more amenable to corrective surgery.³

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