

Two Chambered Heart with Situs Ambiguous Rarest Case Report

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ABSTRACT

We present a rare case of two chambered heart (common atrium with common ventricle) in situs ambiguous in a 2-month-old male child. In view of dyspnea, fatigue, and central cyanosis, the patient was referred to us with a provisional diagnosis of congenital cyanotic heart disease. The baby had a body weight of 3.0 kg and an oxygen saturation of 80% at room air. The auscultatory findings included a single first and second heart sound and soft Grade II/VI ejection systolic murmur. Plain radiograph of the chest in posterior-anterior view revealed the cardiac silhouette on the midline of the thorax with base to apex axis to left suggestive of mesocardia. Trachea bifurcating into morphologically similar bronchi (bronchi are long and more vertical? Right sided), liver is transverse with absent fundic bubble shadow on the left side. Chest radiograph goes in favor of right isomerism. Echocardiogram revealed inferior vena cava (IVC) on left and aorta on right, a common atrium, single ventricle of left ventricular morphology, common atrioventricular valve with moderate regurgitation, IVC opening into right side of common atrium, aorta on left and anterior, pulmonary atresia, left-sided aortic arch with large patent ductus arteriosus. Again, these anomalies are commonly associated with right isomerism. Ultrasonography of the abdomen showed absence of spleen with midline liver and IVC and aorta on the same side of spine favouring right isomerism. The patient was posted for a palliative Blalock–Taussig shunt surgery but died suddenly.

Keywords: Cyanotic heart disease, heterotaxy, mesocardia, right isomerism, situs ambiguous

INTRODUCTION

It has long been recognized that many of the most complex cardiac lesions are found in the hearts of patients with an unusual arrangement of abdominal organs, or so-called heterotaxy. It is also well-recognized that one of the dominant features of such patients is either absence of the spleen or the presence of multiple spleens on each side of the dorsal mesogastrium.¹

Grouped under the broad category of situs ambiguous defects, these often pose diagnostic difficulties due to the confusing anatomy. Since patients rarely survive into adulthood due to cardiovascular complications, natural history of such conditions are not fully understood.

Right isomerism in the context of the congenitally malformed heart is defined as a subset of heterotaxy where some paired structures on opposite sides of the left-right axis of the body are symmetrical mirror images of each other, and have the morphology of the normal right-sided structures. Right atrial isomerism (RAI) is frequently observed in association with a common atrium, a common atrioventricular (AV) valve, and pulmonary outflow tract obstruction.²

Interestingly, Asians show a higher prevalence of heterotaxy syndrome (32%) compared to Westerners.³

CASE REPORT

A 2-month-old male baby born out of non-consanguineous marriage full term normal hospital delivery with uneventful antenatal course with no other obvious external malformations. He was the second-born child and his parents had no history of congenital disease. In view of dyspnea, fatigue, and central cyanosis, patient was referred to us with a provisional diagnosis of congenital cyanotic

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heart disease. The baby had a body weight of 3.0 kg and an oxygen saturation of 80% at room air. The auscultatory findings included a single first and second heart sound and soft Grade II/VI ejection systolic murmur.

Plain radiograph of the chest in posterior-anterior view revealed the cardiac silhouette on the mid line of the thorax with base to apex axis to left suggestive of mesocardia. Trachea bifurcating into morphologically similar bronchi (bronchi are long and more vertical? Right sided), liver is transverse with absent funding bubble shadow on the left side (Figure 1). Chest radiograph goes in favor of right isomerism.

Electrocardiogram (ECG) showed sinus rhythm, left axis deviation with poor R-wave progression with a rate of 150 bpm and P-wave axis of 70° (right sinoatrial node origin) (Figure 2).

Echocardiogram revealed inferior vena cava (IVC) on left and aorta on right, a common atrium, single ventricle of left ventricular (LV) morphology, common AV valve (Figure 3) with moderate regurgitation, IVC opening into right side of common atrium, aorta on left and anterior, pulmonary atresia, left-sided aortic arch with large patent

ductus arteriosus (Video 1). Again, these anomalies are commonly associated with right isomerism.

Ultrasonography of the abdomen showed the absence of spleen with mid line liver and IVC and aorta on the same side of spine favouring right isomerism. The patient was posted for a palliative Blalock–Taussig shunt surgery but died suddenly. If relatives would have permitted for autopsy then diagnosis would have been confirmed.

DISCUSSION

RAI is a severe complex congenital heart defect resulting from embryonic disruption of proper left-right axis determination.⁴ Right isomerism is accompanied by the congenital cardiovascular malformations like common atrium, single ventricle physiology, pulmonary stenosis or atresia, total anomalous pulmonary venous connection, absent coronary sinus. The liver is transverse. The superior vena cavae are bilateral. Bilateral morphologic right bronchi coupled with bilateral morphologic right atrial appendages and bilateral trilobed lungs are present (Figure 4).

The overall outcome of patients with RAI is very poor. RAI is named for its discoverer, Swedish pathologist Biörn Ivemark.

Left isomerism (LAI) is associated with bilateral superior vena cavae, bilateral morphologic left atria (appendages), absent or atretic sinoatrial node. Also associated with bilateral morphologic left bronchi, bilateral morphologic left lungs. Liver is transverse and stomach is usually right sided. Polysplenia is a common but not invariable feature of LAI. Congenital cardiac malformations associated with LAI are inferior vena caval interruption with azygous continuation, partial anomalous pulmonary venous connection, AV septal defect.

There have been a large number of reported cases that could not be classified into either syndrome. Since about 1990,

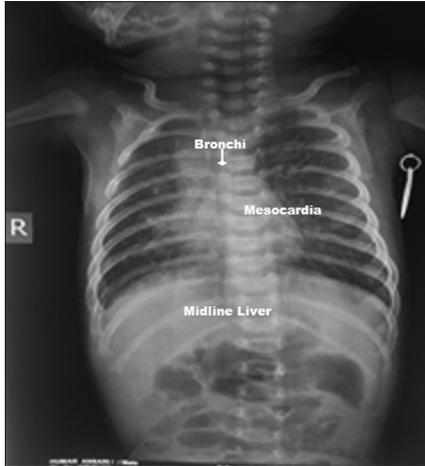


Figure 1. Chest radiograph showing mesocardia, bilateral morphological right bronchi.

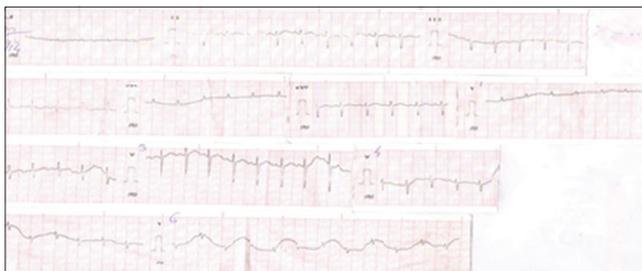


Figure 2. Electrocardiogram showing sinus rhythm.

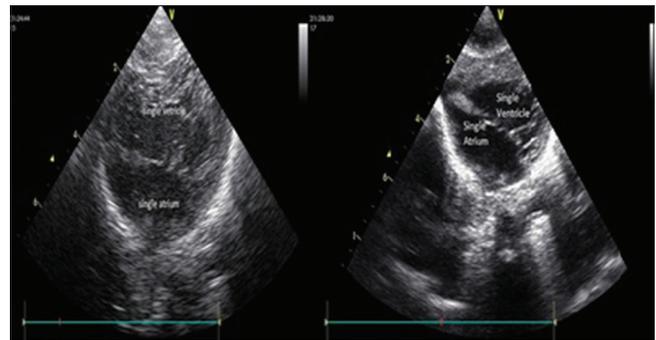


Figure 3. Apical 4C view showing common atrium and common ventricle.

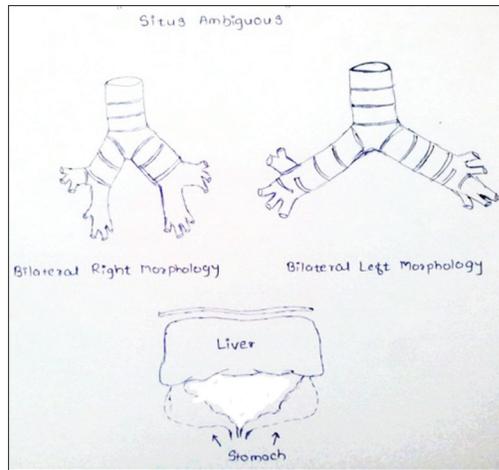


Figure 4. Arrangement of organs in heterotaxy syndrome.

those cases have come to be known as heterotaxy syndrome. Diagnosis of heterotaxy syndrome only requires at least one organ presenting with a left-right differentiation disorder.⁵

In right isomerism, ECGs of the same patient have revealed two types of P waves at different periods: One derived from the right atrium, and the other derived from the left atrium.⁶ On the other hand, in LAI, P-wave derived from the sinus node is uncommon. In addition, although AV block has rarely been observed in right isomerism, 10% of cases of LAI have had coexisting complete AV blocks.⁶

Patients with heterotaxy syndrome present a prevalence of congenital cardiac diseases in 50-100% of the cases.^{7,8} Heterotaxy syndrome with asplenia is associated with congenital cardiac disease in 99-100% of the cases, and it is usually of greater severity as compared with heterotaxy syndrome with polysplenia. Thus, the diagnoses of

heterotaxy syndrome with asplenia are rarely reported in adulthood is explained.⁹

Our patient's clinical and radiological findings go in favor of right isomerism with two chambered heart which is a rare condition. Autopsy would have confirmed the findings. The prognosis of supraventricular with common atrium is poor with high mortality rate of up to 50% in infancy. The median survival of single ventricle of LV morphology is 14 years, while that of the right ventricle morphology is 4 years.⁹ Patients with decreased pulmonary blood flow situation need a modified Blalock–Taussig shunt followed by a bidirectional Glenn operation as the second-stage after 3-6 months and a subsequent Fontan procedure at 2-3 years. The Fontan surgery is usually considered the definitive treatment for such cases which significantly improves the natural history of the disease to a better side.

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