Case Report

Situs ambiguous with three chambered heart (single atrium), evaluation by four-dimensional X-strain color echocardiography – A case report

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ABSTRACT

Situs ambiguous accounts for 4% of all congenital heart disease (CHD) and has an incidence of 1:10,000 newborn births. To standardize the nomenclature for CHD, the international nomenclature society published a globally accepted nomenclature tree for CHD with the 11th Iteration of international classification of disease. According to this publication common atrium, single atrium and atrium communis are denoted as synonyms. The incidence of common atrium among atrial septal defect (ASD) patients is 3–4% only and thus denoting it to be a rare entity. We are presenting an exceedingly rare case of a patient of situs ambiguous, left isomerism, and single atrium with a cleft on the anterior mitral leaflet, evaluated comprehensively by four-dimensional X-strain color Doppler echocardiography.

Key words: Heterotaxy, Levocardia with situs ambiguous, Single atrium, Situs ambiguous

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CASE REPORT

A 5-year-old female child was referred to us from a private pediatric hospital, to rule out the presence of cyanotic CHD. The child was full term delivery by cesarean section, had a normal birth weight, and was delivered at a district hospital, from a primipara woman of 19 year of age. The vaccination of mother and the child was appropriately carried out at the date of presentation to us. There was no history of maternal risk factors of CHD (morbid obesity, diabetes, febrile illness, smoking, alcohol intake, teratogenic drug use, or radiation exposure) on interrogation; her parents gave history of presence of cyanosis since birth, along with recurrent chest infections, failure to thrive and shortness of breath on mild effort. On clinical examination, the child was found to be thin built, irritable, and apprehensive and had frequent bouts of crying while being examined. She had a weight and height of 10 kg and 60 cm respectively, pulse rate was 78/mm, blood pressure was 86/60 mm Hg, and SPO₂ was 75% at room air. The child was deeply cyanosed with bluish coloration of tongue, lips, all the fingers, and toes, together with clubbing. There was a typical pectus carinatum deformity of the chest without any other musculoskeletal anomalies. All the peripheral pulses were normally palpable without any radio femoral delay. Meanwhile, central nervous system, respiratory system, and abdomen were also examined and no abnormality was detected.

On cardiovascular examination, cardiac apex was shifted in the left axillary line, suggestive of cardiomegaly. There was presence of Grade 3/6 pansystolic murmur over left ventricular (LV) apex, radiating to the left sternal edge, and another Grade 3/6 pansystolic murmur was heard at the lower left sternal border, which increased with inspiration, indicative of presence of moderate-to-severe mitral regurgitation (MR), and tricuspid regurgitation (TR), respectively. The second heart sound was closely split and the P2 component was loud. X-ray chest PA view was showing marked cardiomegaly occupying nearly 75% of mediastinum. Moreover, the main and branch pulmonary arteries are strikingly dilated with increased pulmonary blood flow. On more closely scrutinizing the chest X-ray, we found gastric bubble to be on the right side, suggestive of situs inversus/situs ambiguous (Fig. 3).

Echocardiography has substantially expanded the horizons, for accurate non-invasive diagnosis of congenital cardiac defects including complex CHDs. Color Doppler echocardiography [12] not only provides additional information, but also improves overall specificity of two-dimensional (2-D) echocardiography. In current clinical practice, the most often used non-invasive imaging method is echocardiography for sequential analysis of complex CHD. Echocardiography is a non-invasive imaging method that can be performed bedside, no radiation exposure occurs, it is an inexpensive method, and it can be repeated at any time [13]. Transesophageal (TEE) and transthoracic echocardiography (TTE) are reliable in diagnosing most CHD in the pediatric and adult populations [14,15]. Additional imaging is required if the goals of imaging are not achieved with echocardiography. Most often non-invasive imaging options include, cardiac magnetic resonance imaging, and cardiac computed tomography (CT) [16].

We are presenting an exceedingly rare case report of situs ambiguous with three chambered heart (single atrium), comprehensively evaluated by four-dimensional X-strain echocardiography.
Comprehensive four-Dimensional X-Strain Echocardiography

The patient underwent four-dimensional X-strain color echocardiography in supine and left lateral decubitus posture. There were levocardia, concordant D-loop, left aortic arch, and normally related great arteries with confluent pulmonary arterial system. There was noticeable presence of a prominent dilated single atrium, with total absence of remnants of atrial septal tissue, both at the roof of single atrium and atrioventricular junction (Fig. 5).

**Figure 3:** X-ray chest PA – shows massive cardiomegaly with enlarged main and branch pulmonary arteries, suggestive of increase pulmonary blood flow

**Figure 4:** (a) Subcostal view – There is situs ambiguous with liver lying transversely from midline to left hypochondrium, aorta is on the right side and IVC is on the left. (b) ECG suggestive of – left axis deviation with Katz-Wachtel phenomenon characteristic of Biventricular hypertrophy

**Figure 5:** Single atrium with total absence of atrial septal tissue

Superior vena cava (SVC) and IVC were draining into the right lateral side (dextral part) (Fig. 6).

Moreover, three pulmonary veins were opening into the left lateral side (sinistral part) (Fig. 7) of single atrium. The single atrium connects with the left and right ventricle through bileaflet mitral valve (MV) and trileaflet tricuspid valve (TV), respectively (Fig. 8).

Identical left and right atrial appendages were also discerned. Mitral leaflets are large (LAX view) (Fig. 9), with presence of a conspicuous cleft on the anterior mitral leaflet (SAX view) (Fig. 10).

The chordae and papillary muscle of MV and TV were normally attached to the wall of LV and to the ventricular septum and RV apex, respectively.

A classical goose neck deformity was also detected (Fig. 11). There was presence of severe MR, filling up nearly half of single atrium. Two distinct jets of MR were viewed (Fig. 12).

MR jet 1 was produced due to the cleft in the anterior mitral leaflet while, MR jet 2 was due to incomplete coaptation of mitral valve. Immediately after the emergence of MR jets from mitral valve, they merged with each other to produce a large MR jet image in the cavity of single atrium. Total MR jet area was 16.05 sq. cm. (Fig. 13).

Meanwhile, moderately severe TR was also visualized, with a normal tricuspid valve and a dilated tricuspid valve annulus, with an estimated right ventricular systolic pressure/pulmonary arterial pressure to be 59 mmHg. Moreover, there was moderate biventricular dilatation with normal biventricular systolic function; LV ejection fraction was 67%.

**DISCUSSION**

Situs ambiguous is an extremely rare entity, occurring roughly 1/40,000 live births, with a male: female ratio of 2:1 [17]. It does not follow a fixed pattern in all the cases [18]. There are two major clinical sub-divisions of situs ambiguous: (a) Left isomerism and (b) right isomerism. Left isomerism consists of situs ambiguous, polysplenia, two bilateral lung lobes, and identical right and left atrium. Right isomerism comprises situs ambiguous, asplenia, and...
Situs ambiguous with single atrium

three bilateral lung lobes [19]. With situs ambiguous, 90–99% of the patients have severe CHD. Our case had situs ambiguous

with the left isomerism, in the form of the right-sided stomach, polysplenia, both the atrial appendages being similar in morphology and abdominal aorta was on the right side with IVC on the left.
In single atrium patients, the atrial septum is completely absent. The dextral part of the single atrium has the morphology of a right atrium (crista terminalis, pectinate muscle, and right atrial appendage) and receives the superior and inferior vena cava and coronary sinus. The sinistral portion of the single atrium has the morphology of a left atrium (smooth non-trabeculated walls, a left atrial appendage) and receives the pulmonary veins [7].

Single atrium is usually diagnosed in childhood, though adult presentation of the disease has also been reported. Patients present with decrease in exercise tolerance, shortness of breath, cyanosis, upper respiratory tract infections, fatigue, developmental delay, and heart failure [20]. Single atrium may be associated with different ventricular morphologies: Two ventricles, single right ventricle, single left ventricle, or an undifferentiated single ventricle. Associated malformations include intracardiac anomalies (dextrocardia, mesocardiac, levocardial, atrial isomerism, complete AV septal defect, and single AV valve) and anomalies of great vessels (anomalous pulmonary venous connection, left-sided SVC, right-sided aortic arch, persistent ductus arteriosus, pulmonary artery anomalies, and aorta pulmonary collaterals) [21]. It is almost always associated with a cleft in the anterior leaflet of the mitral valve [7]. Our patient had single atrium with identical left and right appendages and a cleft on the anterior mitral leaflet.

The hemodynamic features of shunting in single atrium are very similar to those of a large ASD. However, in patients with single atrium, pulmonary and systemic venous blood tend to mix more easily, and systemic arterial oxygen saturation tends to be lower than in those with a large ASD [6]. The natural course of single atrium is poor. Later, identification of single atrium increases the possibility of progression to arrhythmia, pulmonary vascular disease, or cardiac dysfunction [22]. Moreover, surgical correction can considerably slow progression of the disease [23]. Therefore, early diagnosis is important even in well-tolerated cases.

Detailed anatomic evaluation is mainstay for effective patient management. TEE and TTE play a pivotal role in defining the anatomy of the single atrium and ruling out the classical partial AV septal defect; however, it lacks delineation of the extracardiac structures [14,15]. Patients with single atrium without any associated malformations usually have a good prognosis even without surgical management [20]. However, patients with associated malformations usually require surgical intervention [24].

Isolated anterior mitral leaflet clefts are rare, and isolated posterior mitral leaflet clefts are extremely rare, with only a few cases reported in the literature [25]. Recently, a case series reported, two elderly patients with multiple clefts: one has two clefts on the posterior mitral leaflet, and the other one had one each, on the anterior and posterior leaflet [26]. Mitral valve cleft (MVC) is the most common cause of congenital MR [27], and surgical correction might be an effective treatment for patients with MVC, especially for those with moderate-to-severe MR (even if asymptomatic) [24]. In our patients, there was a single cleft on the anterior mitral leaflet, which along with abnormal coaptation of MV, produced a severe MR occupying nearly 50% of single atrium, thus highlighting, that early surgical correction of this complex anomaly should be contemplated at an intensified manner.

**CONCLUSION**

To the best of our knowledge, this is a first case report of situs ambiguous, let isomerism, single atrium, and cleft on the anterior mitral leaflet, evaluated comprehensively by four-dimensional X-strain color Doppler echocardiography. The presence of severe MR along with intense cyanosis and profound shortness of breath warranted an accelerated action for early surgical correction.

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REFERENCES


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