# Fetal Cardiac Abnormality Left Atrial Isomerism

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*Received: 30 October 2023*

*Accepted: 25 January 2024*

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**DOI 10.5001/omj.2026.20**

## *Abstract*

We report a case of 35 years old G7 P3 pregnant women, who had anomaly scan and Fetal echocardiography (ECHO) that showed Left Atrial Isomerism with interrupted Inferior Vena Cava and hemiazygos continuation into left Superior Vena Cava with fetal bradycardia. The pregnancy ended by lower segment cesarean section at $36^{+4}$ weeks of gestation for patient request. The newborn baby girl required experts Neonatologist and Cardiologist consultation and stayed in neonatal intensive care unit for 4 days after which she was discharged home on stable clinical condition on diuretics with regular follow-ups and plan for surgery at age of 6 months. Left atrial isomerism presents a varied spectrum of cardiac malformations when it is detected prenatally. Complete heart block, complex cardiac abnormalities and fetal hydrops are poor prognostic features. Those with only minor cardiac malformations are at risk postnatally for biliary atresia and for bowel obstruction due to malrotation. This current report presents a case of fetal left atrial isomerism. The condition was identified in pregnancy by expert sonographers and fetal cardiologist despites its complexity. In general, the conditions of a heart with left atrial isomerism seem to be less severe. Hence, we report this case because of rarity of this condition. Left atrial isomerism is a challenging condition that required expert fetal cardiologist input and antenatal counselling. Antenatal monitoring for poor prognostic signs is crucial to set proper plan for intrapartum and postpartum intervention with multidisciplinary team approach.

**Keywords:** Left Atrial Isomerism, Echocardiography, Superior / Inferior Vena Cava, Patent Ductus Arteriosus, Atrio-Ventricular Septal Defect, Ventricular Septal Defect, Non-Stress Test, Neonatal Intensive Care Unit.

## Introduction

The embryologic development of abdominal and thoracic structures follows a spatially controlled and coordinated manner, leading to well-defined right-sided and left-sided anatomic positions within the body. Normal development and positioning of abdominal and thoracic organs is referred to as situs solitus (solitus means common) for the visceral arrangement, and levocardia (heart on the left side) for the thoracic arrangement of organs.[1]

Situs inversus refers to a mirror-image arrangement of the visceral and thoracic structures to that of situs solitus. Any arrangement of visceral and/or thoracic organs other than situs solitus or situs inversus is referred to as situs ambiguous (unknown or complex situs).

Heterotaxy syndrome (in Greek, heteros means different and taxis means arrangement) is a general term that is used to describe the complete spectrum of abnormal organ arrangement, including conditions such as right atrial isomerism (earlier called asplenia) and left atrial isomerism (or earlier polysplenia).

Heterotaxy syndrome, including right and left atrial isomerism, is found in between 2.2% and 4.2% of infants with congenital heart disease. Left atrial isomerism includes a complex spectrum of cardiac and extracardiac anomalies. Hearts demonstrating left atrial isomerism usually are associated with complex congenital cardiac malformations and also has noncardiac anomalies.

Fetal heterotaxy has an increased risk of recurrence in subsequent pregnancies, and that risk has been reported in up to 10% in some series. A genetic etiology for heterotaxy recurrence that may include autosomal dominant, autosomal recessive, X-linked, and single gene disorder, especially primary ciliary dyskinesia, has been suggested.

The identification of the right or left atrial appendage on prenatal ultrasound may be possible in some conditions. but cannot be reliably used for classification. One of the most reliable signs remains the evaluation of vessel arrangement in the upper abdomen as is commonly used in postnatal echocardiography. In general, a reliable suspicion of right or left isomerism can be attained by assessing vessel arrangement in the upper abdomen in combination with intrathoracic findings. The main impact on outcome, however, is primarily dependent on the specific cardiac malformation rather than the actual classification.[2]

There are four common ways in which fetal isomerism is suspected and confirmed:

1. Fetal heart and stomach on opposing sides of the body, leading to the suspicion of situs abnormality and thus directing detailed ultrasound examinations of the chest and abdomen.

2. The presence of complex cardiac malformation on an ultrasound examination, prompting a targeted segmental evaluation of the fetal situs and heart.

3. The presence of complete heart block or other fetal arrhythmia with or without fetal hydrops on an ultrasound examination, which leads to a targeted evaluation of cardiac and abdominal anatomy.

4. The detection of an anomaly of the veno-atrial connection affecting either the inferior vena cava or the pulmonary veins, leading to a segmental evaluation of the cardiac anatomy.

In right atrial isomerism, the two lungs are morphologic right lungs with three lobes each. The liver and stomach can be left-sided, right-sided, or in the midline position. There is commonly asplenia in association with right atrial isomerism. In many cases of right isomerism, abnormal connection of the pulmonary veins can be present.

In left atrial isomerism, the two lungs are morphologic left lungs with two lobes in each. The location of the liver and stomach can be left-sided or right-sided. Polysplenia is also commonly seen. In many cases of left atrial isomerism, there is an interruption of the intrahepatic part of the inferior vena cava with azygos continuation.[3]

The prognosis of right and left isomerism detected in the fetus is generally poor, owing to the severity of the cases detected antenatally.[2] Complete heart block occurs in 40% to 70% of left atrial isomerism cases. Fetuses with left isomerism and heart block are at risk of in utero death following the development of hydrops.[3] On the other hand, newborns with left isomerism and a mild form of cardiac anomaly have an excellent prognosis.

This report presents the case of 35 years old G7 P3 pregnant women, who had anomaly scan and Fetal ECHO that showed Left Atrial Isomerism with interrupted Inferior Vena Cava and hemiazygos continuation into left Superior Vena Cava with fetal bradycardia. A diagnostic amniocentesis was offered but she declined it.

The patient has 3 healthy children at ages of 10, 11 and 7 years, all born by uncomplicated spontaneous vaginal delivery.

The pregnancy ended by lower segment cesarean section at $36^{+4}$ weeks of gestation for patient request. The newborn baby girl required experts Neonatologist and Cardiologist consultation and stayed in neonatal intensive care unit for 4 days after which she was discharged home on stable clinical condition with regular follow-ups at advanced cardiac center in the country.

A written informed consent was obtained from the patient.

## Case Report

35 years old women, G7 P3 with previous 3 miscarriages, with no past medical or surgical history. She is healthy with no family history of cardiac diseases. Husband is 35 years old, healthy, Non-consanguineous marriage. Normal booking investigations. Seen first at 28 weeks of gestation in Fetal medicine clinic at Sultan Qaboos University Hospital, as a referred case from secondary care hospital because anomaly scan showed suspected Truncus arteriosus versus Tricuspid atresia with narrow right and left ventricle outflow tract.

Patient was referred to Royal Hospital for expert Fetal ECHO at 30 weeks which revealed Left Atrial Isomerism (LAI) with interrupted Inferior Vena Cava (IVC) and hemiazygos continuation into left Superior Vena Cava (SVC). Unable to see Right SVC, Levocardia, stomach on the left, Atrio-Ventricular Septal defect (AVSD) [Figure 1] with common atrium and Moderate size Ventricular Septal Defect (VSD), Small TV at this stage with good flow but need assessment. Trivial common valve regurge, at least two pulmonary veins are draining into left side of common atrium. Normal crossing of great arteries, patent both outflow tract.

The Echo Concluded the findings of Complex CHD with LAI, AVSD, small RV, and fetal bradycardia [Figure 2].

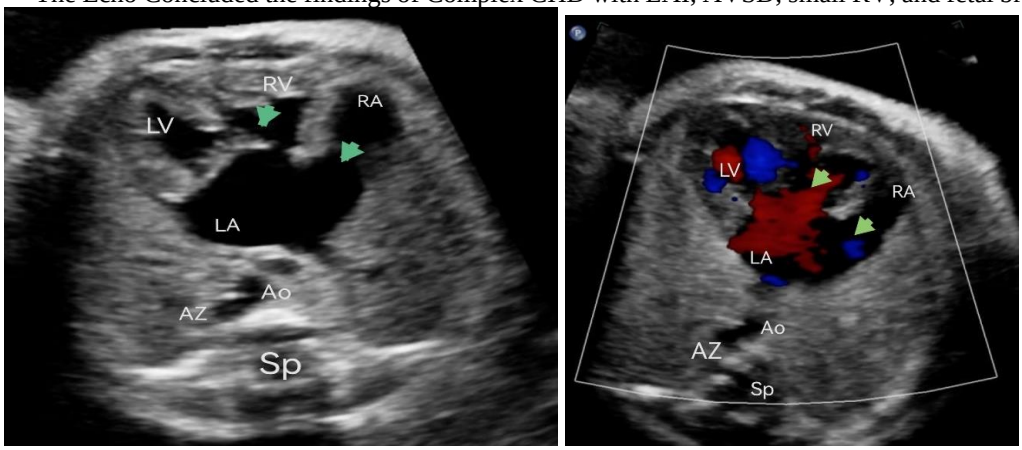


**Figure 1:** Four chambers view of the heart showing the right side of the heart is smaller compared to the left side and the AVSD (arrows). Descending aorta (Ao), azygous vein (AZ) posteriorly. RV: Right ventricle, LV: left ventricle, RA: right atrium, LA: left atrium, Sp: spine.

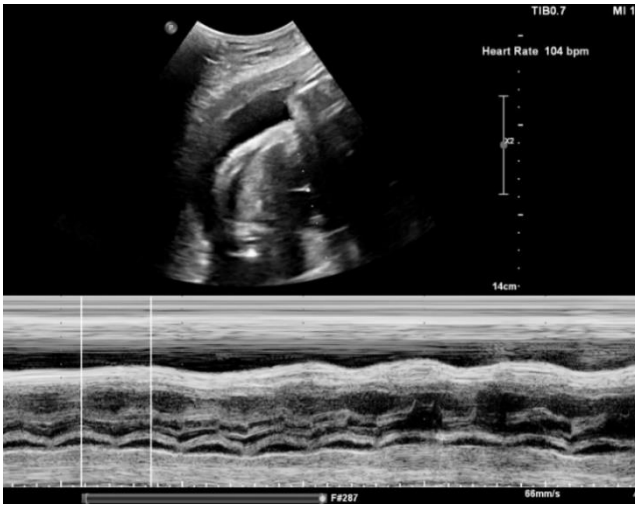


**Figure 2:** Bradycardia, fetal heart rate is 104 with regular beats.

She had regular antenatal visits at fetal medicine clinic, every two weeks. Each visit fetal scan and Non-Stress Test (NST) done to asses fetal wellbeing. Clinical Genetics opinion obtained at initial presentation, advised for karyotype and DNA extraction and storage at time of delivery.

The initial presenting picture was not promising, poor prognosis was discussed with the mother and the plan for no monitoring in labor and no resuscitation was documented.

A follow up Fetal ECHO done at 34 weeks revealed: LAI with interrupted IVC and hemiazygous continuation into left SVC. Unable to see right SVC [Figure 3]. Levocardia. stomach to left. AVSD with common atrium and moderate size VSD. small RV at this stage with good flow but needs assessment postnatally. Trivial common valve regurge. at least two pulmonary veins are draining into left side of common atrium, normal crossing of great arteries, patent both outflow tracts. The pulmonary has flow acceleration with very mild stenosis PG 20 mmhg [Figure 4 and 5]. Left aortic and ductal arches. patent aortic arch. fetal bradycardia (ow atrial rhythm) due to left isomerism. qualitatively good ventricular systolic function.no pericardial effusion at this stage.

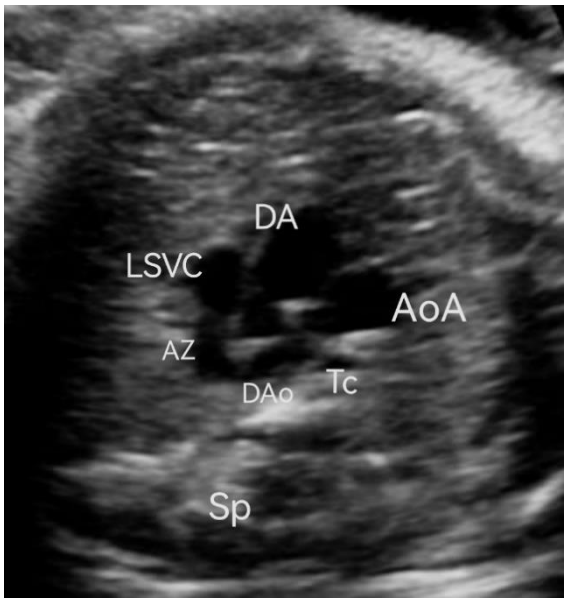


**Figure 3:** Three vessel trachea view demonstrating persistent superior vena cava (PLSVC) with absent right superior vena cava. AOA: aortic arch, DA: Ductal arch, TC: trachea, DAO: descending aorta, AZ: azygous vein, Sp: spine.

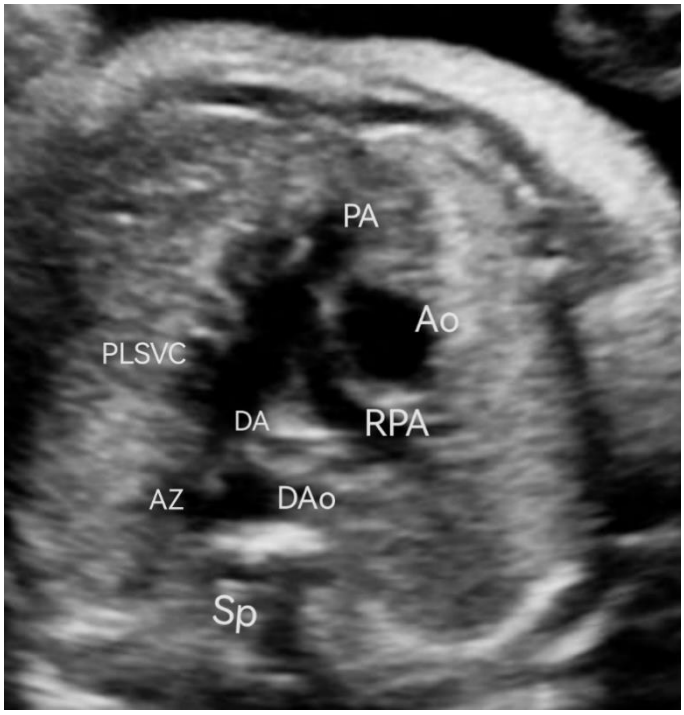


**Figure 4:** Right ventricular outflow tract view demonstrating the pulmonary artery with its branching, RPA: right pulmonary artery and DA: ductus arteriosus. The persistent left superior vena cava (PLSVC), Hemizygous vein (HZ), Descending aorta (DAO), Sp: spine.

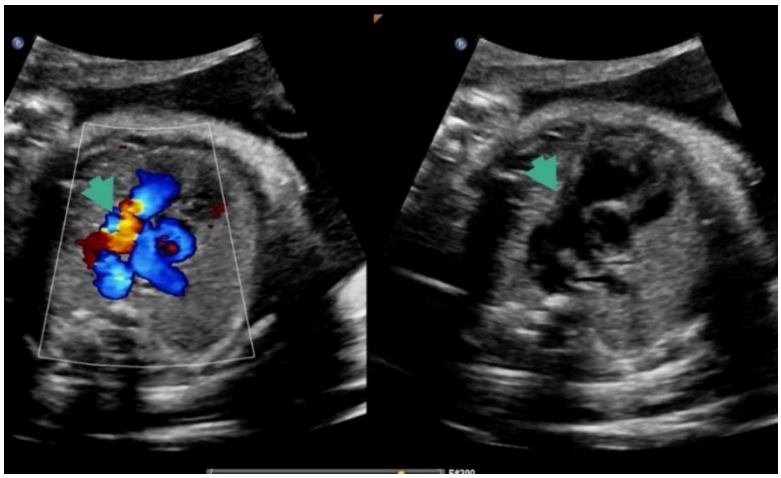

**Figure 5:** Demonstrating right ventricular outflow view with color Doppler showing acceleration in pulmonary valve with mild stenosis.

The scan showed stability in the condition with no hydrops and preserved cardiac function. A consensus made between Fetal medicine consultant, neonatology consultant and Pediatric cardiologist that, full monitoring will be given to the fetus and after birth admission to NICU for Echo and expert intervention accordingly.

Weekly follow up showed progressive increase in liquor from AFI 25cm and 33cm at 32 and 35 of gestation respectively. And non-stress test (NST) done at 35 weeks of gestation showed fetal baseline 80-100 beats per minutes. Another follow-up given at 36 weeks where scan done and revealed active fetus with normal doppler and Deep vertical pocket (DVP) 8 cm.

Patient was insisting on delivery by lower segment cesarean section. A plan was made for delivery at 37 weeks of gestation. But neonatologist requested the cesarean to be done at the beginning of the week as the newborn will need a lot of workup which is difficult to be performed at the end of the week. Patient admitted for the operation and cesarean section performed at $36^{+4}$ weeks of gestation. The mother recovered fully after the operation and discharged home on day two. The newborn baby girl born active with APGAR score of 9 at 1min and 10 at 5min, no dysmorphic features, and did not require any resuscitation. Oxygen saturation 93% in room air, Heart rate136/min and birth weight 2.88kg.

The baby was shifted to NICU for more workup and evaluation, required CPAP initially with low settings. After 4 hours of life, the baby was evaluated by the cardiologist. Chest X-ray reported Cardiomegaly with Mildly increased vascular markings, midline liver with gastric bubble on left. ECHO done and revealed Left atrial isomerism, interrupted IVC, absent right SVC, Large hemiazygous to left SVC to roof of common atrium, common atrium, common atrioventricular valve, mild atrioventricular valve regurgitation, moderate sized inlet VSD, smaller apex forming right ventricle, mild pulmonary stenosis and small PDA.

The newborn required CPAP for one day, weaned to High Flow Nasal Canula (HFNC) second day, weaned to room air at day 3 of life with full tolerable feed and did not required antibiotics. She remained hemodynamically stable with normal heart rate and Blood pressure. 12 lead ECG showed ectopic atrial rhythm and had indirect hyperbilirubinemia (G6PD setup) improved after phototherapy.

Ultrasound head and abdomen done were normal. Karyotyping sent and resulted as normal female karyotype 46XX. The baby discharged home in good condition on day 4 of life, on frusemide 1.5 mg BID and Spironolactone 1.5 mg OD.

After discharge, baby underwent regular follow up by the cardiologist in advanced center specialized in cardiac surgeries in Oman, with repeat ECHO, ECG, and optimizing the diuretic doses according to the baby weight and needs. Lastly seen at 3 months of age with proper development, good head support, social smile and weight gain. A discussion regarding complex surgical correction at age of 6 months is ongoing with the family.

## Discussion

This current report presents a case of fetal left atrial isomerism. The condition was identified in pregnancy by expert sonographers and fetal cardiologist. A published case on 2015 by Simona Duta and his colleagues showed that left atrial isomerism can be detected as early as 12 weeks of gestation and it might be associated with increased risk of fetal demise.[4]

One of the major feature of left atrial isomerism is Interrupted IVC with azygous continuation into the SVC,[5] which is noted as well in this case. Interruption of the IVC with azygous continuation can be detected prenatally as a "double vessel" sign. The sign demonstrates the azygous vein posterior to the descending aorta with the flow of blood in the opposite direction.

The incidence of trisomies in left atrial isomerism is rare, with occasionally reported 22q11 microdeletion. Our case chromosomal study showed normal 46 XX karyotype. In 2020 a case report was published by Aurora Ilian and his colleagues,[6] revealed that. The antenatal detection of corroboration between different structural abnormalities using serial ultrasound examinations and cardiac abnormalities, together with the detection of the affected chromosomes, improves the genetic counseling regarding the prognosis of the fetus and the recurrence rate of the condition for siblings.

Good prognosis is reported for newborns with antenatally mild, isolated cardiac abnormalities with present gallbladder, no heart block, and no other associated structural abnormality. Both cardiac (AV block, SV and coarctation of the aorta) and noncardiac (biliary atresia, other gastrointestinal malformations and low birth weight) anomalies contribute to a high mortality with LAI.[7]

Conclusion

Left atrial isomerism is associated with the presence of "double" left-sided structures with the underdevelopment or absence of right-sided structures. Cardiac defects are less severe in left atrial isomerism and include AVSD, double outlet right ventricle, and others. It is a challenging condition that required expert fetal cardiologist input and antenatal counselling. Antenatal monitoring for poor prognostic signs is crucial to set proper plan for intrapartum and postpartum intervention with multidisciplinary team approach.

We got an opportunity to present such rare case as no previous local studies were addressed similar condition.

## Disclosure

The authors declare that they have no conflict of interests. A written informed consent was obtained from the patient.

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