Journal of Shahrekord University of Medical Sciences
doi: 10.34172/jsums.1027 2025;27(3):142-145 http://j.skums.ac.ir

Case Report

Reporting a case of isolated levocardia

Tahereh Heydarian[1], **Maryam Allahdadian**[2*]

[1]Department of Midwifery, School of Nursing and Midwifery, Isfahan University of Medical Sciences, Isfahan, Iran
[2]Department of Nursing and Midwifery, Fal.C., Islamic Azad University, Isfahan, Iran

***Corresponding Author:** Maryam Allahdadian, Email: Maryamallahdadian@iau.ac.ir

Abstract
Isolated levocardia with situs inversus is a rare congenital anomaly characterized by the abnormal positioning of the heart and abdominal organs. This condition is frequently associated with congenital heart defects and typically carries a poor prognosis. Accurate diagnosis and effective management are crucial for improving clinical outcomes. This report presents a case involving a 25-year-old woman in her first pregnancy who underwent a routine ultrasound at 17 weeks and 2 days of gestation. The ultrasound revealed dextrocardia with situs inversus, prompting a referral to a pediatric cardiologist. Subsequent echocardiography at 18 weeks confirmed the presence of the stomach on the right side and a reversed heart axis, leading to a diagnosis of isolated levocardia with atrioventricular septal defects (AVSDs). The pregnancy was medically terminated at 19 weeks, following appropriate medical approval.
Keywords: Levocardia, Situs inversus, Congenital heart disease, Prenatal diagnosis, Fetus

Received: October 24, 2024, **Revised:** November 13, 2024, **Accepted:** November 16, 2024, **ePublished:** September 8, 2025

Introduction

Isolated levocardia is a rare positional anomaly characterized by the presence of the heart on the left side of the chest, accompanied by visceral heterotaxy. This condition occurs in approximately 1 in 22 000 live births (1). Situs anomalies, which occur in roughly 1 in 10 000 live births, are characterized by the abnormal positioning of major abdominal organs (2). In such cases, the abdominal viscera are located in positions opposite their usual anatomical locations. When the cardiac axis is directed to the left, the condition is referred to as situs inversus with levocardia. Conversely, when the cardiac axis points to the right, it is termed situs inversus with dextrocardia (3).

Approximately 0.4% to 1.2% of patients with congenital heart disease present with isolated levocardia. The five-year survival rate for these patients ranges from 5% to 13%. The prognosis is notably poorer in cases with multiple cardiac anomalies. Only 6% of patients with isolated levocardia survive beyond five years, while 75% die within the first year of life (4). Isolated levocardia is rarely symptomatic during pregnancy and is typically diagnosed postnatally (5). Recent studies underscore the clinical and genetic heterogeneity associated with isolated levocardia. Research conducted by Ding et al indicates that many patients with isolated levocardia may possess underlying genetic abnormalities such as Kartagener syndrome, thereby highlighting the necessity for comprehensive genetic evaluations (6). Additionally, a study by Eitler et al proposed that a multidisciplinary management approach, involving cardiologists, geneticists, and surgeons, can improve long-term outcomes for patients with isolated levocardia (2).

Given the rarity of this condition, clinical reports remain limited. Each case report plays a crucial role in advancing physicians' understanding, particularly when anomalies are identified during pregnancy. Accurate documentation of such cases is essential for refining diagnostic and therapeutic strategies. These findings highlight the importance of early diagnosis and effective management of these anomalies, which can significantly improve treatment outcomes and reduce mortality rates in affected patients.

This case report aimed to highlight a rare instance of isolated levocardia with situs inversus identified during pregnancy and to explore the associated diagnostic and therapeutic challenges. In doing so, it sought to contribute to improved patient management and informed clinical decision-making.

Case Presentation

The patient was a 25-year-old woman in her first pregnancy who presented to a midwifery clinic for prenatal care. Her body mass index (BMI) was 23, and her blood type was A-. She had a history of hypothyroidism, managed with a daily dose of 50 micrograms of levothyroxine. She had no history of infertility, and age has been married for two years. Her husband, a third-degree relative, had no significant medical history. The patient had received standard prenatal care and reported no family history of congenital heart disease.

Given the association between isolated levocardia and

genetic abnormalities, comprehensive genetic testing is strongly recommended to assess potential underlying syndromic associations. Although existing studies indicate that patients with isolated levocardia may be at an increased risk for genetic syndromes, such as Kartagener syndrome and other heterotaxy syndromes, which are characterized by abnormal organ positioning and often associated with respiratory, gastrointestinal, or cardiac complications, genetic evaluations of the fetus revealed no abnormalities. All ultrasound examinations were normal, except for the anomaly scan conducted on September 10, 2023, at 17 weeks of gestation. The anomaly scan indicated dextrocardia with situs inversus, prompting a referral to a pediatric cardiologist for fetal echocardiography at 18 weeks of gestation (Figure 1).

The echocardiography revealed that the stomach was located on the right side, with the heart axis oriented to the left, indicating isolated levocardia. This condition was further associated with an atrioventricular septal defect (AVSD). After two weeks of follow-up and additional evaluation, the diagnosis of isolated levocardia with situs inversus, a rare anomaly, was confirmed. During counseling sessions with the patient and her family, all potential risks and considerations related to either continuing or terminating the pregnancy were thoroughly discussed. These consultations provided comprehensive information about the fetal cardiac condition, including the severe congenital heart defects associated with isolated levocardia, which could significantly affect the fetus's quality of life and long-term health outcomes. Furthermore, concerns regarding potential maternal complications and the potential need for urgent medical interventions in the future were addressed. The discussions aimed to ensure that the patient and her family were fully informed and supported in making a decision that aligned with their values and preferences. Given these considerations, and following a comprehensive evaluation by a multidisciplinary team of specialists in obstetrics and pediatric cardiology, it was decided to proceed with a therapeutic abortion to prevent foreseeable risks. In the 19th week of pregnancy, after obtaining approval from the forensic department, the patient was admitted to the hospital for the procedure.

Situs: **Inversus**
AV connection: **Concordance**
VA connection: **Concordance**
Heart Rate: **152min. PR interval=88msee**
Dysrhythmia: **Absent**
LV contractility: **NL**
RV contractility: **NL**
LV size: **NL**
RV size: **NL**
LA size: **NL**
RA size: **NL**
Aortic valve: **NL**
Pulmonary valve: **NL**
Mitral valve: **NL**
Tricuspid valve: **NL**
Ventricular septum: **Inlet VSD**
Atrial septum: **ASD primum**
Coarctation of aorta: **NO**
Pericardium: **NL**
C/T Ratio: **30%**

Fetal Echo Diagnosis: **Fair echo view, the stomach bubble was seen in the right and at the opposite side of heart axis as (isolated levocardia), complete AVSD, NRGA, moderate AVVR ..**

Plan: Follow-up and reevaluation by another colleague

Figure 1. Fetal Echocardiography Report

Discussion

Situs inversus with levocardia is a rare anomaly characterized by the heart being positioned on the left side of the chest while the abdominal viscera are reversed. This condition is believed to result from incomplete expression of an autosomal recessive gene, although the exact etiology remains unclear (4). Situs inversus may be incidentally diagnosed during unrelated examinations or remain asymptomatic throughout pregnancy, often only being identified postnatally. In our case, this anomaly was detected during pregnancy due to abnormal findings observed in the anomaly scan and subsequent echocardiography.

The term "situs" refers to the arrangement of the atria and abdominal organs in relation to the body's midline. The terms levocardia, dextrocardia, and mesocardia describe the position of the cardiac apex and are distinct from the cardiac structure or the positioning of the abdominal organs (3). Understanding these definitions is crucial for the accurate diagnosis and management of such anomalies, given their significant implications for both maternal and fetal health.

Situs inversus is typically associated with dextrocardia, with a reported likelihood of congenital heart disease ranging from 3% to 5% in these patients (7). However, situs inversus with levocardia, where the cardiac apex is positioned normally on the left side, is extremely rare. The case presented involved situs inversus with levocardia, representing a unique combination. Notably, nearly 95% of situs inversus with levocardia cases are associated with congenital heart disease (4). Diagnosis of this condition can be achieved through a comprehensive medical history, physical examination, chest radiography, electrocardiography, and echocardiography. Depending on the initial findings, further imaging modalities such as computed tomography (CT) or cardiac magnetic resonance imaging (CMR) may be utilized. CT scan provides detailed anatomical visualization of the viscera and cardiac structures but is not recommended during pregnancy due to exposure to ionizing radiation. In contrast, CMR is considered the optimal imaging modality for confirming cases of situs inversus with levocardia, as it avoids radiation exposure and serves as the gold standard for assessing cardiac volume and function (7). This distinction is crucial to ensure maternal and fetal safety while obtaining precise diagnostic information.

Situs inversus with levocardia is generally associated with a poor prognosis, with only 5% to 13% of patients surviving beyond five years. However, rare reports of long-term survival exist, with some individuals living

up to 73 years, particularly in cases where congenital heart disease is absent. Patients with this condition face significant risks, including life-threatening complications such as bowel obstruction and volvulus, which may arise due to the abnormal positioning of the abdominal organs. Additionally, most patients with heterotaxy syndrome, commonly characterized by situs inversus, experience severe congenital heart anomalies, leading to high childhood mortality rates (4).

Goudarzi et al reported a case involving an infant with levocardia, where the heart was positioned on the left side, while the other viscera organs were reversed (5). This highlights the complexity of situs inversus with levocardia, wherein the heart's orientation does not correlate with the typical anatomical positioning of abdominal organs. Campbell et al identified that the most common congenital heart defects associated with isolated levocardia include atrial septal defect, ventricular septal defect, transposition of the great arteries, pulmonary stenosis, and pulmonary atresia (8). In the present case, the fetus exhibited an AVSD, further illustrating the strong association between this condition and congenital heart defects. Likewise, Mesfin et al observed atrial and ventricular septal defects in a 15-year-old adolescent diagnosed with levocardia and situs inversus, reinforcing the notion that cardiac defects are frequently encountered in these cases (4). In contrast, Özdemir et al reported two fetal cases of levocardia in which the heart showed no structural anomalies; however, one fetus did present with sinus syndrome (9).

Wang et al described levocardia as a component of heterotaxy syndrome, which is typically associated with severe cardiac anomalies. In addition to cardiac defects, their findings also noted the presence of other anomalies such as esophageal atresia and polycystic kidney disease (10). This illustrates the complexity of heterotaxy syndrome, wherein multiple organ systems can be affected, leading to significant clinical challenges. Aaron Ong et al reported a patient with levocardia and situs inversus who developed secondary polycythemia and hyperviscosity syndrome (11). These conditions can result from altered blood flow dynamics and oxygenation abnormalities, further complicating the clinical management of patients with these anomalies. Ghawi et al highlighted that levocardia may be associated with serious complications such as bowel obstruction, often necessitating urgent surgical intervention (12).

Nawaz et al reported a neonate presenting with symptoms of bowel obstruction who, in addition to levocardia, was diagnosed with Tetralogy of Fallot, duodenal atresia, and bowel malrotation (13). This case illustrates the complex interplay of congenital anomalies that may occur alongside levocardia, highlighting the potential for significant gastrointestinal and cardiac complications. Abdur Rahman et al described a preterm infant with isolated levocardia who presented with multiple small bowel atresias and obstructive symptoms (14). This case further supports the association between levocardia and gastrointestinal anomalies, which can lead to critical clinical situations requiring immediate surgical intervention. Kumar et al reported a neonate with levocardia and asplenia (15). The absence of the spleen can have serious implications for immune function and susceptibility to infections, necessitating careful monitoring and management for affected patients. Additionally, Luterman et al documented a rare anomaly known as horseshoe lung in a patient with levocardia (16).

Conclusion

Isolated levocardia is strongly associated with various congenital heart defects, making early diagnosis critical for effective clinical decision-making. The identification of this condition during pregnancy enables timely intervention and the development of management strategies that can significantly impact both maternal and fetal outcomes. The findings of this study emphasize the critical importance of identifying rare anomalies such as isolated levocardia during pregnancy. Early detection of these conditions allows healthcare providers to implement appropriate management strategies that can significantly improve therapeutic outcomes for both the mother and the fetus.

Acknowledgments

We would like to express our gratitude to Zahra Maqsoudi (Radiologist), Davood Ramazani-Nejad (Pediatric Cardiologist), and the patient for their invaluable cooperation.

Authors' Contribution

Data curation: Maryam Allahdadian.
Formal analysis: Maryam Allahdadian, Tahereh Heydarian.
Funding acquisition: Maryam Allahdadian, Tahereh Heydarian.
Investigation: Tahereh Heydarian.
Methodology: Tahereh Heydarian.
Project administration: Maryam Allahdadian.
Supervision: Maryam Allahdadian.
Validation: Maryam Allahdadian.
Visualization: Maryam Allahdadian.
Writing –original draft: Tahereh Heydarian.
Writing –review & editing: Maryam Allahdadian.

Competing Interests

The authors declare no conflict of interests.

Ethical Approval

Ethical approval for this study was obtained from the Ethics Committee of Najafabad Islamic Azad University (Ethical Code: IR.IAU.NAJAFABAD.REC.1402.221). Written consent was also obtained from the patient.

Funding

This is a self-funded study.

References

1. Chin Yong A, Wei Chieh JT. Coronary perforation complicating percutaneous coronary intervention - a case illustration and review. ASEAN Heart J. 2013;21(2):3. doi: 10.7603/s40602-013-0002-9.
2. Eitler K, Bibok A, Telkes G. Situs inversus totalis: a clinical review. Int J Gen Med. 2022;15:2437-49. doi: 10.2147/ijgm.S295444.

3. Rao PS, Rao NS. Diagnosis of dextrocardia with a pictorial rendition of terminology and diagnosis. Children (Basel). 2022;9(12):1977. doi: 10.3390/children9121977.
4. Mesfin T, Kedir I, Tilahun T, Seyoum K, Dadi S, Ejigu N, et al. Situs inversus with levocardia in a 15-year-old male adolescent: a case report. J Med Case Rep. 2023;17(1):499. doi: 10.1186/s13256-023-04254-9.
5. Goodarzi R, Rajaei S. Isolated levocardia: a case report. Hormozgan Med J. 2012;16(5):423-7. [Persian].
6. Ding B, Wang J, Wei X, Du Y, Xia L, Sun C, et al. Efficacy and safety of ERCP in patients with situs inversus totalis: multicenter case series and literature review. BMC Gastroenterol. 2022;22(1):497. doi: 10.1186/s12876-022-02593-3.
7. Abdullah NL, Quek SC, Seto KY, Teo LL. Clinics in diagnostic imaging (160). Levocardia with abdominal situs inversus. Singapore Med J. 2015;56(4):198-201. doi: 10.11622/smedj.2015059.
8. Campbell M, Deuchar DC. Dextrocardia and isolated laevocardia. II. Situs inversus and isolated dextrocardia. Br Heart J. 1966;28(4):472-87. doi: 10.1136/hrt.28.4.472.
9. Eriç Özdemir M, Demirci O. Isolated levocardia with situs inversus without cardiac abnormality in fetus: prenatal diagnosis and management. Perinat J. 2020;28(1):48-51. doi: 10.2399/prn.20.0281004.
10. Wang X, Shi Y, Zeng S, Zhou J, Zhou J, Yuan H, et al. Comparing levocardia and dextrocardia in fetuses with heterotaxy syndrome: prenatal features, clinical significance and outcomes. BMC Pregnancy Childbirth. 2017;17(1):393. doi: 10.1186/s12884-017-1579-y.
11. Aaron Ong HJ, Toh SM, Muhammad TJ, Norazrulrizal MN, Sia KK. Complex congenital heart disease: a rare case of situs inversus with isolated levocardia. ASEAN Heart J. 2019;26(4):4-6. doi: 10.31762/ahj1926.0402.
12. Ghawi H, Zghouzi MM, Emahbes TM, Awad SM. Prenatal diagnosis of isolated levocardia and a structurally normal heart: two case reports and a review of the literature. Pediatr Cardiol. 2013;34(4):1034-7. doi: 10.1007/s00246-012-0359-8.
13. Nawaz A, Matta H, Hamchou M, Jacobez A, Trad O, Al Salem AH. Situs inversus abdominus in association with congenital duodenal obstruction: a report of two cases and review of the literature. Pediatr Surg Int. 2005;21(7):589-92. doi: 10.1007/s00383-005-1412-y.
14. Abdur-Rahman LO, Adeniran JO, Taiwo JO. Concurrent dextrogastria, reverse midgut rotation and intestinal atresia in a neonate. J Indian Assoc Pediatr Surg. 2007;12(4):228-30. doi: 10.4103/0971-9261.40843.
15. Kumar A, Singh MK, Yadav N. Dextrocardia and asplenia in situs inversus totalis in a baby: a case report. J Med Case Rep. 2014;8:408. doi: 10.1186/1752-1947-8-408.
16. Lutterman J, Jedeikin R, Cleveland DC. Horseshoe lung with left lung hypoplasia and critical pulmonary venous stenosis. Ann Thorac Surg. 2004;77(3):1085-7. doi: 10.1016/s0003-4975(03)01213-x.