Research Article

SCISET
www.sciset.com

Clinical Trials and Case Reports

2023
Vol. 1 No. 1: 103

Enhancing Prenatal Diagnosis: The Added Diagnostic Utility of Three-Dimensional (3D) Ultrasound and Doppler Angiography Imaging in Left Isomerism

Fujiao He, Yaqin Wang, Yun Xiu, Yixin Zhang, Lizhu Chen*

Department of Ultrasound, Shengjing Hospital of China Medical University, Shenyang, China

Corresponding author:
Lizhu Chen, Department of Ultrasound, Shengjing Hospital of China Medical University, Shenyang, China. E-mail: li_chen@sina.com

Citation: He F, Wang Y, Xiu Y, Zhang Y, Chen L. (2023) Enhancing Prenatal Diagnosis: The Added Diagnostic Utility of Three-Dimensional (3D) Ultrasound and Doppler Angiography Imaging in Left Isomerism. Clin Trial Case Rep. Vol 1(1):103.

Received: June 19, 2023; **Accepted:** July 25, 2023; **Published:** August 02, 2023

Abstract

Objective: To emphasize the significance of utilizing 3D ultrasound and Doppler angiography imaging in the prenatal evaluation of left fetal isomerism.

Methods: A retrospective analysis of volume datasets from 3 fetuses diagnosed with left atrial isomerism was conducted using 3D ultrasound.

Conclusion: We assert that the parasagittal view, showcasing the heart and abdominal vessels, is easily attainable and interpretable, providing a realistic anatomical image without the need for mental reconstruction of spatial relationships. This view proves particularly beneficial in identifying situs anomalies. We advocate for the systematic use of 3D ultrasound in suspected cases of atrial isomerism to enhance our understanding and interpretation of fetal anatomy.

Keywords: Heterotaxy; Isomerism; Polysplenia; Asplenia; Situs ambigus; Situs inversus; Situs solitus, Cardiovascular malformations, Power Doppler imaging, 3D ultrasound. Left atrial isomerism. Malrotation.

Introduction

During early embryonic stages, symmetry is initially observed with respect to the midline. However, during gastrulation, this symmetry is disrupted by cascades of gene activation, leading to the establishment of left-right (LR) patterning and positioning of internal organs and vasculature. This early LR asymmetry is fundamental to the function and positioning of visceral organs within the body [1].

The term "situs solitus" describes the typical asymmetric positioning of cardiac atria and viscera. Defects in LR asymmetric patterning often result in malformations, including heterotaxy syndromes. Heterotaxy syndromes, also known as situs ambiguous, isomerism, asplenia, and polysplenia syndromes, are characterized by abnormal arrangement of thoracic and abdominal organs across the left-right axis, differing from both normal (situs solitus) and mirror-image (situs inversus) arrangements [2].

Heterotaxy syndromes are conventionally categorized into asplenia syndrome or right isomerism, and polysplenia syndrome or left isomerism, based on the status of the spleen and morphology of atrial appendages, bronchi, and lungs. While asplenia syndrome is associated with bilateral right-sidedness, polysplenia syndrome typically involves bilateral left-sidedness. However, spleen presence or absence may not always correlate with these classifications.

The variability in morphological presentations within heterotaxy syndromes makes a single classification challenging. Some clinicians advocate for referring to specific cases as left atrial isomerism, followed by a detailed description, to account for this variability. In this study, we present four cases of left atrial isomerism diagnosed in our department over a 16-month period, aiming to explore definitions, characteristics, and diagnostic approaches for fetal situs anomalies.

Materials and Methods

We present four cases of left atrial isomerism diagnosed in our department over a 16-month period. Through an analysis of these cases and a review of the literature, we aim to explore the definitions and characteristics of left atrial isomerism, while also highlighting the value of 3D ultrasound in the prenatal assessment of atrial isomerism syndromes [3-7].

Examinations were conducted using a Voluson 730 Pro ultrasound system equipped with a volumetric abdominal transducer (4–8 MHz). Stored cardiovascular volumes were prospectively and subsequently analyzed offline by an independent examiner, who was aware of the previous diagnoses of cardiac anomalies obtained via 2D ultrasound. Subsequently, sonography images were transmitted via the internet to a diagnosis reference center (Caen Teaching Hospital, France), where the initial prenatal diagnosis was confirmed or revised. A multidisciplinary team, comprising a pediatric cardiologist, a neonatologist, and a pediatric cardiac surgeon, provided comprehensive prenatal counseling to each expectant mother. Confirmation of the prenatal diagnosis in surviving fetuses was achieved through neonatal echocardiography.

Results

In our study, three patients exhibited interruption of the inferior vena cava (IVC), with severe cardiac malformation observed in one patient and persistent left superior vena cava (LPSVC) in two patients. All four patients displayed ultrasonographic signs indicative of situs ambiguous (left isomerism), with confirmation of the diagnosis in three patients through fetopathological examination. Unfavorable outcomes were observed in two cases: one resulted in pregnancy termination due to severe heart disease, while the other case delivered prematurely at 27+6 weeks' gestation, leading to psychomotor retardation attributed to extreme prematurity. Normal karyotypes were observed in all three cases, with negative results for research 22q11 microdeletion [8,9].

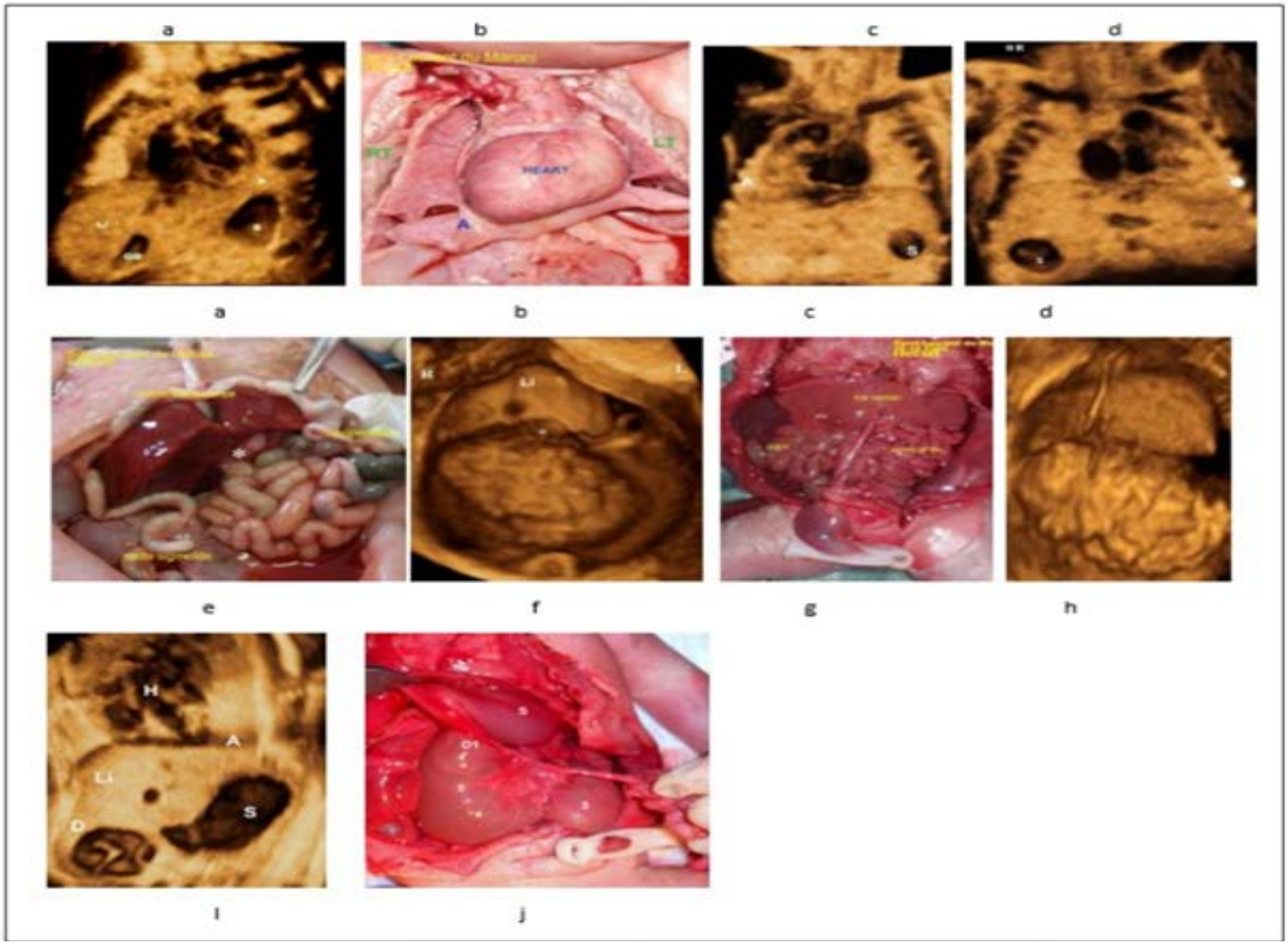

Figure 1: a-Case control, 3D volume of normal situs solitus (28 weeks' Gestation). The heart and stomach are on the left; the gallbladder is on the right, liver is predominantly on the right side.

The "double vessel sign" was consistently identified in all cases with interruption of the IVC and azygos continuation (VCI). Grayscale imaging facilitated the diagnosis of situs, while power Doppler imaging primarily contributed to the prenatal diagnosis of vascular anomalies and aided in detecting situs (Figures 1-2). Additionally, 3D ultrasound and Doppler techniques enhanced visualization of fetal vessels, enabling meticulous offline evaluation and reconstruction of fetal anatomy [10-19].

Among the patients, two had the stomach located on the right side, with one of them exhibiting no congenital heart disease. The images provided depict various echocardiographic and anatomical correlations, including cases of dextrocardia, nonrotation, and median liver position, as well as the characteristic "double-bubble" appearance of the stomach and duodenum. These findings underscore the value of advanced imaging modalities in the comprehensive assessment of fetal situs anomalies.

One patient exhibited dextrocardia, diagnosed via Doppler imaging, where the apex of the heart and the axis of the left hepatic vein were localized on opposite sides. Both the left hepatic vein (LHV) and the apex of the heart were positioned on the same side, pointing downwards. Conversely, in levocardia, the cardiac apex is oriented away from the spine, while in dextrocardia, it is oriented towards the spine. The patients also displayed a liver in the median position, with one patient presenting with the appendix and gall bladder on the left side and the rectosigmoid on the right side. Additionally, one patient showed intestinal nonrotation, with the entire small bowel located in the right abdomen and the colon in the left abdomen. Furthermore, one patient experienced

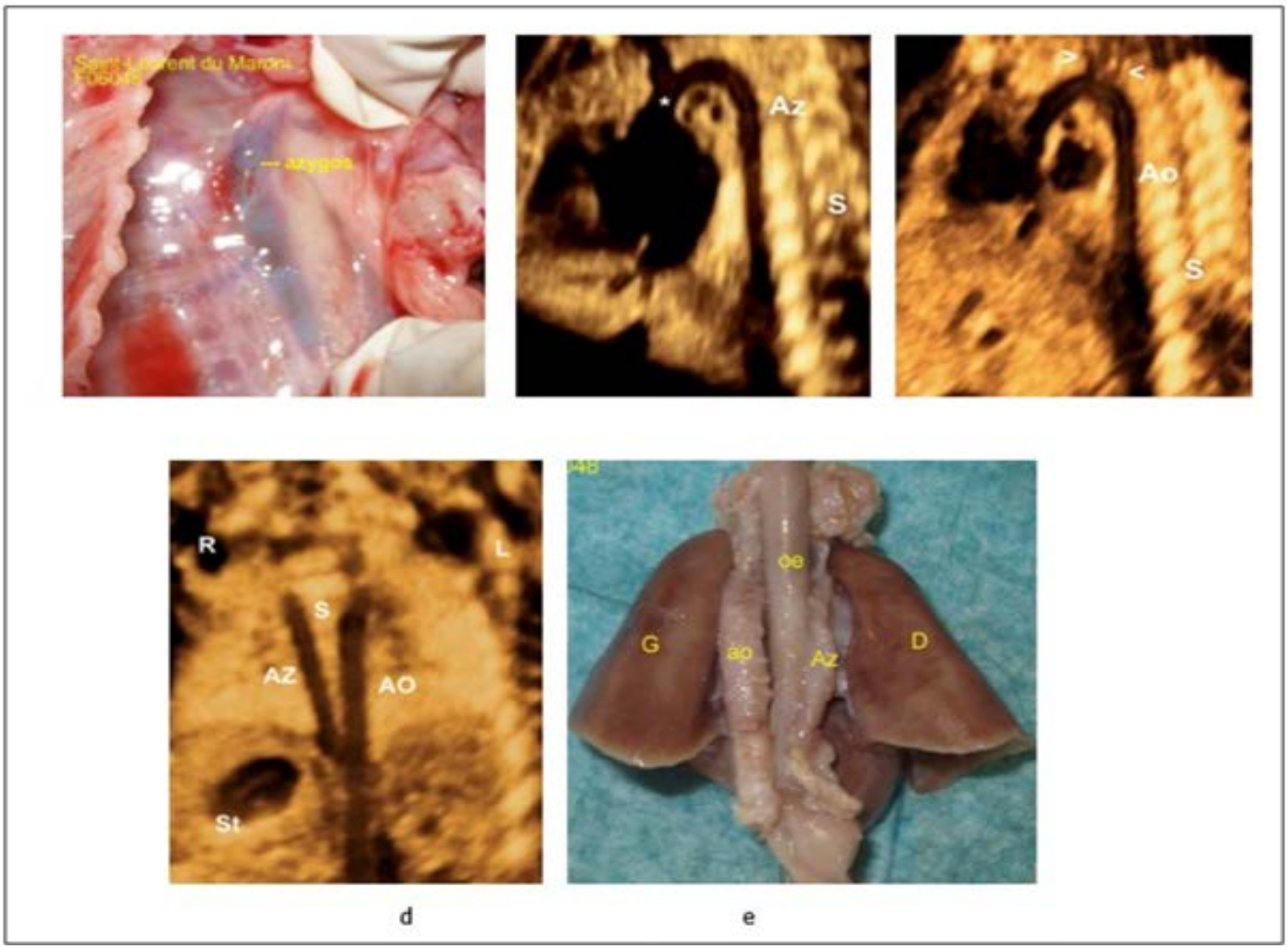

Figure 2: a-Fetopathological examination showing dilated azygos. b- Sagittal image demonstrating the dilated azygos vein and azygos arch connecting to the superior vena cava (*). c-Aortic arche in the same patient, the aortic arche is distinguished from the its branches (arrow heads).e-Coronal planes of the chest and abdomen in a fetus with left atrial isomerism and interrupted inferior vena cava showing the azygos vein (AzV) running parallel and posterior to the descending aorta (on both sides of the spine).f- Fetopathological examination showing azygos and aorta.

congenital duodenal obstruction caused by Ladd's bands. These findings illustrate the diverse anatomical variations observed in cases of left atrial isomerism and underscore the importance of detailed imaging for accurate diagnosis and management [20].

Discussion

The earliest documented case of asplenia with a malformed heart dates back to Martin in 1828. Biron Ivemark's seminal paper in 1955 established the association between spleen anomalies, certain cardiac malformations, and abnormal viscera arrangement, leading to the term "Ivemark's syndrome." Polysplenia was later recognized as a marker of heterotaxy syndrome (HS) by Moller and colleagues in 1967. Van Mierop and colleagues emphasized the correlation between asplenia and bilateral right-sidedness, while Putschar and Mannion described a "symmetrical situs" in historical literature [21].

Van Praagh and others refined the concept of atrial isomerism, emphasizing the distinction between right and left atrial appendage isomerism. Left atrial isomerism syndrome is commonly associated with bilateral left-sidedness and additional anomalies such as left azygos continuation of the inferior vena cava, gastrointestinal malrotation, and absence of the gallbladder. An interrupted inferior vena cava (IVC) with azygos continuation is a hallmark of left atrial isomerism, seen in approximately 80% of cases.

Bilateral superior vena cava (SVC) occurs in a significant percentage of left atrial isomerism cases, with persistent left superior vena cava (PLSVC) being diagnosed in some patients. 3D ultrasound and Doppler imaging allow for detailed reconstruction of vascular anatomy, aiding in the diagnosis of persistent left superior vena cava draining into the coronary sinus and right atrium. Left atrial isomerism may also present with multiple spleens and anomalies of midgut derivatives, including nonrotation or incomplete rotation (Table 1) [22-29].

Mismatch between the positions of the fetal stomach and cardiac apex may indicate atrial isomerism, with the stomach often observed on the right side. Parasagittal views using Doppler imaging can reveal abnormalities such as dextrocardia, with the cardiac apex oriented towards the spine. Any deviation from the standard anatomical position warrants further evaluation, highlighting the utility of Doppler angiography and grayscale imaging in diagnosing situs anomalies.

Conclusion

Left atrial isomerism poses diagnostic challenges due to its complex spectrum of findings. Our study introduces a novel approach to diagnosing atrial isomerism based on the examination of the cardiac apex, the left hepatic vein (LHV), and assessment of the inferior vena cava (IVC) in parasagittal view. In a structurally normal heart with levocardia, a normal IVC, and a right-sided stomach, the likelihood of left isomerism is very low. We propose

Table 1: Summary of patients.

	Case 1	Case 2	Case 3	Case 4
Heart position	Dextrocardia	Levocardia	Levocardia	Levocardia
Stomach	Left	Right	Right	Left
IVC	Interruption	Interruption	No	Interruption
Interruption of IVC with azygos continuation	Yes	Yes	Yes	Yes
Cardiac malformations	AVC- Single atrium (SA) Anomalous hepatic venous drainage directly into the right atrium	LPSVC	AVC Aortic coarc-tation LPSVC	Aortic arche hypoplasia Single ventricle
Digestive anomalies	Midline liver Polysplenia. Intestinal malrotation Appendix on the right side	Midline liver	Midline liver Intestinal nonalrotation	Midline liver duodenal obstruc-tion from Ladd bands,
Caryotype	46XX	46XY	46XY	46XX
Outcome	Pregnancy termination	Delivery at 27+6 weeks	Pregnancy Termination	Pregnancy Termination

that the parasagittal view, which showcases the heart and abdominal vessels, offers a realistic anatomical image without the need for mental reconstruction of spatial relationships. This view is particularly beneficial for detecting situs anomalies and provides an additional tool for diagnosing other anomalies. We recommend incorporating the parasagittal view, along with 3D ultrasound and Doppler angiography, systematically in suspected cases of left atrial isomerism to better understand and interpret fetal anatomy.

References

1. Liang K V, Sanderson S O, Nowakowski G S, Arora A S. (2006) Metastatic malignant melanoma of the gastrointestinal tract. Mayo Clin Proc. 81(4), 511-6.
2. Simons M, Ferreira J, Meunier R, Moss S. (2016) Primary versus Metastatic Gastrointestinal Melanoma: A Rare Case and Review of Current Literature. Case Rep Gastrointest Med. 2306180.
3. El-Sourani N, Troja A, Raab H R, Antolovic D. (2014) Gastric Metastasis of Malignant Melanoma: Report of a Case and Review of Available Literature. Viszeralmedizin. 30(4), 273-5.
4. Wong K, Serafi S W, Bhatia A S, Ibarra I, Allen E A.Melanoma with gastric metastases. , J Community Hosp Intern MedPerspect 6(4), 31972.
5. Augustyn A, de Leon ED, Yopp A C. (2015) Primary gastric melanoma: case report of a rare malignancy. RareTumors. 7(1), 5683.
6. Genova P, Sorce M, Cabibi D, Genova G, Gebbia V et al. (2017) Gastric and Rectal Metastases from Malignant Melanoma Presenting with Hypochromic Anemia and Treated with Immunotherapy. Case Rep Oncol Med. 2079068.
7. Alghisi F, Crispino P, Cocco A, Richetta A G, Nardi F et al. (2008) Morphologically and immunohistochemically undifferentiated gastric neoplasia in a patient with multiple metastatic malignant melanomas: a case report. J Med Case Rep. 2, 134.