Understanding and managing dextrocardia with *situs inversus*: A case report

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Abstract. Dextrocardia with *situs inversus* is an uncommon congenital disorder in which the heart and other organs in the chest and abdomen are reversed. Although it rarely causes symptoms, it can be linked to heart or systemic disorders and is challenging to diagnose due to its unique architecture. The present report describes the case of a 27-year-old man who presented with sudden breathing problems. A physical examination revealed no heartbeat on the left side and odd cardiac noises. Further electrocardiogram tests revealed classic symptoms of dextrocardia, including right axis deviation and inverted P and T waves in Lead 1. Imaging scans confirmed the diagnosis of dextrocardia with *situs inversus totalis*, which placed the liver and gallbladder on the left and the spleen on the right. Despite these uncommon structural abnormalities, the patient's heart functioned normally, as shown by echocardiography. The patient was provided with treatment (expectorant and mucolytics) for the breathing symptoms and directed to cardiologists and pulmonologists. The present case emphasizes the significance of conducting a thorough clinical and diagnostic investigation when encountering odd results. The report highlights the importance of practitioners being aware of unusual illnesses, such as *situs inversus*, to prevent misdiagnoses and unnecessary operations. The case is notable for the absence of structural cardiac damage despite a complete reversal of the internal architecture.

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Introduction

Dextrocardia with *situs inversus*, also known as *situs inversus totalis*, is a rare congenital condition where the major visceral organs are mirrored from their normal positions. The heart is located on the right side of the thoracic cavity instead of the left, and other organs, such as the liver, spleen, stomach and intestines, are also reversed. This condition occurs in ~1 in 10,000 live births (1). The exact cause of dextrocardia with *situs inversus* is not fully understood; however, it is believed to involve a combination of genetic and environmental factors. *Situs inversus* is often inherited in an autosomal recessive manner, meaning both parents must carry a copy of the mutated gene to pass it on to their child (2). Specific genes associated with laterality defects, such as zinc finger protein of the cerebellum 3, located on the X chromosome (Xq26); it encodes a transcription factor critical for early embryonic patterning and regulates signaling pathways (notably NODAL pathway) that establish the left-right body axis. Dynein axonemal heavy chain 5 (DNAH5) encodes a dynein arm motor protein in the outer dynein arm of motile cilia, and DNAH11 is another outer dynein arm heavy chain essential for effective ciliary beating, have been implicated in the development of this condition (3). During normal embryonic development, the organs undergo a complex process of rotation and positioning. In individuals with *situs inversus*, this process is altered, leading to the mirror-image positioning of the organs. This reversal happens early in embryogenesis, typically within the first few weeks of gestation. People with dextrocardia with *situs inversus* usually do not have any symptoms directly related to the positioning of their organs and can lead normal, healthy lives (1). However, they may be at an increased risk for certain complications, such as Kartagener syndrome (OMIM: 244400). Approximately 25% of individuals with *situs inversus* have Kartagener syndrome, which includes chronic *sinusitis*, bronchiectasis and infertility due to ciliary dysfunction. While most individuals with *situs inversus* have a structurally normal heart, there is an

increased incidence of congenital heart defects, such as transposition of the great arteries, atrial septal defects (OMIM: 600747) or ventricular septal defects (OMIM: 600996), and tetralogy of Fallot (4). Dextrocardia with *situs inversus* is typically diagnosed through imaging studies, including chest X-ray, which reveals the heart's position on the right side of the chest, echocardiogram, which provides detailed images of the heart's structure and function, and abdominal ultrasound or CT scans, which confirm the reversed positioning of the abdominal organs. Management of dextrocardia with *situs inversus* focuses on monitoring for and addressing any associated conditions or complications. Regular follow-up with a healthcare provider is essential to ensure any potential issues are identified and treated promptly. The prognosis for individuals with dextrocardia with *situs inversus* is generally good, especially in the absence of significant congenital heart defects or other associated syndromes (5). With appropriate medical care, most individuals can expect to live a normal lifespan.

Case report

Presentation. A 27-year-old man presented to the Akash Institute of Medical Sciences and Research Centre (Bengaluru, India) in May 2023 with a history of a cough with expectoration and fever with chills that had persisted for 4 days. The patient had no known comorbidities, no notable family history of congenital disorders and was born of a non-consanguineous marriage. The patient reported no history of smoking or alcohol consumption.

On examination, the patient's vital signs were as follows: Pulse, 110 bpm (tachycardia); blood pressure, 122/74 mmHg; respiratory rate, 20 cycles per min; SpO_2, 96%; and temperature, 99.8˚F. The apex beat was not palpable on the left side and only faint heart sounds were heard in the left hemithorax. Percussion revealed a tympanic note over the right thorax. On abdominal examination, liver dullness was detected on the left side, suggesting visceral transposition (Table I).

Investigations. An electrocardiogram revealed sinus rhythm with right axis deviation, a negative QRS complex in Lead 1, and inverted P and T waves in Lead 1, with a progressively decreasing R-wave amplitude from V1 to V6. A chest X-ray demonstrated a right-sided cardiac shadow consistent with dextrocardia (Fig. 1). An electrocardiogram revealed sinus rhythm with right axis deviation, a negative QRS complex in Lead 1, and inverted P and T waves in Lead 1, with a progressively decreasing R-wave amplitude from V1 to V6 (Fig. 2). Ultrasonography showed the spleen on the right (Fig. 3) (splenomegaly 14.5 cm), with normal kidneys, consistent with *situs inversus totalis*.

Baseline laboratory investigations were also performed. Hematology showed a white blood cell count of 4,910/mm^3 (normal 4,000-10,500/mm^3), a red blood cell count of 4.29 million/mm^3 (normal range, 4.5-5.9 million/mm^3) hemoglobin 12.5 g/dl (normal range, 13.5-17.5 g/dl), a packed cell volume of 38.2% (normal range, 41-53%) and a platelet count of 1.64 lakh/mm^3 (normal range, 150-400 lakh/mm^3). The differential count revealed 65% neutrophils (normal range, 40-70%), 31% lymphocytes (normal range, 20-40%) and 2% monocytes (normal range, 2-8%), with absent eosinophils (normal range, 1-4%) and basophils (normal range, 0-1%). Biochemical tests showed normal renal and liver function, with 0.9 mg/dl serum creatinine (normal range, 0.7-1.3 mg/dl), 9.4 mg/dl urea (normal range, 20-40 mg/dl) and 3.2 mg/dl uric acid (normal range, 3.5-7.2 mg/dl). Inflammatory markers, including erythrocyte sedimentation rate and C-reactive protein, were within normal limits, and troponin-I was negative, ruling out myocardial ischemia.

Serological screening for infectious diseases was negative, including hepatitis B surface antigen, human immunodeficiency virus 1/2, hepatitis C virus and dengue virus [non-structural protein 1 antigen, immunoglobulin (Ig)M and IgG antibodies]. Urine analysis was normal, with no evidence of proteinuria, hematuria or infection.

These electrocardiogram (ECG) findings are characteristic of dextrocardia with *situs inversus*, which helps in diagnosing the condition. The distinct ECG findings, combined with clinical examination and imaging studies, allowed for a comprehensive understanding of the patient's unique anatomical and physiological presentation.

Outcome and follow-up. The patient was managed with symptomatic treatment, including paracetamol for fever and expectorants for the cough. Paracetamol was provided at 500 mg every 6 h for 3 days and bromhexine was provided at 8 mg three times a day for 3 days. No specific cardiac therapy was required, as echocardiography confirmed normal cardiac function. The patient was referred to the Departments of Cardiology and Pulmonology for further evaluation and follow-up. The respiratory symptoms of the patient improved during the hospital stay, and was discharged in stable condition. At the 3-month follow-up in August 2023, the patient remained asymptomatic and repeat echocardiography showed normal findings. No features were present to suggest Kartagener syndrome, and genetic testing was not pursued. The patient was educated about the implications of *situs inversus* for emergency and surgical care, and was advised to wear a medical alert bracelet.

Discussion

The present case emphasizes the importance of combining physical examination with electrocardiographic and imaging findings when diagnosing dextrocardia with *situs inversus*, especially in patients who present with non-specific symptoms. The present patient, a young adult with no known comorbidities, showed normal cardiac anatomy and function despite a complete mirror-image arrangement of the thoracoabdominal organs. The present case highlights the significance of recognizing unique ECG abnormalities that could otherwise lead to misinterpretation and unnecessary testing. Such clinical vigilance is vital to prevent diagnostic errors and overtreatment.

In the present study, differential diagnoses considered at presentation included pneumonia, bronchitis, tuberculosis, congenital heart disease, pericarditis, gastroesophageal reflux disease and cholecystitis. These were excluded based on normal inflammatory markers, negative serology and supportive imaging studies, which confirmed *situs inversus totalis* with dextrocardia and no structural cardiac defects.

Table I. Clinical and diagnostic summary of the patient with dextrocardia and *situs inversus*.

Parameter	Findings
Age, years	27
Sex	Male
Date of presentation	May 2023
Place of presentation	Akash Institute of Medical Sciences and Research Centre (Bengaluru, India)
Chief complaints	Cough with expectoration, fever with chills (4 days duration)
Habits	Non-smoker and non-drinker
Comorbidities	None
Vital signs	Pulse, 110 bpm (tachycardia); blood pressure, 122/74 mmHg; respiratory rate, 20 cpm; SpO_2, 96%; temperature, 99.8˚F
Cardiovascular exam	Apex beat not palpable on left; faint heart sounds on left
Respiratory exam	Tympanic percussion note on the right thorax
Abdominal exam	Liver dullness on the left side
ECG findings	Sinus rhythm; right axis deviation; negative QRS in Lead 1; inverted P and T waves in Lead 1; decreasing R wave amplitude from V1 to V6
Imaging investigations	Chest X-ray: Dextrocardia suspected. 2D echocardiography: Dextrocardia with normal cardiac function. Abdominal US: Liver and gallbladder on left, spleen on right
Provisional diagnosis	Dextrocardia with *situs inversus*
Laboratory tests conducted at	Akash Institute of Medical Sciences and Research Centre, (Bengaluru, India)
Management	Symptomatic treatment (for fever and coughing), referred to a cardiologist and pulmonologist

ECG, electrocardiogram; US, ultrasonography.

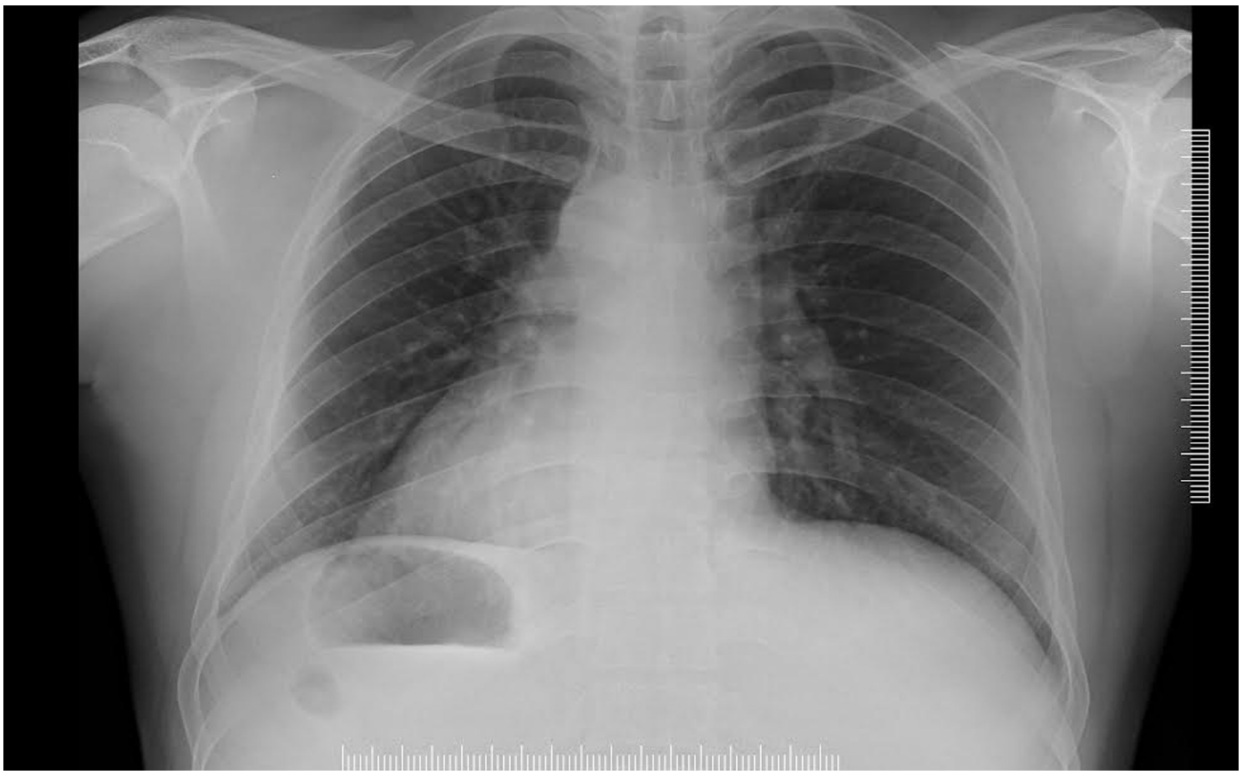

Figure 1. Chest X-ray demonstrating a right-sided cardiac shadow consistent with dextrocardia.

Dextrocardia with *situs inversus* presents a unique diagnostic challenge due to its frequent association with congenital cardiac anomalies. When dextrocardia is associated with *situs solitus*, the incidence of notable cardiac defects, such as atrial septal defects, ventricular septal defects and the transposition of the great vessels, is high, reaching 90-95% (6-8). By contrast, patients with

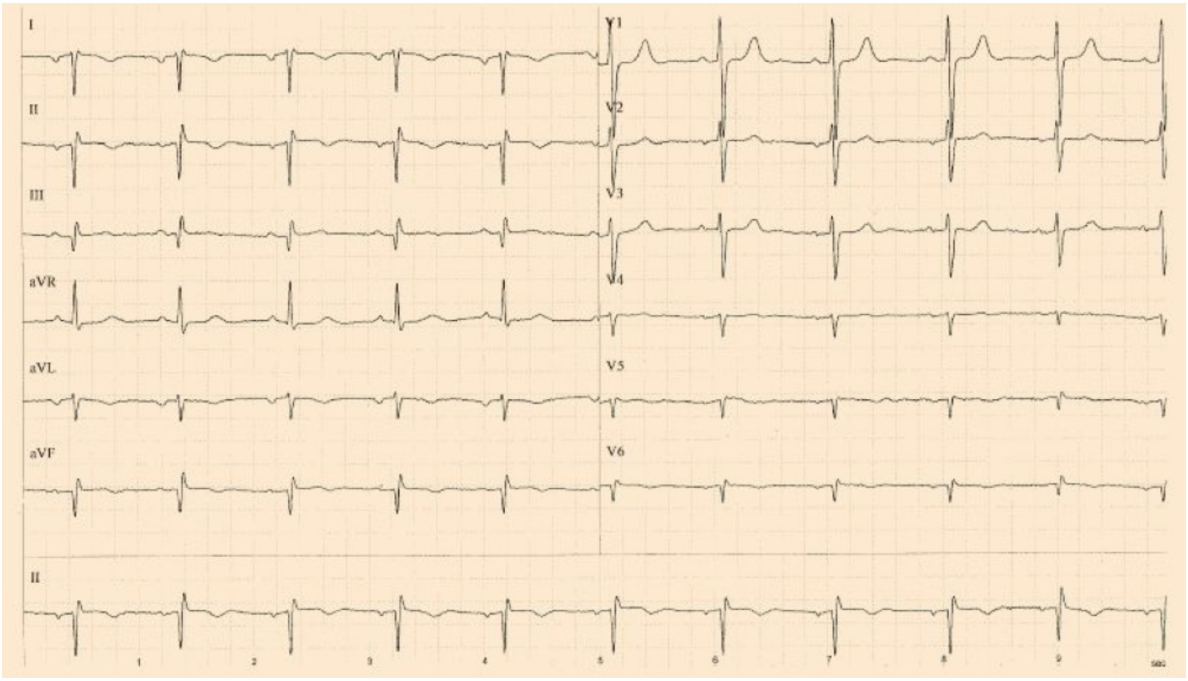

Figure 2. Two-dimensional electrocardiography report of the patient. I, II and III represent the bipolar ECG leads and aVR, aVL and aVF represent the augmented unipolar ECG limb leads. ECG, electrocardiogram.

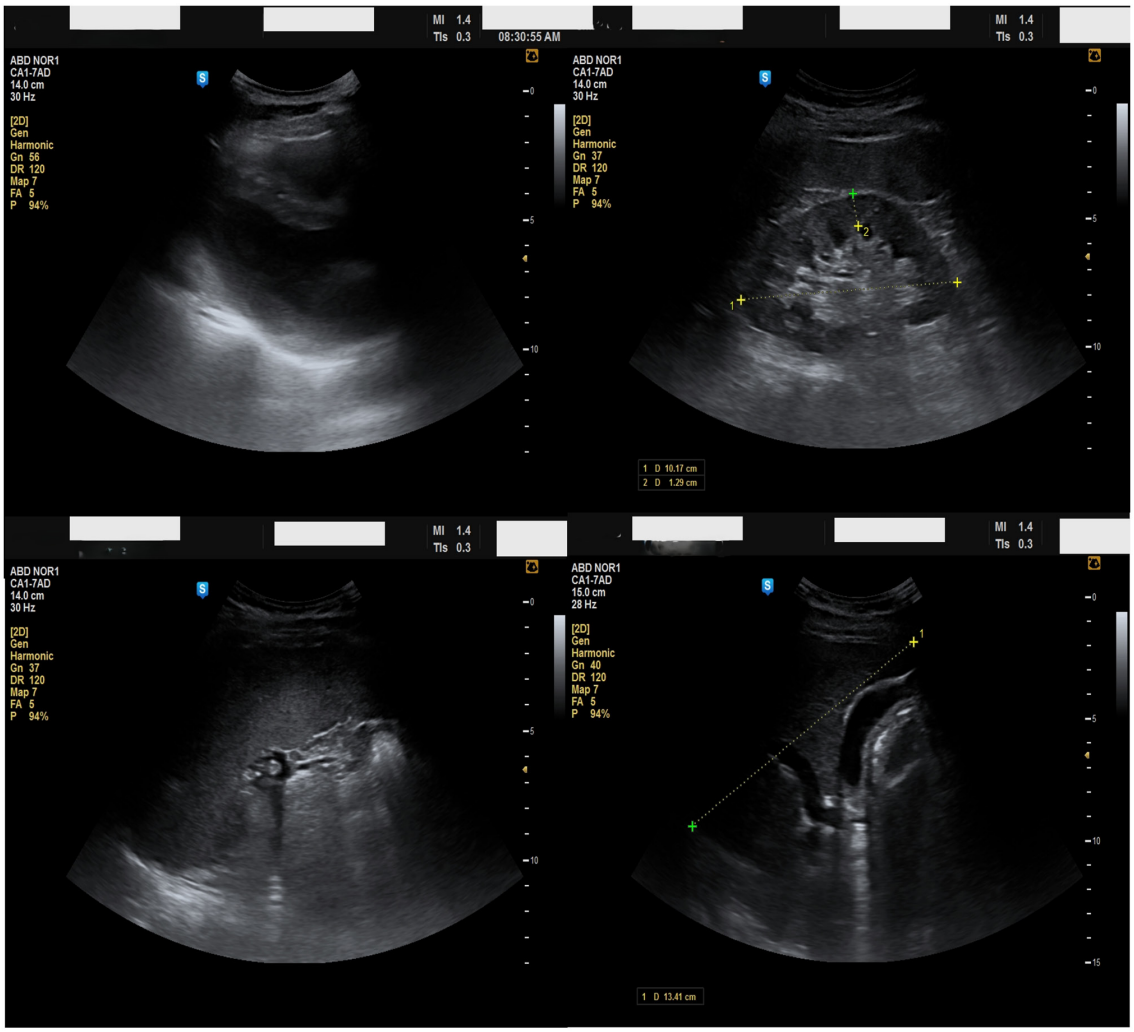

Figure 3. Abdominal ultrasonography showing the spleen in the right hypochondrium.

situs inversus totalis, as in the present case, often have structurally normal hearts, although other anomalies may still be present.

Associated defects in *situs inversus* may go beyond the cardiovascular system and include duodenal atresia (OMIM: 223400), asplenism (OMIM: 271400) or polysplenia, ectopic or horseshoe kidneys and various pulmonary abnormalities, such as chronic sinopulmonary infections and bronchiectasis (8). The presence of *situs inversus* along with primary ciliary dyskinesia (OMIM: 613807) leads to Kartagener syndrome, which is characterized by chronic sinusitis, bronchiectasis and infertility caused by impaired ciliary function (9,10).

The cause of laterality defects such as dextrocardia with *situs inversus* remains multifactorial. Genetic factors, including mutations in the DNAH5 gene, are essential in disrupting left-right asymmetry during embryogenesis (11). Environmental influences such as maternal diabetes, teratogenic exposure and rare cases of conjoined twinning have also been linked (11,12).

Management requires a multidisciplinary approach tailored to the patient's specific anatomical and functional anomalies. A comprehensive cardiac evaluation using echocardiography and ECG is essential for detecting structural or electrical abnormalities (1,2). Respiratory assessments and physiotherapy become particularly important in patients with suspected or confirmed Kartagener syndrome (9,10).

When surgical intervention is needed, careful attention should be paid to reversed organ orientation to prevent intraoperative errors. Genetic counseling should be provided to affected individuals and their families, especially those planning future pregnancies, to understand hereditary risks and implications (11-13). Advances in diagnostic imaging and genetic testing have greatly improved early detection and enabled personalized treatment strategies.

The present case report highlights the educational importance of recognizing such rare congenital conditions and stresses the need for a high index of suspicion in atypical clinical cases. Early detection, proper referral and structured follow-up are essential for improving outcomes in patients with dextrocardia and *situs inversus*.

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Availability of data and materials

The data generated in the present study may be requested from the corresponding author.

Authors' contributions

ND contributed to the conception and design of the study, participated in data collection, analysis and interpretation, conducted the literature review, and drafted the manuscript. MHS contributed to data collection, as well as the analysis and interpretation of the patient data. SCMR contributed to the conception and design of the study, participated in the interpretation of clinical findings, and critically revised the manuscript for important intellectual content, providing substantial scientific input that shaped the discussion and clinical interpretation. KHLG contributed to the interpretation of patient data and clinical decision-making aspects of the case, and critically revised the manuscript for important intellectual content with significant academic and clinical input. ND and KHLG confirm the authenticity of all the raw data. All authors have read and approved the final manuscript and agree to be accountable for all aspects of the work.

Ethics approval and consent to participate

Not applicable.

Patient consent for publication

The patient consented to the publication of the case report and the associated images.

Competing interests

The authors declare that they have no competing interests.

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