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Heterotaxy syndrome, dextrocardia, ureteropelvic obstruction, endometriosis, and pulmonary hypertension in an adult with congenital heart defects: a case report

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Abstract

Background Heterotaxia is characterized by an abnormal positioning of the thoracic and/or abdominal organs, resulting in various physiological and hemodynamic implications. Congenital heart disease involves structural irregularities in the heart or major vessels within the chest, leading to functional challenges.

Case presentation We present a 26-year-old Arab female patient with a complex medical history involving heterotaxy, dextrocardia, congenital heart disease, and ureteropelvic junction obstruction diagnosed in her first year of life, followed by the identification of endometriosis in her early twenties. This combination of disorders is reportedly unique in existing literature. The patient sought emergency care for severe diffuse abdominal pain unrelated to diet or bowel movements, accompanied by nausea. The pain was localized in specific areas and radiated to the shoulder. Despite treatment attempts with hormonal therapy for abdominal pain relief, her symptoms persisted. Upon examination, no immediate surgical or medical emergencies were noted, leading to symptom management with antispasmodics and recommendations for specialist follow-ups. The patient's cardiac condition, including cyanosis and related complications, was discussed along with her past surgeries and diagnostic findings related to her cardiovascular health.

Conclusion This case emphasizes the challenges of managing complex medical conditions in a young individual, highlighting the need for multidisciplinary care and long-term follow-up to ensure the best possible quality of life for the patient. An interdisciplinary approach is crucial for the comprehensive care of patients with heterotaxy, as they may present with a wide range of anatomical abnormalities affecting different organ systems. Clinicians need to be well informed about the complexities of heterotaxy syndromes and their diverse manifestations to provide effective and personalized care.

Keywords Heterotaxy, Dextrocardia, Congenital heart disease, Ureteropelvic junction obstruction, Endometriosis

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Background

Heterotaxia is characterized by an abnormal positioning of the thoracic and/or abdominal organs, resulting in various physiological and hemodynamic implications. It differs from situs inversus, although it may not align entirely with that condition. It is observed in approximately 1 in 10,000 births and is often linked to congenital heart defects, with a co-occurrence rate as

high as 100%. Common heart anomalies linked to heterotaxy include atrial septal defects, partial anomalous pulmonary venous return, and common atrioventricular canals. Individuals with heterotaxy typically exhibit a disruption of the inferior vena cava, which may be replaced by the azygous or hemiazygous vein [1, 2].

Congenital heart disease (CHD) involves structural irregularities in the heart or major vessels within the chest, leading to functional challenges. Between 22% and 45% of patients with CHD also have accompanying extracardiac anomalies or genetic disorders. Notably, the incidence of renal and urinary tract abnormalities in individuals with CHD ranges from 7.5% to 12.5%, surpassing that in the general population. Strikingly, there is no apparent correlation between specific cardiac defects and renal or urinary tract abnormalities [3]. In contrast, pulmonary hypertension is an uncommon occurrence in individuals with heterotaxy syndrome [4].

Endometriosis is a common issue that impacts women in their childbearing years. Data suggest that it affects 2–10% of women in general and might impact as many as half of women dealing with infertility. It is a noncancerous, estrogen-sensitive, and ongoing inflammatory condition distinguished by the existence of endometrial tissue found outside the uterus. Typical symptoms include painful menstruation, discomfort during intercourse, persistent pelvic pain, and challenges with fertility [5].

The precise origins of endometriosis remain unclear, but it is believed that a combination of genetic elements and environmental influences, such as exposure to toxins and pollutants, play a role in its onset [6].

Ureteropelvic junction (UPJ) obstruction is a frequent type of kidney blockage that occurs when there is a barrier at the point where the renal pelvis meets the start of the ureter. This obstruction can arise from either inherent factors present since birth or conditions acquired later in life. Most cases are congenital, meaning they are present from birth, and can be triggered by internal or external factors. UPJ obstruction usually presents in individuals either incidentally during an ultrasound or when an individual complains of flank pain [7, 8].

In this report, we present the case of a 26-year-old Arab female patient with significant medical history of heterotaxy, dextrocardia, and ureteropelvic junction obstruction, all of which were diagnosed in her first year of life. Additionally, in her early twenties, she was diagnosed with endometriosis. To the best of our knowledge, this is the first case in literature with this combination of disorders.

Case presentation

A 26-year-old Arab female patient presented to the emergency room owing to severe, vague abdominal pain, which was generalized. This pain was dull and did not correlate with food intake or bowel movements. There were no associated symptoms other than nausea; it was sometimes located in the epigastric and right chondral areas and sometimes spread to the right shoulder without spreading to the back or the left shoulder. Therefore, she was treated with antispasmodics and told to follow up with a cardiologist and a gynecologist.

During her cardiological follow-up, the patient stated that her life is nearly normal with oxygen supplementation; she goes to college, walks, and dances. Her dyspnea is classified as New York Heart Association (NYHA) class 2, with no orthopnea or paroxysmal nocturnal dyspnea. At one point, she became cyanotic, with a saturation of 65%, during minimal activities such as climbing stairs, and it may have declined to 45%.

Her medical history is significant for congenital heart disease, endometriosis with repeated episodes of abdominal pain, and unrepaired unilateral ureteropelvic junction obstruction. Her family history was not significant. No maternal history of smoking or drug use (other than amoxicillin/clavulanic acid and over-the-counter mucolytics) during pregnancy. However, her mother had a history of mild viral upper respiratory tract infection during the first trimester. Moreover, there was no maternal history of stillbirth, but there was a history of a spontaneous abortion in the second trimester.

The patient did not have a cyanotic appearance at birth but became cyanotic at the age of 2 months, which was more prominent during breastfeeding and crying. Unfortunately, no reported investigations support first-time diagnoses.

At the age of 2 years, the patient was hospitalized in preparation for pulmonary artery banding, a surgical intervention to reduce overcirculation in the pulmonary tree. Unfortunately, it was canceled for unknown reasons. At the time, the parents were told about their daughter's poor prognosis and were advised to monitor their daughter's health with digitalis treatment.

When she was 3 years old, the patient was diagnosed with left ureteropelvic junction obstruction. Owing to cardiac disease, her surgery was canceled.

At 8 years of age, echocardiography revealed dextrocardia with atrioventricular concordance and ventriculoarterial disconcordance. Additionally, it showed that the inferior vena cava was continuous with the azygos vein, the presence of an atrial septal defect (ASD), a ventricular septal defect (VSD), mitral valve atresia, right pulmonary artery aneurysm, and signs of pulmonary hypertension.

At the age of 18 years, she was reportedly diagnosed with dextrocardia, hypoplastic left heart syndrome, ASD, VSD, Eisenmenger syndrome, and situs inversus totalis.

An echocardiography was performed showing the following: dextrocardia with mirror image heart; left superior vena cava and hepatic veins draining into the left-sided atrium; interrupted inferior vena cava that was continuous with the hemi-azygos vein; pulmonary veins draining to the right-sided atrium; mitral valve atresia with a single left AV valve (tricuspid valve); a hypoplastic left ventricle located posteriorly and to the left; a dominant right ventricle which was significantly dilated, heavily trabeculated, and located anteriorly and to the right; large muscular VSD that measured 10 mm, with a bidirectional shunt; moderate to severe depression of ventricular systolic function in the right ventricle (RV); both atria were dilated; a large ASD that measured 26 mm with a bidirectional shunt; big vessels side-by-side (the aorta to the left arising from the dominant RV and the pulmonary artery to the right arising from the hypoplastic left ventricle (LV)); posterior deviation of the conus septum causing slight sub-pulmonary obstruction; significantly enlarged pulmonary arteries especially on the right side (aneurysmal dilatation); the main pulmonary artery (MPA) had a diameter of 50 mm, the left pulmonary artery (LPA) had a 26 diameter of , and the right pulmonary artery (RPA) had a diameter of 26 mm; the right aortic arch with no evidence of coarctation or patent ductus arteriosus (PDA); the coronary artery origins were normal. The color Doppler study showed mild pulmonary regurgitation (PR) with elevated pulmonary end-diastolic pressure (PEDP) (PEDP = 57 mmHg and mean pulmonary artery pressure (MPAP) = 88 mmHg) and mild left AV valve incompetence. The continuous wave (CW)/pulsed wave (PW) Doppler studies demonstrated indirect evidence of severe pulmonary hypertension (Eisenmenger), flow velocity across the sub-pulmonary and pulmonary valve = 2.3 m/second, and a peak instantaneous pressure gradient (PIPG) = 21 mmHg. Flow velocity across the pulmonary artery was 1.6 m/second with mid-systolic closure and flow velocity across the aortic valve was 1.2 m/second.

At the time, the patient was managed with digoxin, furosemide, sildenafil, and aspirin.

At 22 years old, she was diagnosed with endometriosis.

Her cardiovascular examination revealed the following: a centric cyanotic appearance with severe clubbing in her fingers and toes; no distended jugular veins; no peripheral edema; no pulmonary edema; apical pulse was palpable at the right fifth intercostal space at the anterior axillary line; heart auscultation revealed holosystolic murmur with 3/6 intensity best heard at the apex with no spreading. Her abdomen was soft, nontender, with normal bowel sounds. The liver was not palpable or percussed at either side, and there was no palpable spleen. Her vitals included oxygen saturation of 85% and 80% in her upper and lower extremities, respectively; blood pressure of 90/60 mmHg in both arms; heart rate 72 beats/minute and irregular; and no fever.

A typical electrocardiogram (ECG) was performed (Figs. 1, 2). It showed sinus rhythm with left axis deviation and a regular rhythm at a speed of 74 beats/minute, some premature atrial contractions, signs of left atrial enlargement, signs of complete right bundle branch block, and signs of biventricular hypertrophy.

The echocardiography was performed at her follow-up in 2024 revealing the exact malformations but with slightly different numbers (Figs. 3, 4).

A chest–abdomen–pelvis computed tomography (CT) scan with contrast was performed, revealing a bilobed lung with hyparterial bronchus bilaterally; dextrocardia with malformations; suprahepatic veins draining through one vein into the right atrium, and it was noted that the right inferior pulmonary vein was also draining through this vein; left lobe of liver extending to the left epigastric area forming "beaver tail liver" around the spleen; bilobed spleen with a notch in its central portion; extrarenal pelvis on the right side; anteriorly rotated pelvis with obstruction of the ureteropelvic junction and second-degree hydronephrosis on the left side; and hematosalpinx bilaterally. These anomalies gave the impression of situs ambiguous with a left heterotaxy pattern, with dextrocardia and other congenital malformations (including partial anomalous pulmonary venous return (PAPVR)), UPJ obstruction, and endometriosis (Figs. 5, 6 and 7).

In addition to the CT scan, an abdominal ultrasound was done for further follow-up. It revealed the presence of a cyst with turbid content, measuring 3.6 cm by 5.5 cm on the right Fallopian tube and a similar cyst measuring 6.5 cm by 7.3 cm on the left tube, more commonly associated with endometriosis (Fig. 8).

After a couple of weeks, a gynecologic consultation was conducted to manage the patient's abdominal pain. The advice was to handle the pain with only over-the-counter analgesics to avoid the side effects of hormonal therapy or medical oophorectomy at this age. To avoid direct trauma to the abdomen and pelvis regions as much as possible, the doctor stated that surgical options were not considered owing to her cardiac status.

The patient was advised to monitor her general health, especially mouth hygiene and wound care. In addition, it was highly recommended to avoid pregnancy, and there is a possibility of a prescription for birth control pills. She was advised to get infective endocarditis prophylaxis before dental procedures. She is managed with furosemide, digoxin, aspirin,

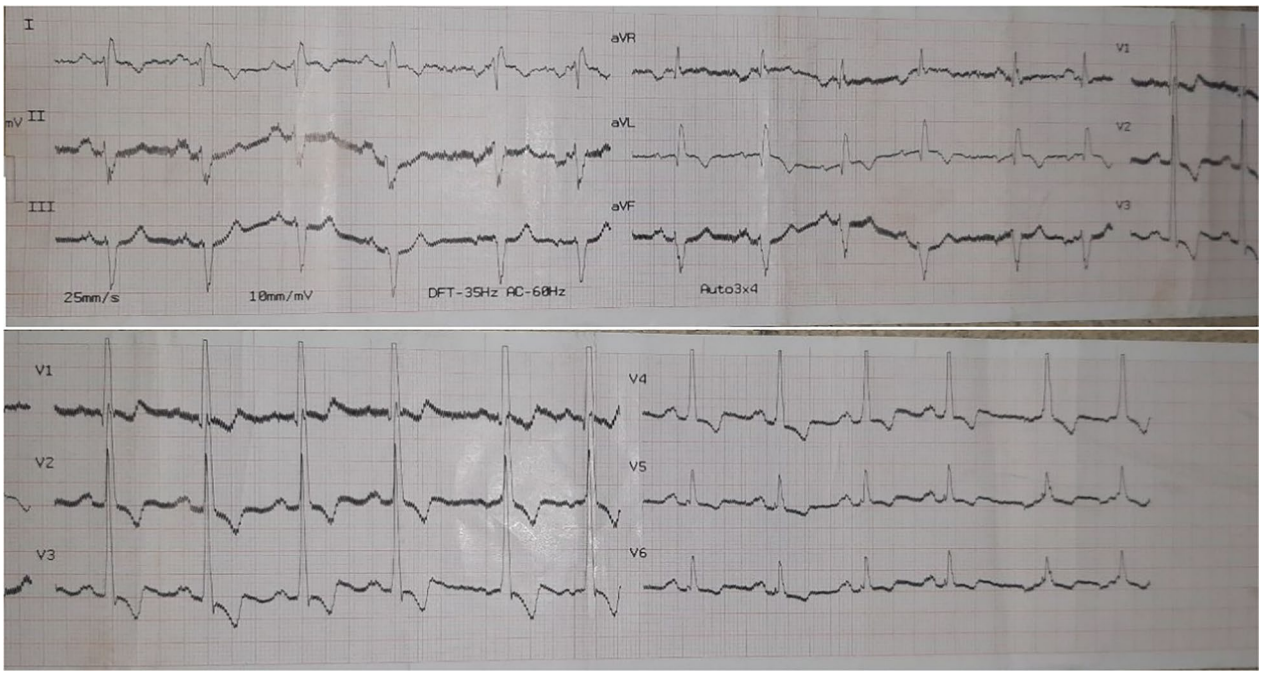

Fig. 1 Electrocardiogram showing a sinus rhythm with left axis deviation and a regular rhythm at a speed of 74 beats/minute, some premature atrial contractions, left atrial enlargement, complete right bundle branch block, and biventricular hypertrophy

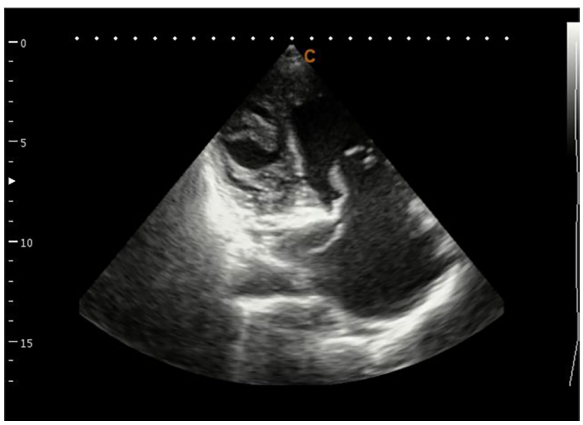

Fig. 2 Echocardiogram demonstrating the following findings: atrial septal defect, ventricular septal defect, hypertrophic right ventricle, and hypoplastic left ventricle

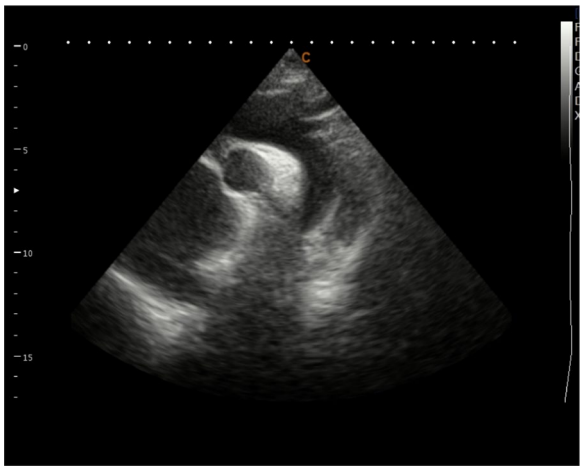

Fig. 3 Echocardiogram demonstrating the aortic arch with normal appearance and no sign of patent ductus arteriosus

sildenafil, and analgesics to relieve her abdominal pain, and was told to follow-up every 6 months or when her symptoms worsen.

The prognosis of the patient is poor, with high pulmonary arterial pressure and lack of ability to do complex interventions such as heart and lung transplantation. Additionally, pregnancy is contraindicated and, without emergent needs, noncardiac surgery is highly dangerous.

Discussion and conclusion

In literature, the combination of the abnormalities presented in this case (heterotaxy syndrome: mirror-image dextrocardia, PHTN, hemiazygous continuation of the interrupted inferior vena cava, bilobed lungs, bilobed spleen, PAPVR, extrarenal pelvis, and ureteropelvic junction obstruction (UPJO)) has never been described. Therefore, the presented case illustrates a complex and

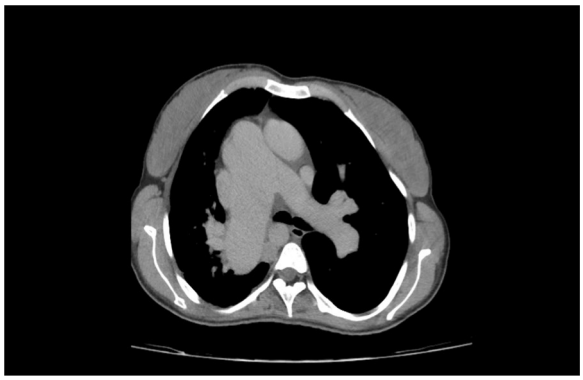

Fig. 4 An axial view of a computed tomography scan showing the pulmonary arteries and right pulmonary artery aneurysm

unique anatomy that will offer valuable insights for the future.

Situs ambiguous, or heterotaxy syndrome, refers to an abnormal positioning of internal organs compared with that of normal organs. There have been no specific risk factors identified, and it is thought that the underlying etiology of these abnormalities is a primary defect in lateralization around day 28 of gestation, which leads to a deviation from the normal position of the viscera. Such arrangements are thought to be multifactorial in their inheritance pattern, and the incidence of aneuploidy or other chromosomal abnormalities is very low. The incidence of heterotaxy syndrome is 1:10,000 newborns, but heterotaxy syndrome accounts for approximately 4% of all congenital heart disease (CHD) cases. Situs ambiguous is traditionally divided into two general categories: left isomerism (situs ambiguous with polysplenia) and right isomerism (situs ambiguous with asplenia).

Situs ambiguous with polysplenia, has an incidence of between 1:10,000 and 1:20,000 in new-born births and a female predominance. CHD often presents to medical attention owing to a suspected heart defect, as the incidence of CHD is between 50% and 90%. Commonly associated cardiac malformations include partial anomalous pulmonary venous return, atrial septal defect (ASD), and a common atrioventricular (AV) canal. Although the term polysplenia is used, the number of spleens ranges from one to ten or more; spleens may be located either bilaterally or, if unilateral, ipsilateral to the stomach because the spleen arises from the dorsal mesogastrium. Another common finding is inferior vena cava (IVC) interruption with azygos or hemiazygos continuation; this has been reported to be the most specific finding of situs ambiguous with polysplenia. Consistent with the term "left isomerism," the lungs are bilaterally bilobed with hyparterial bronchi.

Situs ambiguous with asplenia, which has an incidence of 1:10,000 and 1:20,000 in new-born births and a male predominance, has a very high incidence of CHD (nearly 100%) and, thus, often presents in childhood with a cyanotic heart defect such as a common AV canal, univentricular heart, transposition of the great arteries, or total anomalous pulmonary venous return. In contrast to left isomerism, when the spleen is absent, the IVC typically runs ipsilateral to the aorta, and the patient will have bilateral trilobed lungs with eparterial bronchi.

Patients with heterotaxy syndromes present in a number of ways owing to the wide spectrum of anatomic

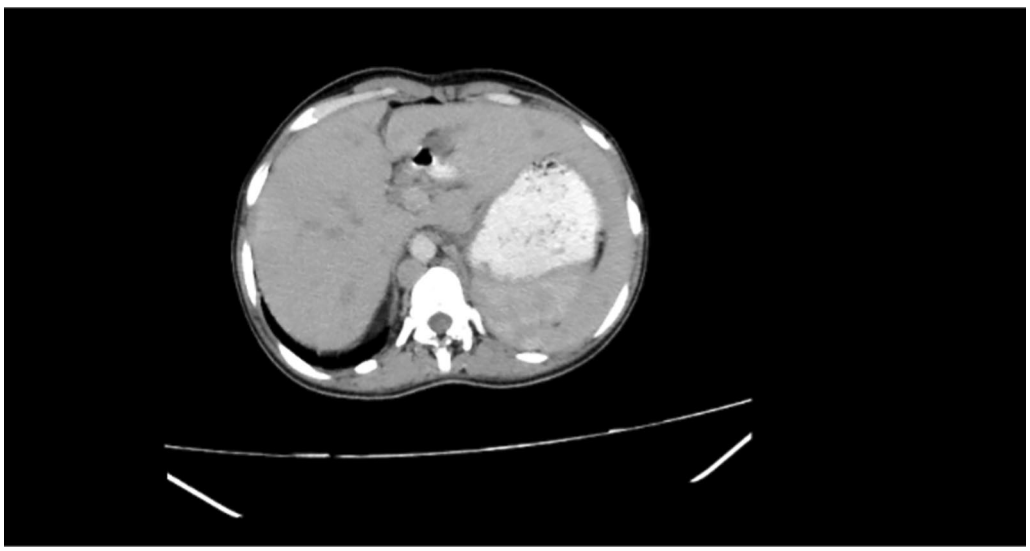

Fig. 5 An axial view of a computed tomography scan showing the liver and spleen, with "beaver tail liver" noted around the spleen

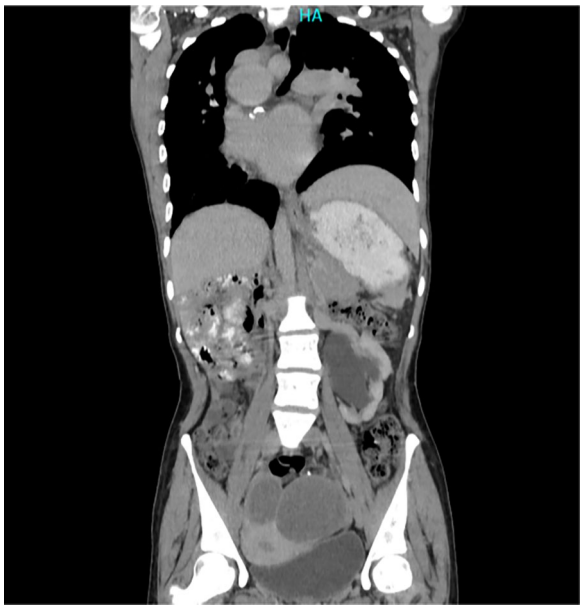

Fig. 6 A coronal view of a computed tomography scan showing the kidneys and uterus, left side hydronephrosis, and sacs around the uterus

abnormalities that may be present. Unlike situs inversus totalis, a more benign arrangement that often occurs in adulthood because of its relatively few associated abnormalities, the anatomy of situs ambiguous does not follow a specific pattern. Because most patients will not fit neatly into one of the two categories outlined above, it is thought that asplenia and polysplenia are on two ends of a spectrum, with many patients falling somewhere in between, and that the specific individual abnormalities should be stated in the diagnosis rather than simply using the terms left isomerism or polysplenia and right-sided isomerism or asplenia [9].

The severity of heterotaxy syndrome ranges from mild to life-threatening, significantly impacting infants and children. The syndrome, especially in cases of polysplenia, leads to a high mortality rate, mainly owing to severe cardiac abnormalities, with a majority of affected children not surviving past age 5 years. Notably, a small percentage of patients with minor cardiac defects may reach adulthood without symptoms. With complex cardiac issues, the 1-year mortality rate is alarmingly high. The scarcity of data on adult patients highlights the challenge of managing these patients, especially regarding surgical interventions and health maintenance. The syndrome also encompasses genitourinary abnormalities but lacks detailed data on sex-specific anomalies and has no strong links to female anatomy problems [10].

This lack of information raises questions about whether patients with heterotaxy syndrome should follow standard screening and health maintenance guidelines. Despite the data gaps, there is an emphasis on the necessity for intensified monitoring and a multidisciplinary approach

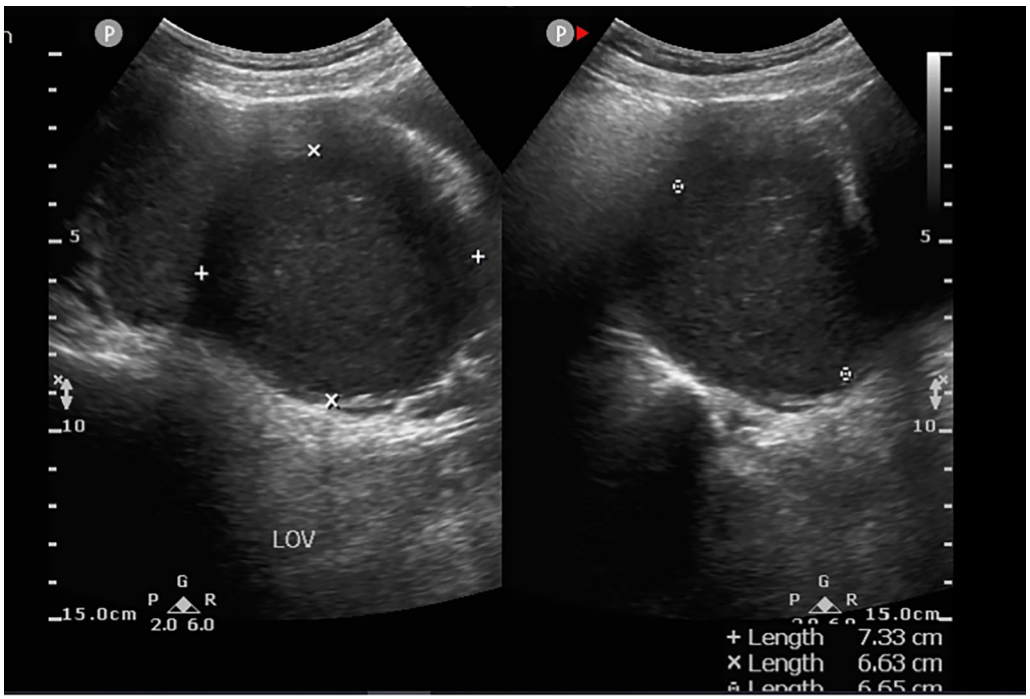

Fig. 7 An echograph showing the left ovary and cyst with turbid content

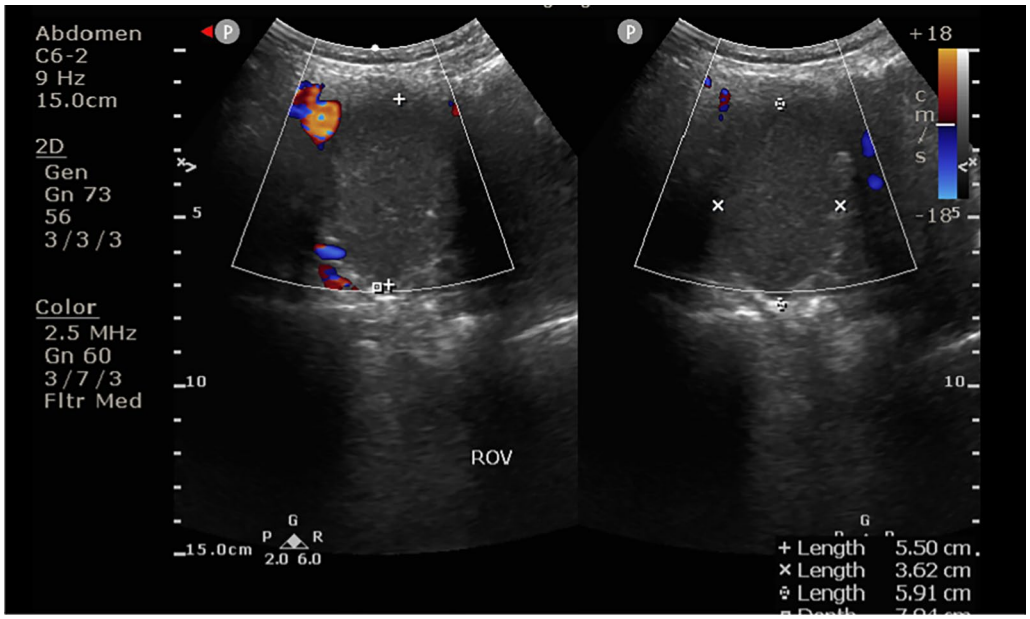

Fig. 8 An echograph showing the right ovary and cyst with turbid content

to care, with the hope that medical advancements will enhance outcomes for these patients.

Moreover, situs ambiguous is more prevalent in closely related Asian and Muslim communities, with a frequency of 1 in 2700, than in the English population, where it occurs at a rate of 1 in 24,000. Asian populations have a greater incidence of heterotaxy syndrome. Additionally, polysplenia syndrome is more frequently observed in females. Determining the exact occurrence of polysplenia in adults is challenging owing to the absence of severe congenital heart disease. Given the absence of a single definitive anomaly that defines this rare condition, it is recommended that the diverse range of anomalies be individually described [11].

Congenital heart disease (CHD) is described as an abnormality in the heart structure or the intrathoracic great vessels that leads to functional problems. Generally, 22–45% of patients with CHD have concomitant extracardiac anomalies or genetic syndromes. The prevalence of renal and urinary tract abnormalities in CHD patients is estimated to be approximately 7.5–12.5%, which is higher than that in the general population [12].

Congenital heart disease is one of the major causes of morbidity and mortality in patients with situs abnormalities, and its incidence can be predicted when the patient's situs is known. Specifically, the incidence of CHD in patients with situs solitus totalis (with levocardia) is very low (0.6–0.8%) [5]. However, patients with situs solitus and dextrocardia have a very high incidence of CHD, reported as greater than 90% [5]. In patients with situs inversus totalis, the incidence of CHD is relatively low, at 3–5% [9].

Mirror-image dextrocardia is a condition in which the heart is a left–right mirror image of a normal heart. Dextrocardia can be associated with situs inversus and other cyanotic and acyanotic congenital heart diseases, including ventricular septal defects. The most common form of dextrocardia in adults is characterized by L-loop ventricles and inverted great vessels (situs inversus totalis or mirror-image dextrocardia), which are present in 1 to 2 out of every 20,000 individuals in the normal population. Dextroversion is the second most common type of dextrocardia associated with situs solitus, D-loop ventricles, and normal great arteries. Dextrocardia can also be associated with heterotaxy syndromes of asplenia and polysplenia, with a tendency toward noncyanotic congenital heart defects [13].

In our case, patients with generalized situs ambiguously presented with an abnormal arrangement of thoracoabdominal organs that did not follow a specific pattern. More specifically, cardiac looping is often affected, and the abdominal organs may be frankly reversed or arranged in an unusual fashion, such as in the midline or in a bilaterally symmetric manner. Commonly associated findings are intestinal malrotation, abnormal venous drainage patterns, and congenital heart disease.

Endometriosis is a chronic gynecological disease that is estimated to affect 10% of reproductive-age women. Recent insights have linked endometriosis to several pathological mechanisms, including systemic inflammation,

a proatherogenic lipid profile, and enhanced oxidative stress, and endothelial dysfunction. The pathogenesis of endometriosis, which is considered a systemic disease, and the impact of the disease remains poorly understood. The true prevalence of endometriosis remains unknown since, historically, diagnoses have only been made using laparoscopy and, more recently, multimodal imaging techniques. Misdiagnosis remains common, and diagnosis is often largely delayed by years [14].

Ureteropelvic junction obstruction typically refers to blockage at the junction of the renal pelvis and the beginning of the ureter. The etiology of UPJ obstruction includes congenital and acquired conditions. Most cases are congenital owing to intrinsic and/or extrinsic causes, and approximately 13,000 newborns are diagnosed with this condition each year in the USA [7].

Ureteropelvic junction (UPJ) obstruction is one of the most frequent cases of obstructive nephropathy. The usual presentations of UPJ obstruction in patients include incidental detection via ultrasonography or the detection of flank pain. The main goals of radiological investigations used in the diagnosis of UPJ obstruction are to determine the anatomical site of the obstruction and the functional significance of the obstruction [8].

Ureteropelvic junction obstruction (UPJO) is more common in pediatric patients than in adults, and this anatomical pathology is more common in boys than in girls, with up to twice the number of cases in males compared with females. The left side is also affected twice as often as the right side [15].

The extrarenal pelvis refers to the renal pelvis located outside the confines of the renal hilum, representing a normal anatomical variant found in approximately 10% of the population. On ultrasound, the extrarenal pelvis may appear dilated, which could mistakenly suggest an obstructive pathology. However, further investigations with CT often correct this misinterpretation. By using CT/magnetic resonance imaging (MRI), a normal extrarenal pelvis can be identified by the presence of normal renal cortical thickness, bilateral symmetrical contrast excretion, and calyces that appear normal. It might be considered in the differential diagnosis of hydronephrosis, regardless of the cause, such as UPJO [16].

We can attribute UPJO and the extrarenal pelvis to situs ambiguity as potential manifestations of the condition. Situs ambiguous is known for its lack of a specific pattern, with the urinary system and genitals being among the most affected systems by its abnormalities. Nevertheless, it cannot be conclusively stated that endometriosis is directly linked to situs ambiguity.

In light of the above, heterotaxy syndromes may involve numerous anatomical abnormalities across multiple organ systems, with variations in ambiguous situs cases differing. While attempts have been made to classify cases into two main groups, the spectrum of variations is broad, making a single description impossible.

Here, we present the case of a 26-year-old female patient with a significant medical history of heterotaxy, congenital heart disease (CHD), dextrocardia, and ureteropelvic junction obstruction, all of which were diagnosed in the second month after birth. Additionally, she was more recently diagnosed with endometriosis. Given the rarity of heterotaxy syndrome, further data are needed to ensure optimal medical care for this population. As mortality and morbidity rates decrease and more individuals with heterotaxy reach adulthood, it is crucial to utilize imaging modalities to monitor these variations closely and establish a multidisciplinary team for comprehensive care.

Clinicians should also be knowledgeable about heterotaxy syndrome and its rare manifestations, such as pulmonary hypertension (PHTN), urinary system abnormalities, and dextrocardia.

Abbreviations

HS	Heterotaxy syndrome
PHTN	Pulmonary hypertension
PAPVR	Partial anomalous pulmonary venous return
UPJO	Ureteropelvic junction obstruction
CHD	Congenital heart disease
ASD	Atrial septal defect
VSD	Ventricular septal defect

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Author contributions

MHM (physician and supervisor) analyzed and interpreted the patient data. MAZ was responsible for communication with the patient and was a minor contributor in writing the manuscript. AA was a major contributor in writing the manuscript. HA was responsible for the radiological approach. All authors read and approved the final manuscript.

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Availability of data and materials

Data sharing does not apply to this article as no datasets were generated or analyzed during the current study.

Declarations

Ethics approval and consent to participate

Not applicable.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests

The authors declare that they have no competing interests.

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