



Dextrocardia with Situs Inversus Totalis and Subaortic-Type Double Outlet Right Ventricle in an Adult: A Case Report

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ABSTRACT

Background: Dextrocardia with situs inversus totalis (SIT) is a rare congenital condition, and significant congenital heart disease (CHD) complicates only approximately 0% to 10% of affected individuals. The coexistence of subaortic-type double outlet right ventricle (DORV) in adults is exceptionally uncommon. Mirror-image anatomy further complicates cardiovascular assessment and may lead to diagnostic misinterpretation without systematic segmental analysis and multimodal imaging.

Case Presentation: A 29-year-old man presented with chronic cyanosis, exertional dyspnea, and a three-month history of worsening symptoms accompanied by 5 kg of unintentional weight loss. Chest radiography demonstrated a right-sided gastric bubble. Transthoracic echocardiography identified a large subaortic ventricular septal defect (VSD) measuring 27.3 mm and

a left ventricular ejection fraction of 51%. Contrast-enhanced cardiac computed tomography (CT) with three-dimensional reconstruction confirmed dextrocardia, SIT, atrial situs inversus, a right aortic arch, subaortic-type DORV with aortic override, and imaging features suggestive of pulmonary hypertension.

Conclusion: This case underscores the diagnostic value of cardiac CT in definitively classifying complex conotruncal anatomy in the setting of mirror-image laterality. Early referral to an adult congenital heart disease center and formal pulmonary vascular assessment are essential when corrective intervention remains under consideration. Radiologists should maintain high suspicion for visceral situs inversus when a right-sided gastric bubble is identified on chest radiography.

Keywords: situs inversus totalis; cardiac computed tomography; dextrocardia; double outlet right ventricle; pulmonary hypertension

INTRODUCTION

The global burden of adult congenital heart disease (ACHD) continues to increase as improved diagnostics and interventions extend survival into adulthood.¹ Dextrocardia is defined as congenital placement of the heart in the right hemithorax with rightward apex orientation.² When accompanied by mirror-image reversal of thoracoabdominal viscera, the condition is termed situs inversus totalis (SIT).³ SIT affects approximately 1 in 6,000 to 1 in 35,000 live births and is frequently diagnosed incidentally in asymptomatic individuals.⁴ Unlike dextrocardia with situs solitus, significant congenital heart disease (CHD) occurs in only 0% to 10% of patients with SIT; however, accurate segmental cardiovascular analysis remains essential when cardiac anomalies coexist.^{2,5}

Double outlet right ventricle (DORV) is a complex conotruncal anomaly in which both great arteries arise predominantly from the morphologic right ventricle, encompassing diverse anatomic subtypes with distinct hemodynamic and surgical implications.^{1,6} Subaortic-type DORV, characterized by a subaortic ventricular septal defect (VSD) and an anterior rightward aortic position, represents one of the most surgically relevant forms because intraventricular biventricular repair is often feasible.^{6,7} Multimodal cardiovascular imaging including echocardiography, cardiac computed tomography (CT), and cardiovascular magnetic resonance (CMR) enables systematic segmental assessment of atrial situs, ventricular looping, and great artery relationships.^{8,9}

The coexistence of dextrocardia, SIT, and DORV is exceptionally uncommon, particularly in adults, as most published reports describe incidental SIT or simple associated defects such as isolated VSD.^{5,10} Mirror-image anatomy further complicates diagnosis and may lead to misinterpretation if standard imaging conventions are not adapted.^{2,11} Here, we report an adult case of dextrocardia with SIT and subaortic-type DORV, emphasizing multimodal imaging findings, segmental cardiovascular analysis, and clinical implications for diagnosis and management.

CASE PRESENTATION

A 29-year-old man presented with a long-standing history of congenital heart disease since childhood, manifesting as chronic cyanosis, exertional dyspnea, and easy fatigability. Over the three months preceding presentation, he reported progressive worsening of exertional dyspnea and fatigue, accompanied by an unintentional weight loss of 5 kg. He denied syncope, chest pain, hemoptysis, or recurrent lower respiratory tract infections. There was no history of prior cardiac surgery, cardiac catheterization, cigarette smoking, alcohol use, or familial congenital heart disease. The patient had been hospitalized previously for heart-related complaints.

Initial cardiovascular evaluation included chest radiography and transthoracic echocardiography. Chest radiography demonstrated cardiomegaly, a right-sided gastric bubble suggestive of abdominal situs inversus, and normal pulmonary vascularity (Figure 1). Transthoracic echocardiography revealed a left ventricular ejection fraction of 51%. The right atrium and right ventricle were normal in size. The left ventricle demonstrated preserved linear dimensions with concentric hypertrophy. A large subaortic VSD measuring 27.3 mm in diameter was identified. No additional shunt lesions, including atrial septal defect (ASD) or patent ductus arteriosus (PDA), were observed.

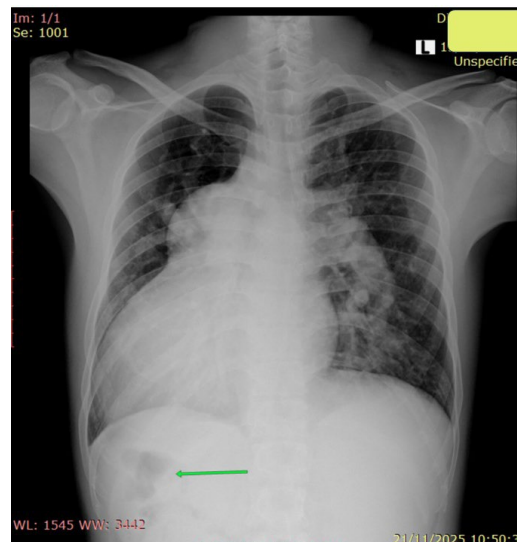


Figure 1. Posteroanterior chest radiograph demonstrating dextrocardia with the cardiac silhouette predominantly in the right hemithorax, cardiomegaly, and a right-sided gastric bubble (arrow) suggestive of visceral situs inversus.

Because of clinical suspicion of DORV, contrast-enhanced cardiac CT with three-dimensional reconstruction was subsequently performed for comprehensive anatomic characterization. CT confirmed dextrocardia with the heart positioned in the right hemithorax. Situs inversus totalis was demonstrated by mirror-image arrangement of the abdominal viscera, including a left-sided liver, right-sided spleen, and right-sided stomach (Figure 2). Dextrocardia with the heart in the right hemithorax, atrial situs inversus, and a right aortic arch were also identified on subsequent CT images.

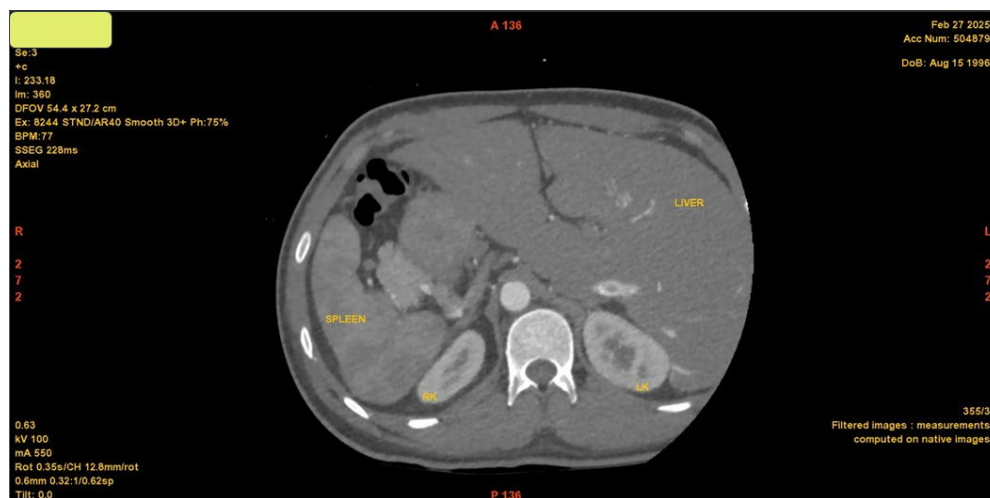


Figure 2. Contrast-enhanced cardiac CT in axial view confirming mirror-image arrangement of abdominal viscera (left-sided liver and right-sided spleen and stomach).

Short-axis CT images at the ventricular level showed a large subaortic VSD, concentric left ventricular hypertrophy, and right ventricular hypertrophy (Figure 3). Long-axis CT images further delineated subaortic-type DORV with aortic override of the large VSD (Figure 4).

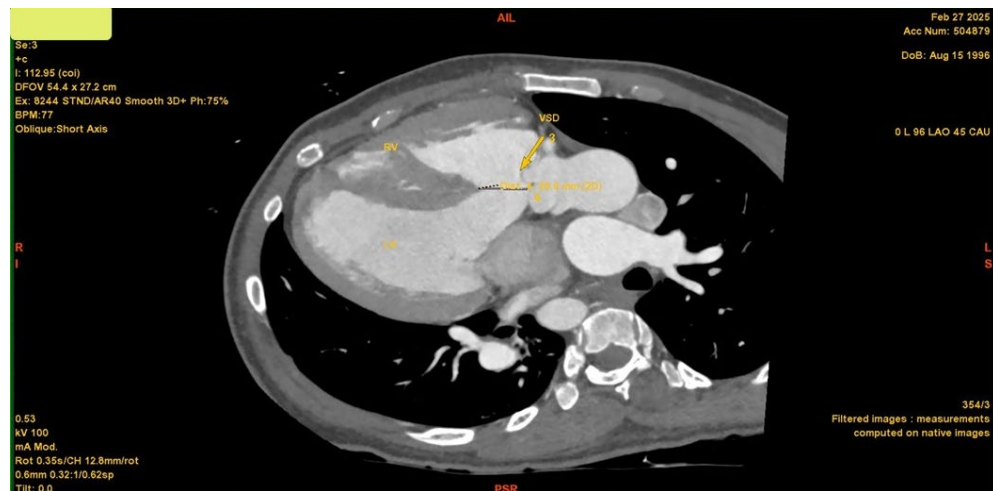


Figure 3. Contrast-enhanced cardiac computed tomography in short-axis view at the ventricular level showing a large subaortic VSD, concentric left ventricular hypertrophy, and right ventricular hypertrophy. LV, left ventricle; RV, right ventricle; VSD, ventricular septal defect.

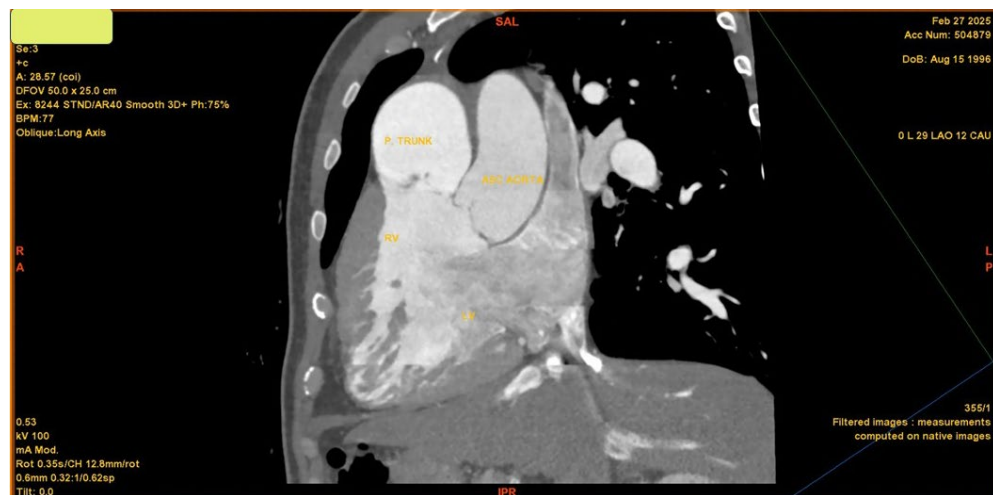


Figure 4. Contrast-enhanced cardiac computed tomography in long-axis view demonstrating subaortic-type double outlet right ventricle (DORV) with aortic override of the large ventricular septal defect. A, aorta; LV, left ventricle; PA, pulmonary artery; RV, right ventricle; VSD, ventricular septal defect.

Cardiac CT confirmed subaortic-type DORV with both the aorta and pulmonary artery arising predominantly from the morphologic right ventricle, a large subaortic VSD measuring 27.3 mm with aortic override, and no subaortic, subpulmonic, or pulmonary valvular stenosis. The right ventricle showed hypertrophy. Although echocardiography demonstrated preserved linear left ventricular dimensions with concentric hypertrophy, cardiac CT showed left ventricular dilatation with concentric remodeling, a finding more consistent with chronic pressure and volume loading on cross-sectional assessment. The right and left atria were normal in size. The main pulmonary artery was dilated, and imaging features were suggestive of pulmonary hypertension. No associated ASD, PDA, coronary artery anomaly, coarctation of the aorta, or atrioventricular valvular abnormality was identified.

The cardiac CT impression was summarized as follows: (1) dextrocardia with situs inversus; (2) double outlet right ventricle, subaortic type, with both great arteries arising from the right ventricle; (3) large VSD measuring 27.3 mm with overriding aorta; (4) dilated right and left ventricles with concentric left ventricular hypertrophy; and (5) findings suggestive of pulmonary hypertension.

DISCUSSION

This case illustrates an exceptionally rare adult presentation of dextrocardia with situs inversus totalis (SIT) complicated by subaortic-type double outlet right ventricle (DORV), a large ventricular septal defect (VSD), and imaging features suggestive of pulmonary hypertension. Although SIT occurs in approximately 1 in 6,000 to 1 in 35,000 live births and is frequently identified incidentally in asymptomatic individuals, significant congenital heart disease (CHD) is found in only a small subset of patients, with reported rates of approximately 0% to 10%.^{4,5,12} Conversely, dextrocardia associated with situs solitus is much more commonly accompanied by complex cardiac malformations.² The present patient therefore represents an uncommon subgroup of mirror-image laterality in whom a major conotruncal anomaly, rather than an isolated positional variant, accounts for chronic cyanosis and progressive functional decline into adulthood.

The clinical course of this 29-year-old man is consistent with long-standing uncorrected complex CHD. Chronic cyanosis, exertional dyspnea, and easy fatigability since childhood reflect persistent intracardiac shunting and reduced systemic oxygen delivery. The three-month history of worsening symptoms accompanied by unintentional weight loss raises concern for deteriorating cardiopulmonary status, which in unrepaired shunt lesions may be related to progressive pulmonary hypertension, declining ventricular function, or superimposed volume overload.¹ Unlike many published adult cases of SIT discovered incidentally on routine imaging without cardiovascular symptoms^{4,12}, this patient presented with overt hemodynamic compromise and had required prior hospitalization for cardiac complaints, underscoring the severity of the underlying anatomy.

From a morphologic standpoint, DORV is defined as a ventriculoarterial connection in which both great arteries arise entirely or predominantly from the morphologic right ventricle, yet it encompasses a broad spectrum of anatomic and hemodynamic subtypes.^{6,13} Subaortic-type DORV, as identified in this case, is characterized by a subaortic VSD in close relationship to the aortic outflow tract and represents one of the most surgically relevant forms because intraventricular biventricular repair is often feasible when pulmonary vascular disease has not become irreversible.^{6,7} The large subaortic VSD measuring 27.3 mm, together with aortic override and the absence of subaortic, subpulmonic, or pulmonary valvular stenosis, likely permitted high pulmonary blood flow from early life. Over time, this physiology may promote pulmonary vascular remodeling and pulmonary hypertension, supported in the present patient by main pulmonary artery dilatation on cardiac CT.^{1,6} A preserved left ventricular ejection fraction of 51% and concentric left ventricular hypertrophy on echocardiography are more consistent with chronic pressure and volume loading than with univentricular physiology. This distinction is clinically important because adult DORV with situs inversus has been reported in association with univentricular physiology and surgically prohibitive anatomy¹⁰, whereas the present case demonstrates morphologically distinct biventricular chambers with a large VSD committed to the aorta, a profile more closely aligned with potentially corrective subaortic DORV described in the surgical literature.^{6,7}

Accurate diagnosis in this setting requires systematic segmental cardiovascular analysis and cannot rely on conventional anatomic assumptions alone. Mirror-image arrangement of the

thoracoabdominal viscera, including a left-sided liver, right-sided spleen, and right-sided stomach, confirmed SIT and corresponded with the right-sided gastric bubble initially suspected on chest radiography. Cardiac CT subsequently established dextrocardia, atrial situs inversus, and a right aortic arch, thereby completing the anatomic framework required for classification of the conotruncal malformation.^{2,3} This stepwise evaluation reflects the recommended diagnostic approach to disorders of laterality, in which chest radiography may provide early clues to visceral situs, echocardiography supplies functional and hemodynamic information, and cross-sectional imaging delineates complex spatial relationships that are often difficult to assess on transthoracic echocardiography alone.^{9,11} In this case, echocardiography identified a large subaortic VSD and provided information on ventricular size, whereas contrast-enhanced cardiac CT with three-dimensional reconstruction was crucial in confirming DORV subtype, aortic override, great artery origin, and aortic arch anatomy. These findings align with current evidence emphasizing the value of cardiac CT in the preoperative assessment of DORV, including the relationship of the VSD to the great arteries, outflow tract geometry, and coronary anatomy.^{6,9}

Comparison with previous reports reveals both similarities and important differences. Darbari et al. (2024) described an adult with dextrocardia, situs inversus, and a small perimembranous VSD, with emphasis on surgical complexity related to mirror-image exposure and cannulation rather than the severity of the intracardiac defect itself. The present patient differs substantially by harboring a large subaortic VSD with DORV, a far more complex conotruncal lesion with greater hemodynamic consequences. Arenas-Fabbri et al. (2025) reported another rare adult combination of total situs inversus and DORV, but in the setting of univentricular physiology with marked left ventricular hypoplasia, rendering corrective surgery unfeasible. By contrast, the current case demonstrates morphologically distinguishable ventricles without evidence of functionally single-ventricle physiology, thereby warranting further evaluation by an adult congenital heart disease team for possible corrective or palliative intervention, provided formal assessment of pulmonary vascular resistance is performed.¹ Survival into the third decade of life without prior cardiac surgery is noteworthy and may reflect a balance between high pulmonary flow, progressive pulmonary hypertension, and partially compensated ventricular function;

however, the lack of detailed historical records limits interpretation regarding potentially missed opportunities for earlier intervention.

This case offers several practical lessons for radiologists and clinicians. First, a right-sided gastric bubble on chest radiography should raise suspicion for visceral situs inversus and prompt segmental interpretation of all subsequent cardiac imaging.¹¹ Second, clinical suspicion of DORV in a cyanotic adult should lead to comprehensive cross-sectional imaging even when echocardiography has already identified a large VSD, because subtype classification and surgical planning require precise definition of ventriculoarterial connections, the degree of override, and associated aortic arch and coronary anatomy.^{6,9} Third, the combination of dextrocardia, SIT, and subaortic-type DORV should be recognized as an exceptionally rare but clinically consequential entity that may present in adulthood with progressive cyanotic heart disease rather than as an incidental finding. Fourth, imaging features suggestive of pulmonary hypertension in unrepaired DORV should be communicated promptly to the referring clinical team, as they may indicate diminishing corrective options and the need for advanced pulmonary vasodilator therapy or palliative strategies.¹

Several limitations should be acknowledged. Physical examination findings, electrocardiographic data, pulmonary artery systolic pressure estimation, and cardiac catheterization results were unavailable for review. Long-term therapeutic decisions, surgical candidacy, and clinical outcome after CT were not documented in the available medical records. Accordingly, the principal contribution of this report to the literature lies in the description of a rare anatomic combination characterized through a multimodal imaging approach in an adult patient, rather than in the evaluation of therapeutic outcomes.

CONCLUSION

Dextrocardia with situs inversus totalis and subaortic-type double outlet right ventricle is an exceptionally rare combination, particularly when presenting symptomatically in adulthood with chronic cyanosis and progressive functional deterioration. In this 29-year-old man, a large subaortic

ventricular septal defect with aortic override and imaging features suggestive of pulmonary hypertension were identified in the setting of uncorrected complex congenital heart disease. Multimodal cardiovascular imaging, especially contrast-enhanced cardiac computed tomography with three-dimensional reconstruction, was essential for segmental anatomic characterization and definitive classification of the conotruncal malformation after initial evaluation with chest radiography and echocardiography.

This case underscores the need for heightened clinical and radiologic awareness of mirror-image laterality, early specialist referral to an adult congenital heart disease center, and formal pulmonary vascular assessment when corrective intervention remains under consideration. Prompt communication of findings suggestive of pulmonary hypertension is critical, as they may influence surgical candidacy and long-term management. We believe this report adds a valuable imaging-based description of a rare adult presentation to the existing literature and may assist clinicians and radiologists in recognizing and evaluating similar complex congenital cardiac anomalies.

DISCLOSURE OF POTENTIAL CONFLICT OF INTEREST

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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