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# Beyond Cyanosis: CT Angiographic Insights into Heterotaxy Syndrome with Left Isomerism in a Post Bilateral Bidirectional Glenn Shunt Patient

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**Abstract:** ***Heterotaxy syndrome represents a rare spectrum of congenital anomalies characterized by abnormal arrangement of thoracoabdominal organs along the left-right body axis. Left isomerism (polysplenia syndrome) is frequently associated with complex congenital cardiac malformations, interrupted inferior vena cava (IVC) with azygos continuation, and abnormal pulmonary and systemic venous drainage. We present a case of a pediatric patient with known dextrocardia and status post bilateral bidirectional Glenn shunt who underwent CT pulmonary angiography for comprehensive postoperative evaluation. Imaging demonstrated a constellation of classic left isomerism findings: dextrocardia, transverse liver, polysplenia, bilateral morphologic left atrial appendages, common atrium with large atrial septal defect, membranous ventricular septal defect, hypoplastic right ventricle, double outlet right ventricle with supravalvular pulmonary stenosis, and interrupted left-sided IVC with azygos continuation. Bilateral superior vena cavae were identified draining into patent bilateral bidirectional Glenn shunts. Bronchopulmonary findings included bilateral hyparterial bronchi with discordant lung lobation. No pulmonary thromboembolism was detected. This case underscores the indispensable role of multidetector CT angiography in providing comprehensive characterization of complex cardiovascular and thoracoabdominal anatomy in heterotaxy syndrome, particularly in the postoperative setting where echocardiography may be limited.***

**Keywords:** Heterotaxy syndrome; Left isomerism; Polysplenia syndrome; Dextrocardia; Bidirectional Glenn shunt; Double outlet right ventricle; Interrupted inferior vena cava; CT pulmonary angiography; Congenital heart disease

## 1. Introduction

Heterotaxy syndrome is a rare but clinically significant congenital disorder defined by the failure of normal thoracoabdominal organ lateralization along the left-right body axis. The resultant anatomical configurations range from partial to near-complete mirror imaging of normally positioned structures, producing a highly variable and complex phenotype that encompasses both cardiac and extracardiac anomalies. The estimated incidence ranges from approximately 1 in 10,000 to 1 in 40,000 live births, accounting for nearly 3% of all congenital heart diseases [1, 2].

The embryologic basis of heterotaxy syndrome lies in disruption of the molecular signaling cascade governing left-right axis determination during early gastrulation and cardiac looping. Several genes have been implicated in this process, including ZIC3, NODAL, LEFTY1, LEFTY2, PITX2, and CFC1, which orchestrate the Nodal signaling pathway critical for establishment of left-right asymmetry [3,4]. Approximately 20-25% of cases are associated with primary ciliary dyskinesia, including Kartagener syndrome, underscoring the role of motile cilia in left-right patterning [5].

Heterotaxy syndrome is broadly classified into two subtypes based on atrial appendage morphology. Right isomerism (asplenia syndrome) is characterized by bilateral right-sidedness, asplenia, bilateral trilobed lungs with eparterial bronchi, and frequently severe cyanotic heart disease. Left isomerism (polysplenia syndrome), the subject of this report, is characterized by bilateral left-sidedness and is classically associated with polysplenia, interrupted inferior vena cava (IVC) with azygos or hemiazygos continuation, bilateral morphologic left atrial appendages, bilateral bilobed lungs with hyparterial bronchi, and variable but often complex congenital cardiovascular anomalies [1,6].

The cardiovascular phenotype of left isomerism is highly variable and may range from relatively mild septal defects to severe cyanotic malformations including double outlet right ventricle (DORV), complete atrioventricular septal defects, and single ventricle physiology. Associated anomalies frequently necessitate staged surgical palliation, including the bidirectional Glenn shunt and ultimately Fontan circulation [7]. In patients with bilateral superior vena cavae, bilateral bidirectional Glenn shunts- connecting each SVC to the ipsilateral pulmonary artery—may be performed to redirect superior caval return into the pulmonary circulation.

Accurate anatomical delineation is paramount in these patients, as surgical planning and long-term prognosis critically depend on precise understanding of intracardiac anatomy, systemic and pulmonary venous pathways, great vessel relationships, and extracardiac anomalies. While echocardiography remains the first-line modality for initial cardiac evaluation, its acoustic window limitations and inability to fully characterize extracardiac vascular anatomy—particularly in postoperative patients—may necessitate complementary cross-sectional imaging [8]. Multidetector CT (MDCT) angiography has emerged as an invaluable tool in this context, offering high spatial resolution, rapid acquisition, and the capability for multiplanar and three-dimensional reconstruction, enabling comprehensive evaluation of complex congenital cardiovascular anatomy [8,9].

We present a case of complex cyanotic congenital heart disease with heterotaxy syndrome and left isomerism in a post bilateral bidirectional Glenn shunt patient, evaluated by CT pulmonary angiography. This case illustrates the full imaging spectrum of left isomerism and demonstrates the indispensable role of MDCT in postoperative evaluation and future surgical planning.

## 2. Case Report

A pediatric patient, aged 9 years with a known diagnosis of dextrocardia and complex cyanotic congenital heart disease presented for CT pulmonary angiography evaluation. The patient had previously undergone bilateral bidirectional Glenn shunt surgery at 3 months of age. The clinical indication for CT imaging included comprehensive postoperative vascular anatomical assessment and exclusion of pulmonary thromboembolism.

CT pulmonary angiography was performed using a multidetector CT scanner with intravenous iodinated contrast administration, following an age-appropriate low-dose pediatric protocol with bolus-tracking technique. Images were reconstructed in axial, coronal, and sagittal planes, with additional curved multiplanar and maximum intensity projection (MIP) reformations generated for detailed vascular assessment.

## 3. Imaging Findings

**Chest Radiograph**

The frontal chest radiograph demonstrated dextrocardia with the cardiac apex directed toward the right hemithorax. Median sternotomy sutures were identified, consistent with prior cardiac surgery. No acute cardiopulmonary process was identified on the plain radiograph.

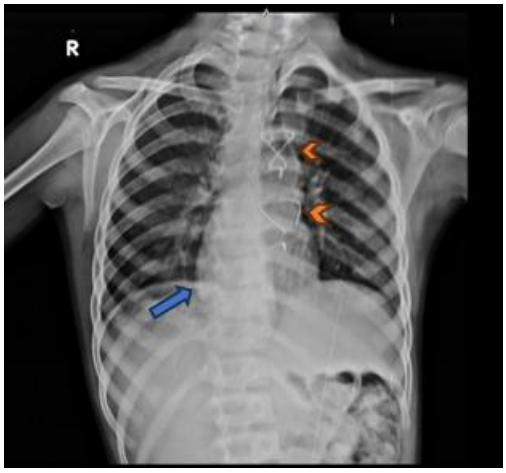



**Figure 1:** Frontal Chest Radiograph

Frontal chest radiograph demonstrating dextrocardia with right-sided cardiac apex (blue arrow). Median sternotomy wires are present in situ (orange arrowheads), consistent with prior cardiac surgery.

- Blue arrow: right-sided cardiac apex confirming dextrocardia
- Arrowheads: median sternotomy sutures from prior bilateral bidirectional Glenn shunt surgery

Note: Absence of a normal left-sided cardiac silhouette

**Cardiac Position and Situs**

CT angiography confirmed dextrocardia with the cardiac apex directed to the right hemithorax. The liver demonstrated a transverse, midline orientation—a hallmark of heterotaxy syndrome. The gallbladder and the remnant of the infra-hepatic IVC were positioned to the left of the spine, while the stomach and descending aorta were situated on the right side. Multiple small splenic nodules were identified in the upper left abdomen, consistent with polysplenia syndrome. These findings established an overall situs ambiguus with left isomerism.

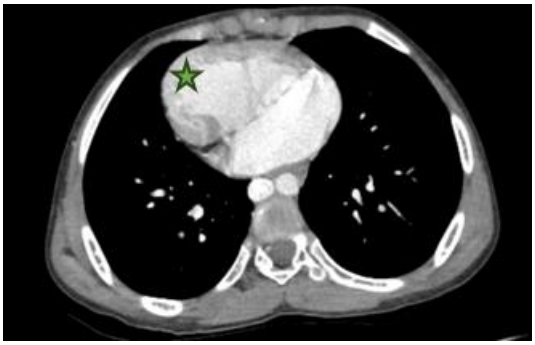
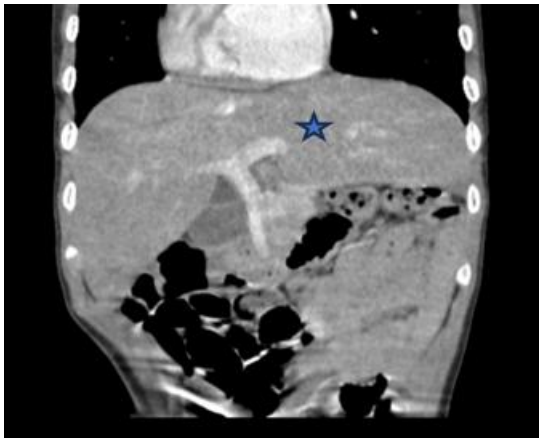


**Figure 2A**



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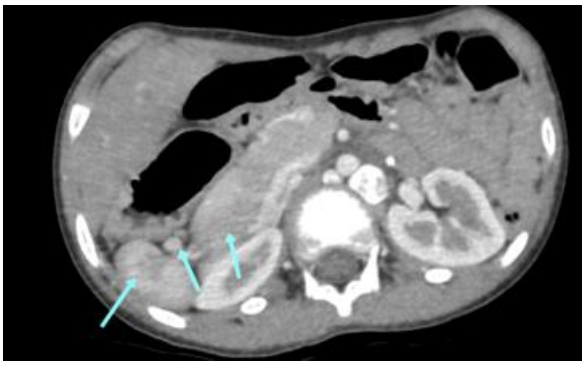

**Figure 2C**

**Figure 2:** Cardiac and Abdominal Situs and Polysplenia

A) Axial contrast-enhanced CT images demonstrating right sided heart apex (green star).
B) Coronal contrast enhanced CT images shows the liver in a transverse, midline position.
C) Axial contrast enhanced CT image shows multiple splenic nodules (polysplenia) in the left upper quadrant.
   - Green star: Right sided heart apex, confirming dextrocardia

- Blue star: Transverse, midline liver- classic heterotaxy finding
- Blue arrows: Multiple polysplenic nodules in the left upper quadrant

**Atria, Atrial Appendages, and Ventricular Findings**

Evaluation of the intracardiac anatomy revealed complete absence of the interatrial septum, resulting in a common atrial chamber compatible with a large atrial septal defect (common atrium). All visualized hepatic veins drained directly and without obstruction into this common atrial chamber. Bilateral atrial appendages demonstrated morphologic left atrial characteristics- with broad, finger-like trabeculated appendages- consistent with bilateral left atrial appendages and left isomerism.

A membranous ventricular septal defect measuring approximately 15.0 mm was identified. The right ventricle appeared hypoplastic, with a disproportionately smaller cavity size compared to the left ventricle. The aorta was identified arising from the hypoplastic right ventricle, a finding consistent with double outlet right ventricle (DORV) in conjunction with the pulmonary artery also originating from the right ventricle.

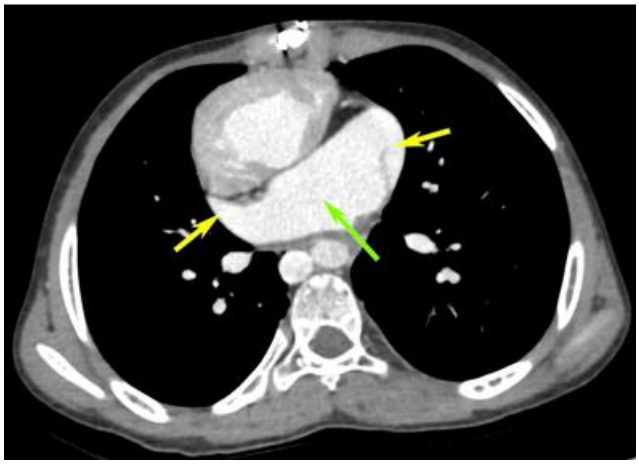

**Figure 3:** Intracardiac Anatomy: Common Atrium and Bilateral Left Atrial Appendages

Axial CT angiography image at the level of the atria demonstrating absence of the interatrial septum forming a common atrial chamber (green arrow). Bilateral atrial appendages (yellow arrows) exhibit morphologic left atrial characteristics with broad, finger-like configurations, consistent with bilateral left atrial appendages (left isomerism).

- Green arrow: common atrial chamber (absent interatrial septum)
- Yellow arrows: bilateral left atrial appendages with characteristic finger-like morphology

**Pulmonary and Systemic Venous Anatomy**

Bilateral superior vena cavae were present. The right superior vena cava was surgically anastomosed to the right pulmonary artery, and the left superior vena cava was anastomosed to the left pulmonary artery, consistent with patent bilateral bidirectional Glenn shunts. The anastomoses appeared widely patent without evidence of stenosis or thrombosis.

A key diagnostic finding was interruption of the infrahepatic/suprarenal portion of the left-sided inferior vena cava with azygos continuation. A mildly prominent hemiazygos vein was identified draining into a dilated azygos vein, which ascended in the posterior mediastinum and drained into the right superior vena cava. All four pulmonary veins drained into the common atrial chamber without evidence of anomalous pulmonary venous return or obstruction.

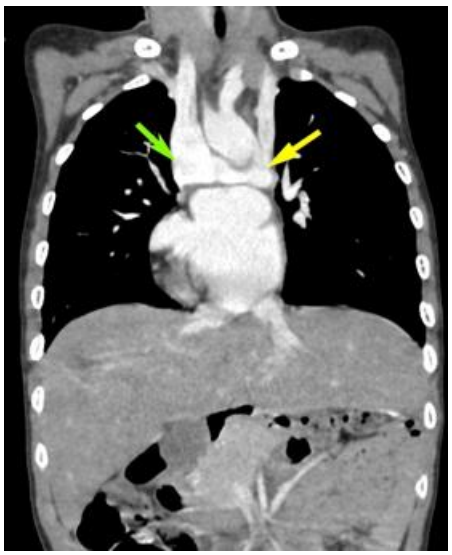

**Figure 4:** Bilateral Glenn Shunts

- Coronal CT angiography image demonstrating bilateral bidirectional Glenn shunts with right superior vena cava (green arrow) anastomosed to the right pulmonary artery and left superior vena cava (yellow arrow) anastomosed to the left pulmonary artery. Note: Patent anastomoses bilaterally without evidence of stenosis or thrombosis
- Green arrow: right superior vena cava anastomosed to right pulmonary artery (right Glenn shunt)
- Yellow arrow: left superior vena cava anastomosed to left pulmonary artery (left Glenn shunt)

**Great Vessel Abnormalities**

A left-sided aortic arch was present. The aorta arose from the hypoplastic right ventricle. The main pulmonary artery also originated from the right ventricle and demonstrated abrupt narrowing at its supravalvular level, measuring approximately 5.5 mm in cross-sectional diameter, consistent with supravalvular pulmonary stenosis. These findings confirmed double outlet right ventricle (DORV) with associated supravalvular pulmonary artery stenosis.

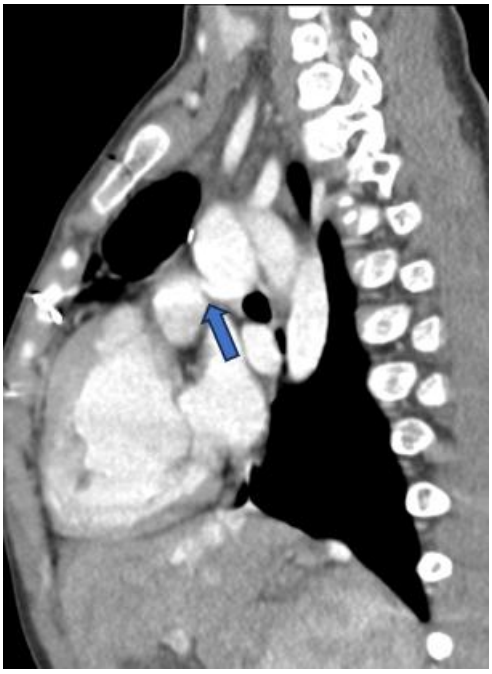

**Figure 5:** Supravalvular Pulmonary Stenosis
Sagittal oblique CT angiography reconstruction demonstrating the main pulmonary artery with abrupt supravalvular narrowing (blue arrow), consistent with supravalvular pulmonary stenosis.

- Blue arrow: supravalvular pulmonary stenosis- abrupt narrowing

**Bronchopulmonary Findings**

Evaluation of the bronchopulmonary anatomy revealed bilateral hyparterial bronchi—where the main bronchus courses below the level of the ipsilateral pulmonary artery on both sides. The right lung was bilobed and the left lung was trilobed, indicating discordant pulmonary lobation with reversal of the typical lobar pattern. These bronchopulmonary findings are characteristic of bilateral left-sidedness in left isomerism.

No focal consolidation, pulmonary mass, pleural effusion, or parenchymal airspace disease was identified. Critically, no pulmonary thromboembolism was detected; pulmonary arterial branches demonstrated adequate contrast opacification without filling defects. Median sternotomy sutures from prior surgery were noted in situ.

## 4. Discussion

This case illustrates a comprehensive imaging constellation of left isomerism with complex cyanotic congenital heart disease evaluated by CT pulmonary angiography in the postoperative setting. The spectrum of findings—from the characteristic polysplenia and interrupted IVC to the severe intracardiac malformations requiring bilateral Glenn palliation—exemplifies the diagnostic complexity that characterizes this syndrome.

**Embryology and Classification**

Heterotaxy syndrome results from disruption of the molecular lateralization cascade during early embryogenesis, specifically during the establishment of left-right asymmetry at the nodal cilia level. The motile cilia of the embryonic node generate directional fluid flow that activates asymmetric Nodal signaling on the left side, leading to left-sided expression of LEFTY1/2 and PITX2, which suppress right-sided identity and establish organ laterality. Mutations in genes encoding components of this pathway- including ZIC3 (X-linked), NODAL, LEFTY2, ACVR2B, and CFC1- have been identified in heterotaxy patients [3,4]. The association with primary ciliary dyskinesia in approximately 20-25% of cases provides further evidence for the cilia-dependent mechanism [5].

The classification into right and left isomerism reflects the type of bilateral symmetry imposed upon normally asymmetric structures. Left isomerism, as in this case, results in bilateral left-sidedness: bilateral morphologic left atria and atrial appendages, bilateral bilobed lungs with hyparterial bronchi, and multiple spleens (polysplenia)—all of which are normally left-sided structures present bilaterally [1,6].

**Characteristic Imaging Features of Left Isomerism**

The present case demonstrated multiple pathognomonic features of left isomerism, which we discuss in the context of the existing literature:

**Interrupted IVC with azygos continuation** is the single most characteristic imaging finding of left isomerism, occurring in approximately 65-90% of cases [1,2]. In this pattern, the infrahepatic IVC is absent and inferior venous return is redirected via the azygos (right-sided) or hemiazygos (left-sided) vein into the SVC, bypassing the heart. In our patient, the left-sided IVC was interrupted at the suprarenal level, with azygos continuation draining into the right SVC. This finding has important surgical implications, as it affects cardiopulmonary bypass cannulation strategies and Fontan pathway planning [7,10].

**Bilateral left atrial appendages** reflect the bilateral left-sidedness of left isomerism. The morphologic left atrial appendage is characterized by a broad-based, finger-like, trabeculated configuration with a narrow junction to the atrial body, contrasting with the broader, triangular morphology of the right atrial appendage. Identifying bilateral LAAs on CT is a reliable marker for left isomerism and can be assessed on axial or reformatted images [6,9].

**Polysplenia** results from bilateral development of what is normally the left-sided splenic tissue, producing multiple small splenic nodules rather than a single spleen. While the name implies multiple spleens, these are functionally active and thus patients with left isomerism have preserved (though sometimes variable) splenic function, distinguishing them immunologically from right isomerism patients who have functional asplenia and are at risk for overwhelming sepsis [1,5].

**Bilateral hyparterial bronchi** and discordant lung lobation are the bronchopulmonary correlates of left isomerism. Normally, the right bronchus is eparterial (crossing above the right pulmonary artery) and the left is hyparterial (crossing below the left pulmonary artery). In left isomerism, both bronchi adopt the hyparterial morphology, and the lung lobation may be discordant-as in our case, where the right lung was bilobed and the left trilobed, the reverse of normal [2,6].

**Double Outlet Right Ventricle in Heterotaxy Syndrome**

DORV is defined by the origin of both great arteries predominantly from the morphologic right ventricle and represents one of the more severe cardiac manifestations of heterotaxy syndrome. Its association with left isomerism, while less common than with right isomerism, is well documented and carries significant prognostic implications [7]. In the present case, DORV was associated with supravalvular pulmonary stenosis, further limiting pulmonary blood flow and necessitating early palliative surgery. The combination of DORV, hypoplastic right ventricle, common atrium, and large VSD in this patient represents a complex single-ventricle-type physiology that guided the decision for bilateral Glenn palliation.

**The Bilateral Bidirectional Glenn Shunt**

The bidirectional Glenn shunt connects the superior vena cava end-to-side to the ipsilateral pulmonary artery, directing passive superior caval venous return directly into the pulmonary circulation without requiring ventricular pumping. This reduces the volume load on the single functioning ventricle while improving pulmonary blood flow [7,10]. In patients with bilateral SVCs—as in this case, a common finding in left isomerism where the left SVC typically persists due to failure of regression—bilateral bidirectional Glenn shunts are required to capture all superior caval return. CT angiography in this patient confirmed bilateral patent Glenn anastomoses without stenosis or thrombosis, providing reassurance regarding shunt function and informing the timeline for Fontan completion.

**Role of CT Angiography in Heterotaxy Syndrome**

Multidetector CT angiography has become an indispensable imaging tool in the evaluation of complex congenital heart disease, particularly in the postoperative setting [8,9]. Its key advantages include: (1) high spatial resolution allowing delineation of small vascular structures and anastomoses; (2) rapid acquisition minimizing motion artifact; (3) simultaneous evaluation of the airways, lung parenchyma, abdominal viscera, and cardiovascular structures in a single examination; and (4) the ability to generate multiplanar and three-dimensional reconstructions that facilitate surgical planning and communication.

In heterotaxy syndrome specifically, MDCT provides information that often cannot be reliably obtained by echocardiography alone, including characterization of IVC continuity, azygos anatomy, Glenn shunt patency, pulmonary artery anatomy, and extracardiac anomalies such as abdominal situs and bronchopulmonary morphology [2,8,9]. The radiation dose concern in pediatric patients is mitigated by modern low-dose protocols, iterative reconstruction algorithms, and the diagnostic yield of CT in reducing the need for additional invasive procedures.

## 5. Take-Home Message

**Key Learning Points**

- Left isomerism (polysplenia syndrome) presents with a characteristic imaging constellation: interrupted IVC with azygos continuation, bilateral morphologic left atrial appendages, polysplenia, bilateral hyparterial bronchi, and complex congenital cardiovascular anomalies.
- CT pulmonary angiography provides comprehensive one-stop assessment of intracardiac anatomy, systemic and pulmonary venous pathways, great vessel relationships, airway morphology, and abdominal situs—information that is critical for both initial diagnosis and postoperative management.
- In postoperative Glenn shunt patients, CT angiography can confirm shunt patency, detect pulmonary thromboembolism, and characterize residual or progressive cardiovascular anomalies, directly informing the timing and planning of subsequent Fontan completion.
- Recognition of discordant lung lobation and bilateral hyparterial bronchi on CT, when combined with cardiovascular anomalies, should prompt systematic evaluation for the full spectrum of heterotaxy syndrome findings.

## 6. Conclusion

This case illustrates a rare and complex presentation of heterotaxy syndrome with left isomerism, encompassing severe cyanotic congenital heart disease including dextrocardia, common atrium, large VSD, hypoplastic right ventricle, DORV with supravalvular pulmonary stenosis, and interrupted IVC with azygos continuation, in a patient status post bilateral bidirectional Glenn shunts. CT pulmonary angiography provided comprehensive anatomical delineation of intracardiac defects, systemic and pulmonary venous anomalies, bronchopulmonary morphology, and postoperative vascular anatomy in a single examination.

Radiologists and clinicians should be familiar with the characteristic imaging features of left isomerism and appreciate that the cardiac phenotype can be highly variable and severe, extending well beyond the traditionally quoted mild-to-moderate spectrum. Multidetector CT angiography remains an indispensable complementary imaging modality to echocardiography in the evaluation of heterotaxy syndrome—particularly postoperatively—and plays a pivotal role in guiding staged surgical management toward Fontan completion. Early and accurate imaging diagnosis, with clear communication of the complex anatomy to the surgical team, is fundamental to optimizing outcomes in these challenging patients.

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