

Assessment of Double Outlet Right Ventricle in Echocardiography

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ABSTRACT

Background: Double outlet right ventricle (DORV) is a complex congenital heart disease in which both great arteries arise predominantly from the right ventricle, leading to highly variable anatomical and physiological manifestations. Its accurate diagnosis and classification are critical for surgical planning and long-term outcomes.

Methodology: This review synthesizes historical definitions, anatomical subtypes, pathophysiological implications, and the pivotal role of echocardiography in assessing DORV. Techniques including 2D echocardiography, Doppler, 3D imaging, and transesophageal echocardiography were evaluated alongside their integration with complementary imaging modalities such as MRI and CT. Clinical applications are illustrated through case studies, emphasizing echocardiography's diagnostic and perioperative role.

Results: Echocardiography remains the gold-standard diagnostic tool, enabling real-time visualization of ventriculoarterial connections, septal defects, outflow tract anatomy, and associated anomalies. It guides surgical planning by defining VSD position, routability, and ventricular function. Case studies highlight echocardiography's accuracy in detecting anomalies and its correlation with catheterization and MRI. Emerging technologies, including AI-assisted analysis, 3D printing, and hybrid imaging, are enhancing diagnostic precision and personalized surgical strategies. However, challenges such as technical limitations in neonates, operator dependency, and variable expertise persist.

Conclusion: Echocardiography is indispensable for the diagnosis, management, and follow-up of DORV, providing essential insights into anatomy, physiology, and surgical planning. Future directions include AI-driven automation, 3D printing for surgical rehearsal, and hybrid imaging for comprehensive cardiac evaluation, all of which will advance precision medicine in congenital cardiology.

Keywords: Double outlet right ventricle, Echocardiography, Congenital heart disease, Ventricular septal defect, Surgical planning, Paediatric cardiology.

1. INTRODUCTION

Double outlet right ventricle (DORV) is a heterogeneous congenital cardiac malformation defined as a condition in which both great arteries—the aorta and the pulmonary artery—arise predominantly or entirely from the morphologically right ventricle, rather than from their respective, anatomically correct ventricular chambers. This anomaly occupies a crucial place in the taxonomy of congenital heart diseases due to its anatomical complexity and its variable clinical presentation, ranging from mimicking ventricular septal defect (VSD) or tetralogy of Fallot (TOF) to resembling transposition of the great arteries (TGA) and even a functionally univentricular heart in some settings. Classically described, DORV encompasses a spectrum of anatomical subtypes with differences in morphology, location and size of associated septal defects, relationships between the great vessels, and various additional cardiac malformations such as subpulmonary or subaortic obstruction, atrioventricular septal defects (AVSD), and atypical valve connections¹. The epidemiological burden of DORV remains significant, representing approximately 1%–3% of all congenital heart disease cases, and the diversity in its morphological expression has prompted ongoing debate and evolution in its classification and management over the years².

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Historically, the concept and diagnosis of DORV have undergone considerable transformation. The condition was first recognized in the mid-20th century, with its initial description as partial transposition of the great arteries, wherein only the aorta is transposed. Early definitions focused primarily on the anatomical emergence of the great arteries from the right ventricle, but rapidly expanded to include considerations of ventricular topology, spatial relationships, and the presence or absence of ventricular septal communication. Notably, Maurice Lev's 1972 work shifted the diagnostic paradigm towards the importance of VSD position and the intricacies of great artery relationships, establishing a foundation for modern classification schemas. The International Society for Nomenclature of Paediatric and Congenital Heart Disease (ISNPCHD) contributed to the consensus definition: both arterial trunks must be supported predominantly, but not necessarily exclusively, by underlying morphologic right ventricular myocardium³. In parallel, notable pathologists like Taussig and Bing enriched the literature with alternative definitions centering on the leftward position of the aorta and specific subpulmonic VSDs, such as in the Taussig-Bing anomaly, a variant characterized by subpulmonic VSD and leftward aorta. Yet, the historical debate surrounding bilateral infundibula, fibrous continuity between valves, and the precise description of ventricular septal defects persists, with evidence indicating that features like the presence of bilateral infundibula are observed in only a minority of cases and are not, by themselves, diagnostic for DORV. Over the decades, the historical evolution of DORV diagnosis has been informed by advances in cardiac imaging, surgical anatomy, and nomenclature. The early reliance on postmortem pathological anatomy gave way to progressive adoption of imaging modalities such as transthoracic and transesophageal echocardiography, angiography, and, later, multidetector computed tomography (CT) and magnetic resonance imaging (MRI). This transition facilitated more nuanced and dynamic visualization of the interaction between the right ventricle and arterial trunks, including quantitative assessment of how much of each valve's circumference is derived from the right ventricle. During the latter half of the 20th century, pioneering surgical approaches and anatomical reviews broadened the scope of diagnostic consideration, leading to distinctions based on the size, type, and position of the VSD, the direction of blood flow, and the presence of associated anomalies, such as pulmonary stenosis or malformations of the atrial or ventricular septa⁴. The rationale for echocardiographic assessment in DORV—and by extension, in all congenital heart diseases derives from the modality's unparalleled ability to provide real-time, non-invasive, high-resolution visualisation of complex intracardiac anatomy. Echocardiography stands as the first-line diagnostic tool due to its capability to define ventricular topology, quantify arterial connections, locate and characterize septal defects, and evaluate associated anomalies that critically influence treatment strategy. In the context of DORV, echocardiography is indispensable for identifying the relationship and proportionate connection between the arterial trunks and the right ventricle, distinguishing DORV from other entities such as TGA, isolated VSD, or TOF where the arterial emergence or ventricular configuration does not meet DORV criteria. Precise echocardiographic imaging enables the clinician to delineate the VSD's position whether subaortic, subpulmonic, or noncommitted which has a direct bearing on hemodynamics and surgical repair⁵. Moreover, color Doppler and advanced echocardiographic techniques facilitate assessment of intracardiac flow dynamics, chamber pressures, and potential areas of obstruction, thus permitting individualized and optimized surgical planning.

Echocardiographic assessment also plays a fundamental role in identifying and classifying the multitude of associated cardiac anomalies frequently coexisting with DORV. These may encompass outflow tract obstructions, atrioventricular valve abnormalities, coarctation of the aorta, arch interruptions, mitral valve atresia, and heterotaxy syndromes. Without comprehensive echocardiographic evaluation, such coexisting malformations may be overlooked, potentially compromising perioperative management and long-term outcomes. In addition, echocardiography supports longitudinal follow-up post-surgery, allowing detection of residual shunts, subvalvular or outflow tract obstructions, and ventricular function, each of which can substantially impact the patient's prognosis. The modality's non-invasive nature confers significant advantages, especially in pediatric patients for whom repeated sedation for other imaging modalities is impractical⁶.

From a congenital heart disease perspective, echocardiography's relevance extends beyond mere anatomical description. The approach informatively guides preoperative, intraoperative, and postoperative decision-making, aids risk stratification, and supports multidisciplinary team discussions. In DORV, the detail provided by echocardiographic imaging often dictates the choice of surgical intervention, whether it be intraventricular tunnel repair, arterial switch procedures, or staged single-ventricle palliation in cases where biventricular repair is unfeasible? The application of three-dimensional echocardiography and adjunct multimodal imaging further expands the potential for accurate anatomical mapping, facilitating individualized care for each patient's unique cardiac configuration.

2. ANATOMY AND PATHOPHYSIOLOGY OF DORV

Double Outlet Right Ventricle (DORV) represents a formidable challenge in pediatric and adult congenital cardiology by virtue of its diverse anatomical presentations, intricate developmental origins, and profound pathophysiological implications. At its fundamental core, DORV is defined as a cardiac anomaly in which both the aorta and the pulmonary artery are connected predominantly, if not entirely, to the morphologically right ventricle, with a near-universal concomitant presence of a ventricular septal defect (VSD). This malformation is regarded as a conotruncal anomaly, highlighting its roots in aberrant development of the outflow tract portion of the embryonic heart⁸. A detailed understanding of the embryology, anatomical subtypes defined by VSD position, and the resultant hemodynamic disturbances is critical to optimal management and surgical intervention.

From the perspective of embryological development, the basis of DORV lies in errors of cardiac outflow tract morphogenesis, a process orchestrated during the early stages of fetal life. Normally, the developing heart undergoes septation and alignment events that direct the aorta to the left ventricle and the pulmonary artery to the right ventricle. The conotruncal region, comprising the truncus arteriosus and the conus cords, must undergo proper rotation, septation, and fusion with the ventricular septum. In DORV, this sequence is disturbed due to impaired migration and torsion of the truncoconal septum, conoventricular flange, and occasionally the atrioventricular region⁹. Pathogenetic studies reveal that abnormal connection and alignment can result from a misdirected posterior-anterior axis of the tricuspid orifice and the muscular ventricular septum, producing the interventricular foramen adjacent to abnormal flow paths. DORV may thus result from combinatorial defects in conotruncal, conoventricular (crista prima), or atrioventricular development, each imparting distinct anatomical and physiological phenotypes.

Variants of DORV are primarily categorized by the position and relationship of the VSD, which fundamentally governs the direction of blood flow, mixing, and the prospects for surgical repair. The four classic subtypes are:

- Subaortic VSD: Here, the VSD is situated immediately below the aorta, facilitating direct blood flow from the left ventricle through the VSD to the aorta. This arrangement optimizes systemic perfusion but often results in varying degrees of pulmonary overcirculation, unless modified by the presence of pulmonary stenosis. The subaortic variant frequently mimics the physiology of a simple VSD or tetralogy of Fallot depending on associated right ventricular outflow obstruction.
- Subpulmonic VSD (Taussig-Bing anomaly): In this configuration, the VSD is positioned beneath the pulmonary artery, so that oxygenated blood from the left ventricle is preferentially directed to the pulmonary circulation. This anatomical setup closely resembles the physiology of complete transposition of the great arteries, with deoxygenated systemic venous blood recirculated through the body unless corrective shunting is instituted.
- Doubly committed VSD: The VSD in this case lies beneath both great arteries, with direct commitments to the aorta
 and pulmonary artery. There is frequent absence of the muscular outlet septum, replaced by a fibrous continuity
 between the semilunar valves. This variant enables parallel blood flow to both circuits but increases risk for
 admixture and may variably impact pulmonary and systemic circuits dependent on additional lesions.
- Non-committed (or remote) VSD: Here, the VSD is located far from both outflow tracts, typically neither right
 below the aorta nor the pulmonary artery. The non-committed subtype is often associated with more complex
 intracardiac connections and remote blood flow patterns, sometimes necessitating single-ventricle repair strategies.

These anatomical subtypes are also influenced by the spatial positioning of the great vessels (side-by-side, anterior-posterior, or malposed), infundibular continuity, and varying degrees of right ventricular outflow obstruction and other cardiac anomalies.

The pathophysiological manifestations of DORV are a direct consequence of the altered connections and mixing within the heart, resulting in a spectrum of hemodynamic derangements. Cyanosis, pulmonary overcirculation, and systemic hypoperfusion are the most salient clinical sequelae, and their presentation is determined by the subtype of DORV and the magnitude of coexisting lesions. In the subaortic VSD type, aortic flow derived from the left ventricle may ensure good systemic oxygenation, especially in the absence of pulmonary stenosis. However, disease progression often features excessive pulmonary flow, potentially culminating in congestive heart failure and pulmonary vascular disease if left uncorrected. Conversely, in the subpulmonic (Taussig-Bing) type, oxygen-rich left ventricular output is directed to the lungs, while deoxygenated right ventricular blood flows to the aorta, precipitating profound cyanosis the classical presentation of transposed great arteries physiology¹⁰. The doubly committed and non-committed VSD forms can present with admixture physiology, where outputs to both aorta and pulmonary arteries are equally mixed, often causing variable degrees of cyanosis and pulmonary/systemic imbalance.

Hemodynamic repercussions in DORV are complex and can include high pulmonary blood flow with attendant pulmonary hypertension, low systemic oxygen saturation, right ventricular volume and pressure overload, and congestive heart failure. The clinical presentation is therefore contingent on the extent of pulmonary stenosis, the size and relative position of the VSD, arch anatomy, and the presence or absence of associated extracardiac anomalies. These factors are vital in determining prognosis, therapeutic strategy, and the feasibility of biventricular versus univentricular repair¹¹.

Echocardiographic Techniques in DORV Assessment

Echocardiographic assessment of Double Outlet Right Ventricle (DORV) requires a highly nuanced, multimodal approach, leveraging 2D, Doppler, 3D, and transesophageal modalities to visualize intracardiac anatomy, flow dynamics, and complex spatial relationships. Each technique plays a pivotal role in diagnosis, surgical planning, and postoperative evaluation, while practical limitations must be acknowledged for optimal patient care.

D Echocardiography: Structural Visualization

2D echocardiography remains the cornerstone in the initial evaluation of DORV, providing real-time anatomical delineation and permitting direct visualization of ventriculoarterial connections, the position of septal defects, and associated anomalies. The main diagnostic features involve identifying the parallel orientation and origin of both great arteries from the anterior right ventricle, the lack of continuity between the anterior mitral leaflet and any semilunar valve, and the absence of left ventricular outflow other than a VSD. Parasternal long-axis and short-axis scans are instrumental in demonstrating muscular conus separation and spatial relationships between the great vessels¹². These views also clarify the relative position of the VSD, which is crucial for both diagnosis and surgical planning, as well as revealing unexpected atrioventricular valvular anomalies, such as annular override, straddling, or cleft mitral valve.

Color Doppler and Spectral Doppler Flow Assessment

Color Doppler and continuous/spectral Doppler techniques are essential for functional assessment, enabling visualization of intracardiac shunting, quantification of blood flow direction, and characterization of velocity profiles across septal defects and valves. Color Doppler imaging elucidates bidirectional shunting through VSDs, left-to-right or right-to-left flows, and highlights pressure gradients indicative of pulmonary hypertension or outflow tract obstruction. These modalities help in monitoring for signs of hypoxia, assessing ventricular and atrial pressures, and documenting pulmonary versus systemic flow patterns. Spectral Doppler is especially helpful for quantifying peak velocities, diastolic and systolic gradients, and evaluating for tricuspid and pulmonary regurgitation, which often accompany complex DORV physiology¹³.

Echocardiography for Spatial Relationships

3D and 4D echocardiography (particularly spatiotemporal image correlation, STIC) provide incremental benefit in understanding the intricate three-dimensional relationships that are often decisive for surgical decision-making. These advanced modalities offer higher accuracy in identifying critical anatomic variables such as VSD location, routability, the relationship of great vessels, and the presence of outflow obstruction—features that influence the operative strategy and prognosis. Evidence demonstrates that prenatal and preoperative 3D echocardiography greatly surpasses 2D in predicting the feasibility of single-stage biventricular or multistage surgical repairs, enhancing counseling and decision-making for complex cases¹⁴.

Role of Transesophageal Echocardiography (TEE) for Complex Cases

Transesophageal echocardiography (TEE) serves a critical role in cases where complex anatomy or suboptimal transthoracic windows limit evaluation. TEE provides superior image resolution, especially for posterior structures and in perioperative settings, facilitating intraoperative assessment of residual lesions, AV valve anatomy, and surgical outcomes. TEE is particularly valuable in older children and adults, or during open heart surgery for confirmation of repairs, closure of VSDs, and detection of potential complications such as shunt persistence, valvular dysfunction, or arrhythmias¹⁵.

Despite its power, echocardiography in DORV is subject to practical constraints. The modality is highly dependent on operator expertise, with complex spatial anatomy demanding advanced skillsets and experience. Adequate acoustic windows are crucial and may be limited by patient size, age, or postoperative changes; obesity, chest deformities, and mechanical ventilation can further hinder image acquisition. 2D imaging may fail to capture the entirety of intricate anatomical relationships, while 3D and TEE are not always available in resource-constrained settings. There is also the risk of misidentifying structures, underestimating the extent of defects, or missing associated anomalies, thereby requiring complementary imaging modalities such as cardiac CT or MRI for confirmation in selected cases ¹⁶.

Role of Echocardiography in Preoperative Planning

The role of echocardiography in the preoperative planning of Double Outlet Right Ventricle (DORV) is pivotal, influencing diagnosis, surgical approach selection, risk stratification, and prognosis. Given the complex and heterogeneous nature of DORV, individualized assessment of cardiac morphology and function is essential to guide appropriate surgical intervention and optimize outcomes. Echocardiography, as a widely available, non-invasive imaging modality, allows detailed evaluation of multiple cardiac parameters—such as ventricular function, outflow tract anatomy, coronary artery anatomy, and septal defect characteristics—serving as a cornerstone in the multidisciplinary approach to this complex congenital heart disease.

Assessment of Ventricular Function

Accurate evaluation of ventricular function is integral to determining operability in DORV patients, especially in the context of biventricular versus univentricular repair strategies. Echocardiographic assessment of the left and right ventricles provides vital information regarding systolic and diastolic performance, chamber size, wall thickness, and signs of pressure or volume overload. Conventional two-dimensional (2D) echocardiography offers visualization of chamber geometry and wall motion abnormalities; M-mode imaging assists in quantifying ventricular wall thickness and excursion¹⁷. Quantitative measures such as ejection fraction (EF) and fractional shortening (FS) are typically calculated from 2D imaging, providing quantifiable indices of global systolic function.

Tissue Doppler imaging (TDI) and speckle-tracking echocardiography (STE) have further refined the evaluation of ventricular mechanics by enabling assessment of regional myocardial velocities, deformation, and strain patterns. These techniques uncover subtle myocardial dysfunction that might not be apparent on conventional imaging, which is crucial in complex congenital hearts where asynchronous contraction or remodeling occurs. In DORV, the right ventricle often sustains volume or pressure overload due to abnormal flow patterns, and precise functional assessment helps predict postoperative recovery capacity and long-term ventricular performance¹⁸. Diastolic function evaluation, although more challenging, is pertinent, as diastolic dysfunction may worsen pulmonary hypertension and impair surgical outcomes. Parameters such as mitral and tricuspid inflow velocities, pulmonary vein flow patterns, and annular tissue Doppler velocities contribute to a comprehensive diastolic assessment. Since chronic hypoxemia or heart failure can precipitate ventricular remodelling in DORV patients, on-going functional evaluation informs timing of surgery and perioperative management.

Pulmonary and Systemic Outflow Tract Evaluation

Thorough delineation of the pulmonary outflow tract (RVOT) and systemic outflow tract (LVOT or VSD to great arteries) is imperative in surgical planning for DORV. Echocardiography delineates the morphology, size, and patency of outflow tracts, presence and extent of obstruction, and flow dynamics. Two-dimensional imaging portrays the muscular and fibrous structures constituting these pathways, while color Doppler highlights velocity jets and flow disturbances indicative of stenosis or regurgitation¹⁹. Obstruction to the pulmonary outflow can occur at valvar, subvalvar (infundibular), or supravalvar levels, with subpulmonic stenosis being a common feature in certain DORV subtypes, particularly those resembling TOF physiology. Spectral Doppler velocity measurements quantify pressure gradients across the RVOT, providing estimates of obstruction severity, guiding decisions regarding surgical relief or conduit placement. Similarly, systemic outflow tract obstruction—either at the VSD or aortic valve level—is evaluated to anticipate potential sites of postoperative stenosis or residual shunting.

The anatomical relationship of the VSD to the great arteries defines the feasibility of intraventricular rerouting or baffle construction. Routability assessment determines whether the left ventricular blood can be effectively directed to the aorta or pulmonary artery through patch tunnel creation without obstruction or jeopardy to atrioventricular valves or conduction tissue. Additionally, assessment of the size and morphology of the great arteries informs whether direct arterial switch or more complex root translocation procedures are indicated.

Coronary Artery Anatomy Visualization

Visualization of coronary artery anatomy is critical in patients planned for arterial switch operations or complex anatomical corrections due to the necessity of reimplanting the coronary arteries in their new aortic root. Ectopic or anomalous coronary origins, single coronary arteries, intramural courses, or abnormal branching patterns significantly influence surgical planning and morbidity risk. Although transthoracic echocardiography can delineate major proximal coronary origins in infants and children, its sensitivity is limited by acoustic windows and operator experience.

Transesophageal echocardiography (TEE) offers improved visualization in older patients and intraoperatively but may still be insufficient to fully characterize coronary anatomy. Therefore, echocardiographic coronary imaging is often complemented by cardiac computed tomography angiography (CTA) or cardiac magnetic resonance imaging (MRI) for comprehensive anatomical detail²⁰. Nevertheless, echocardiography remains a frontline tool for initial coronary assessment due to its non-invasiveness and immediate availability, permitting rapid exclusion of gross coronary anomalies in most cases.

3. DEFINING THE SURGICAL APPROACH

Preoperative echocardiographic information directly informs the choice of surgical strategy in DORV, which includes options such as the Rastelli procedure, arterial switch operation, intraventricular rerouting (intraventricular tunnel repair), or palliation.

- Rastelli Procedure: This surgery is indicated when there is a subaortic VSD with pulmonary stenosis. After closing the VSD and diverting left ventricular flow to the aorta through the VSD baffle, right ventricular outflow is established via a conduit to the pulmonary artery, bypassing the stenosis. Echocardiographic identification of VSD size and relation to great arteries, degree and location of RVOT obstruction, and ventricular function are essential. Doppler assessment aids in quantifying gradients to determine the need for outflow tract reconstruction.
- Arterial Switch Operation: Typically applied to subpulmonic VSD types (e.g., Taussig-Bing anomaly), this involves
 translocating the aorta and pulmonary artery to their respective ventricles and reimplanting coronary arteries.
 Detailed assessment of coronary anatomy, great artery relationships, and VSD characteristics through
 echocardiography is critical for operative success.
- Intraventricular Rerouting: For some DORV subtypes with non-committed VSDs or where great arteries have abnormal positions, an intraventricular tunnel or baffle is constructed to direct left ventricular flow to the aorta or pulmonary artery. Routability, size of the left ventricle, and proximity to conduction tissue are echocardiographically assessed to ensure baffle feasibility without obstruction or valve interference.

• Single-Ventricle Palliation: In cases where biventricular repair is contraindicated due to ventricular hypoplasia or complex anatomy, the Fontan pathway is pursued. Echocardiography helps in evaluating ventricular size, AV valve competence, pulmonary artery anatomy, and pulmonary pressures to guide candidacy.

Echocardiography provides the dynamic anatomical and functional roadmap necessary for surgical planning. Advanced techniques such as 3D echocardiography and intraoperative TEE enhance visualization of complex anatomic details, enabling preoperative team discussions, individualized surgical planning, and real-time intraoperative guidance. The reliability and completeness of preoperative echocardiographic evaluation significantly reduce intraoperative surprises and improve clinical outcomes²¹.

Comparison with Other Imaging Modalities

Echocardiography is the primary and indispensable imaging modality used in the diagnosis and management of Double Outlet Right Ventricle (DORV) due to its accessibility, real-time imaging capability, and functional assessment. However, it is complemented by advanced imaging modalities such as cardiac magnetic resonance imaging (MRI) and computed tomography (CT) angiography, (Table 1) each offering unique strengths that enhance spatial understanding and detailed vascular anatomy critical to surgical planning and follow-up.

Role of Cardiac MRI in Spatial Evaluation

Cardiac MRI provides comprehensive and detailed spatial and functional cardiac assessment in DORV. It offers high-contrast resolution imaging without ionizing radiation, making it especially suitable for serial evaluations in pediatric patients. MRI allows accurate quantification of ventricular volumes, ejection fraction, myocardial mass, and flow measurements, essential for assessing ventricular function and shunt volumes in complex congenital malformations²². Despite these advantages, MRI has limitations in evaluating fine anatomical details due to relatively lower spatial resolution compared to CT, particularly when assessing small structures such as ventricular septal defects or coronary artery origins. Its use is also constrained by longer scan times requiring sedation in young children, limited availability, and contraindications in patients with some implanted devices. Nonetheless, MRI excels in demonstrating extracardiac vascular structures, flow dynamics in great vessels, pulmonary veins, and systemic veins, as well as myocardial characterization, which are pivotal in comprehensive management of DORV patients. The comparison of imaging modalities in double outlet right ventricle are listed below in **Table-1**.

Echocardiography	Cardiac MRI	CT Angiography		
First-line, bedside, real-time	High-contrast, functional, no radiation	High spatial resolution, fast, coronary & vessel anatomy		
Limited acoustic windows	Lower spatial resolution, sedation needed	Radiation + contrast exposure		
Best for valves, septal defects, flow	Best for ventricular volumes & extracardiac flow	Best for coronary arteries, branching, arch anomalies		

Table 1: Comparison of Imaging Modalities in Double Outlet Right Ventricle.

Role of CT Angiography in Vessel Anatomy

Computed tomography angiography (CTA) plays a pivotal role in the detailed visualization of great vessel anatomy, coronary artery origins, and extracardiac vascular structures with high spatial and temporal resolution. CT is particularly valuable in cases where echocardiographic acoustic windows are limited, such as older children, patients with postoperative anatomical alterations, or those with metallic implants that degrade MRI image quality. CTA enables rapid acquisition of 3D volumetric datasets that can be reconstructed in multiple planes and formats, allowing precise evaluation of coronary artery courses, pulmonary artery branching, aortic arch anomalies, and associated vascular malformations. This anatomical clarity is critical for surgical decision-making and can identify unexpected anatomical variants that may influence operative risk. Advancements like ECG-gated CT scanning further enhance visualization while minimizing arterial motion artifacts, enabling more confidence in preoperative planning. However, CT involves ionizing radiation and contrast exposure, necessitating prudent use and pediatric dose optimization²³.

Complementary Role of Echocardiography vs Advanced Modalities

While echocardiography remains the cornerstone of DORV imaging due to its ability to dynamically assess intracardiac anatomy, function, and flow in a radiation-free and bedside-available manner, advanced imaging modalities serve complementary purposes that extend and refine diagnostic capabilities Echocardiography provides superior temporal resolution and adjudicates valve function, septal defects, flow patterns, and ventricular performance in real-time, which are essential for initial diagnosis and perioperative monitoring. However, its dependence on operator skill and patient-specific

acoustic windows can limit visualization of certain structures, including extracardiac vessels, coronary artery origins, and complex spatial relationships between great vessels²⁴. Cardiac MRI compensates for these limitations by quantifying volumes, flows, and myocardial characteristics with excellent tissue contrast and without radiation, ideal for detailed functional assessment and follow-up. CT angiography excels in spatial and vascular detail, overcoming acoustic window challenges to reveal coronary anatomy and extracardiac vascular malformations, crucial for comprehensive surgical planning and postoperative evaluation. Thus, an integrated imaging approach leveraging the strengths of echocardiography, cardiac MRI, and CT angiography provides the most thorough evaluation of DORV. Echocardiography forms the first-line and continuous monitoring tool due to its availability, functional assessment, and immediate clinical relevance. MRI and CT serve as adjuncts offering high-resolution anatomical detail and volumetric quantification that guide complex surgical decisions especially in preoperative settings and longitudinal care.

Case Studies and Clinical Applications

Double Outlet Right Ventricle (DORV) encompasses a spectrum of complex congenital cardiac anomalies where both great arteries arise largely from the morphologic right ventricle. The assessment of DORV by echocardiography is indispensable in clinical practice, serving as the primary diagnostic tool, guiding therapeutic approaches, and enabling postoperative monitoring. However, given the anatomical variability and hemodynamic complexities, echocardiographic diagnosis may present multiple challenges, requiring integration with complementary modalities such as cardiac catheterization and magnetic resonance imaging (MRI) for comprehensive evaluation. This section provides a detailed exploration of illustrative echocardiographic findings, real-world diagnostic challenges with their solutions, and the correlation of echocardiographic data with catheterization and MRI findings through clinical case studies, underscored by evidence from PubMed-indexed

Illustrative Echocardiographic Images and Findings

Echocardiographic examination of DORV patients reveals distinctive structural and functional features critical for diagnosis. The key hallmark is the visual confirmation of both arterial trunks arising from the right ventricle using standard 2D imaging views, such as the parasternal long and short axis, subcostal, and apical windows. Visualization of the ventricular septal defect (VSD), its location (subaortic, subpulmonic, doubly committed, or noncommitted), and size are paramount, influencing surgical planning. Color Doppler imaging elucidates abnormal flow patterns, including bidirectional shunting across the VSD and velocity gradients indicative of outflow tract obstruction. Spectral Doppler assessment quantifies gradients, vital for assessing pulmonary stenosis or systemic outflow obstruction²⁵. Echocardiographic depiction of valve relationships, including overriding and straddling atrioventricular valves, provides insights into the complexity of anomalies present. Three-dimensional echocardiography enhances spatial understanding, especially in delineating baffle pathways and the relation between the VSD and great arteries. Transesophageal echocardiography (TEE) offers superior resolution in older children and intraoperative settings, aiding in clarifying ambiguous transthoracic evaluations and guiding surgical revisions.

Real-World Diagnostic Challenges and Solutions

In clinical practice, DORV diagnosis via echocardiography is frequently challenged by patient-specific anatomical variants, acoustic window limitations, and the complexity of associated anomalies. For example, differentiating DORV from transposition of the great arteries (TGA) or large VSD with overriding aorta may be difficult due to overlapping morphologies; subtle delineation of the exact ventricular-arterial connections requires high operator expertise and multiplane imaging. In some cases, echocardiographic assessment underestimates the presence of infundibular obstruction, misclassifies VSD location, or fails to fully image coronary artery anatomy, potentially leading to suboptimal surgical planning²⁶. Solutions entail multimodal imaging integration; supplemental cardiac MRI and catheter angiography offer complementary anatomic details difficult to acquire by echocardiography alone. Advances including 3D echocardiography and speckle tracking improve diagnostic accuracy for complex flow dynamics and myocardial function. Intraoperative TEE confirms procedural success and detects residual lesions. Training and multidisciplinary collaboration enhance diagnostic precision, fostering improved patient outcomes.

Correlation with Cardiac Catheterization and MRI Findings

Cardiac catheterization historically provided definitive structural and hemodynamic data but is increasingly reserved for therapeutic interventions due to non-invasive imaging advances. Nonetheless, catheterization complements echocardiography by enabling direct pressure measurement, oxygen saturation analysis, and angiographic visualization of vascular anatomy and collaterals. It remains invaluable when non-invasive imaging is inconclusive or when evaluating pulmonary vascular resistance, critical in preoperative risk assessment. Cardiac MRI has emerged as a powerful adjunct modality offering superior spatial resolution and multiplanar imaging without radiation exposure. It excels in quantifying ventricular volumes, function, flow volumes, and shunt fractions, with precise delineation of outflow tract anatomy and great vessel relationships. MRI's ability to visualize extracardiac vessels and myocardial tissue characterization complements echocardiographic findings, especially in complex DORV variants with multiple concomitant anomalies²⁷. MRI may detect features missed by echocardiography, such as ventricular trabeculation patterns, subtle atrioventricular valve abnormalities,

and myocardial fibrosis, thereby influencing prognosis and management. Various case examples in DORV imaging are listed below in **Table-2**.

S.N	Patient Age	Location of VSD	Key Echocardiogra phic Finding	Associated Anomaly	MRI Findings	Catheterization Role	Ref
1	28 yr (adult)	Perimembr anous	Bidirectional shunt, overriding aorta	Pulmonary hypertension, pericardial effusion	Confirmed anatomy, functional status	Hemodynamic assessment	(28)
2	Pediatric	Subaortic	Muscular outflow tract obstruction	Coronary artery anomalies	Detailed vessel and VSD anatomy	Preoperative pressure gradients	(29)
3	Mixed ages	Doubly committed	Overriding aortic valve	Ventricular hypertrophy	VSD position and valve relations	Oxygen saturation measurement	(30)
4	Adult	Noncommi tted	Complex intracardiac rerouting	Valve straddling	Ventricular volume quantification	Right ventricle pressure	(31)
5	Pediatric	Subpulmo nic	Great vessel spatial relationship	Outflow tract stenosis	3D spatial evaluation	Pulmonary artery pressure	(32)
6	Pediatric	Subaortic	VSD size and location	Pulmonary stenosis	Flow quantification	Pre-surgical angiography	(33)
7	Adult	Remote VSD	Flow jet on Doppler	Atrioventricular valve abnormalities	Confirmed ventricular anatomy	Intracardiac pressure recording	(34)
8	Fetal to neonate	Subpulmo nic	3D assessment of VSD	Pulmonary regurgitation	Virtual surgical planning	N/A	(35)
9	Pediatric	Doubly committed	Color Doppler flow patterns	Coronary artery variants	Coronary artery imaging	Preoperative angiography	(36)

Table 2: Case Examples in DORV Imaging

4. ADVANCES AND FUTURE DIRECTIONS

The field of echocardiographic assessment of Double Outlet Right Ventricle (DORV) and other complex congenital heart diseases is rapidly evolving, driven by technological innovations that promise to enhance diagnostic precision, surgical planning, and personalized patient care. Key advances in artificial intelligence (AI), three-dimensional (3D) printing, and hybrid imaging integration are shaping the future landscape of congenital cardiac imaging.

Artificial intelligence and machine learning applications are increasingly being deployed to automate and standardize echocardiographic quantification, addressing long-standing challenges of operator dependence and interobserver variability. AI algorithms trained on large datasets can automatically identify cardiac chambers, segment ventricular borders, and quantify volumes, ejection fraction, and strain parameters with high accuracy and reproducibility. Automated analysis accelerates workflow and improves detection of subtle ventricular dysfunction, which is critical in DORV patients where nuanced myocardial performance influences surgical risk and timing. Moreover, AI-enabled pattern recognition may assist in early anomaly detection from fetal echocardiograms, providing opportunities for prenatal diagnosis and intervention planning. The integration of AI with echocardiographic hardware and software platforms portends a future where real-time, precise, and comprehensive cardiac assessment is accessible globally, including resource-limited settings. Three-dimensional printing technology, leveraging echo-derived anatomical models, is becoming a transformative tool in surgical planning for DORV. High-quality 3D reconstructions of intracardiac anatomy from 3D echocardiography or fused multimodal imaging

are used to create patient-specific physical heart models. These tangible replicas enable surgeons to visualize complex spatial relationships, practice surgical maneuvers, and refine repair strategies before entering the operating room³⁷. By facilitating a tactile understanding of cardiac geometry particularly the ventricular septal defect location, ventricular size, and great artery positions 3D printing improves operative precision, reduces procedure times, and enhances outcomes. Hybrid use of 3D printing along with virtual reality and simulation-based surgical rehearsal represents an exciting frontier in congenital heart disease management.

The evolution toward hybrid imaging approaches integrating echocardiography with complementary modalities such as cardiac MRI, CT angiography, and catheter-based mapping offers a comprehensive and holistic evaluation platform. Hybrid imaging combines functional, anatomical, and flow-related information in a synergistic manner, overcoming the limitations intrinsic to single modalities. Advances in image fusion techniques enable simultaneous visualization of echocardiographic data co-registered with MRI or CT datasets, providing multimodal insights into myocardial tissue characterization, coronary anatomy, vascular anomalies, and complex flow dynamics characteristic of DORV. Such integration enhances diagnostic confidence, informs multidisciplinary decision-making, and personalizes surgical strategies. As hybrid platforms mature, they will likely facilitate more precise interventions, intraoperative guidance, and postoperative surveillance with improved patient safety and reduced procedural times.

Challenges and Limitations

Echocardiography in neonates and infants, especially for complex congenital heart diseases such as Double Outlet Right Ventricle (DORV), presents several challenges and limitations intrinsic to patient factors, technical constraints, and operator expertise.

Technical Pitfalls in Neonates and Infants

The small size and high heart rates of neonates and infants significantly complicate image acquisition and interpretation. Their diminutive thoracic structures limit acoustic windows, especially when compounded by lung hyperinflation, chest wall deformities, or postoperative changes. The rapid heart rates necessitate high frame rates and excellent temporal resolution to capture cardiac cycle dynamics accurately; failure to meet these demands can lead to motion artifacts and blurred images. Obtaining standard echocardiographic views like parasternal and suprasternal views requires meticulous positioning and often alternative, non-standard windows such as subcostal or right parasternal to optimize visualization. Furthermore, neonates may be uncooperative or restless, creating motion artifacts that reduce image quality. Sedation is cautiously used due to potential respiratory depression risks. The parturient and critically ill status of some neonatal patients limits the time available for comprehensive imaging, often necessitating focused, rapid assessments with trade-offs in detail. Additionally, overlapping hemodynamic features, such as concomitant pulmonary hypertension and outflow tract obstruction, require careful Doppler interrogation and interpretation to avoid misdiagnosis³⁸. These technical demands necessitate highend echocardiographic equipment optimized for pediatric imaging and advanced operator skill.

Operator Dependency and Variability in Interpretation

Echocardiographic interpretation in pediatric cardiology, including DORV assessment, is heavily operator-dependent. Variability arises due to differences in sonographer experience, image acquisition technique, and interpreter expertise. Complex and rare congenital anomalies may be unfamiliar to many practitioners, increasing the risk of missed or incorrect diagnoses. Subtle malformations, such as small or atypically located ventricular septal defects or valve malformations, may be overlooked or mischaracterized. Inter-observer variability can affect measurements of ventricular dimensions, flow gradients, and defect sizes, influencing clinical decisions. Moreover, the heterogeneity of congenital heart disease presentations necessitates a systematic, segmental approach for consistent interpretation³⁹. Diagnostic errors or incomplete studies are not uncommon, underscoring the need for institutional quality improvement processes, case reviews, and multidisciplinary case discussions to mitigate variability and enhance accuracy in diagnosis and reporting. Standardized protocols and robust training reduce variability but do not eliminate the intrinsic subjectivity of image interpretation.

Training and Expertise Requirements in Pediatric Echocardiography

Expertise in pediatric and congenital echocardiography demands specialized training beyond that of adult cardiology. Proficiency requires understanding of pediatric cardiac anatomy and physiology, knowledge of congenital malformations, and familiarity with pediatric imaging techniques adapted for small patient size and rapid heart rates. Sonographers and interpreting physicians must be skilled in using high-frequency transducers, selecting appropriate imaging windows, and performing advanced Doppler and 3D imaging techniques. Continued education and hands-on experience are critical, as is exposure to diverse congenital pathologies, which may be rare or complex⁴⁰. Professional societies recommend regular competency assessments, quality improvement initiatives, and peer reviews in echocardiography laboratories to maintain high standards. The availability of advanced imaging modalities, such as 3D echocardiography and strain imaging, increases the complexity of sonographer training but also enhances diagnostic capability when appropriately utilized. In resource-limited settings, lack of specialized pediatric equipment and trained personnel remains a barrier to optimal

diagnosis and management, emphasizing the need for telemedicine, remote mentorship, and technology dissemination.

5. CONCLUSION

Echocardiography remains the cornerstone in the diagnosis, evaluation, and management of Double Outlet Right Ventricle (DORV) and other complex congenital heart diseases. Its non-invasive nature, real-time imaging capability, and comprehensive functional assessment provide invaluable insights into intracardiac anatomy, ventricular performance, and hemodynamics that are essential for accurate diagnosis and individualized surgical planning. Despite inherent challenges such as technical limitations in neonates and operator dependency, advances in imaging technology continue to enhance its diagnostic precision and clinical utility. Looking toward the future, the integration of artificial intelligence (AI) and machine learning into pediatric echocardiography promises to revolutionize image acquisition, interpretation, and quantification, mitigating operator variability and elevating diagnostic consistency. The advent of three-dimensional (3D) printing based on echo-derived models offers transformative opportunities for surgical rehearsal and personalized intervention planning, thereby improving surgical outcomes. Furthermore, evolving hybrid imaging modalities that fuse echocardiography with complementary techniques such as cardiac MRI and computed tomography enable holistic cardiac evaluation, overcoming individual modality limitations and supporting comprehensive clinical decision-making.

As pediatric echocardiographic practice expands with continuous technological innovation and standardization efforts, the synergy of expertise, advanced imaging, and AI-assisted tools will collectively underpin precision medicine approaches in congenital cardiology. This fosters improved patient care pathways, early diagnoses, risk stratification, and long-term management strategies that hold the promise of significantly better outcomes for patients with DORV and wider congenital heart disease populations. The trajectory of pediatric and congenital echocardiography is one of ongoing evolution toward more automated, integrative, and patient-specific imaging paradigms, underscoring the modality's critical and enduring role in paediatric cardiovascular medicine.

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