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Evaluation of Right and Left Ventricular Function in Ebstein Anomaly Using RVGLS and LVGLS: A Narrative Literature Review

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ABSTRACT

Ebstein anomaly is a rare congenital heart defect characterized by tricuspid valve malformation, atrialization of the RV, and hemodynamic disturbances that may affect both ventricles. Assessing ventricular function in this condition is essential because it is closely related to disease progression, clinical symptoms, and long term prognosis. Conventional parameters such as TAPSE, RVFAC, and LVEF are commonly used but have limitations. Speckle Tracking Echocardiography (STE), through Right Ventricular Global Longitudinal Strain (RVGLS) and Left Ventricular Global Longitudinal Strain (LVGLS), provides higher sensitivity for detecting ventricular dysfunction. This narrative literature review evaluates the role of RVGLS and LVGLS in patients with Ebstein anomaly. Evidence shows that RVGLS is more accurate in identifying RV dysfunction compared with TAPSE and RVFAC, and possesses strong prognostic value for mortality. LVGLS can detect LV dysfunction at a subclinical stage even when LVEF remains preserved, reflecting involvement of longitudinal myocardial fibers due to ventricular interdependence and secondary remodeling. After tricuspid valve surgery, both RVGLS and LVGLS remain sensitive indicators of changes in ventricular performance. Overall, RVGLS and LVGLS demonstrate superiority over conventional systolic parameters and are recommended as primary tools for evaluating biventricular function in patients with Ebstein anomaly.

Keywords: Ebstein anomaly**,** RVGLS, LVGLS, Left ventricular dysfunction, Right ventricular dysfunction.

INTRODUCTION

Ebstein Anomaly is a rare congenital heart defect characterized by a tricuspid valve positioned abnormally toward the right ventricle (RV), resulting in a smaller RV and causing atrialization of the RV. This abnormality has a relatively low prevalence, accounting less than 1% of all congenital heart disease cases (Holst et al., 2019; Singh et al., 2025).

In Ebstein Anomaly, the tricuspid valve undergoes displacement of the posterior and septal leaflets, abnormalities of the anterior leaflet, and dilation of the tricuspid valve annulus. These abnormalities result in tricuspid valve regurgitation, which over time leads to RV dilatation due to volume overload (Jost et al., 2007). The more severe the degree of tricuspid regurgitation, the larger the functional RV becomes (Fratz et al., 2013). This can ultimately cause RV dysfunction. Because the right and the Left Ventricle (LV) share the interventricular

septum and pericardium structures, structural or pressure changes in the RV may affect the LV through structural and hemodynamic interactions, eventually leading to LV dysfunction (Egbe et al., 2021a).

The cardiac ventricles play a role in circulating blood throughout the body and the lungs. Evaluation of RV and LV function is important because it can help to determine hemodynamics, progression, and prognosis of patients with Ebstein Anomaly. RV function can be assessed using Tricuspid Annular Plane Systolic Excursion (TAPSE) and Right Ventricular Fractional Area Change (RVFAC), whereas LV function can be evaluated using Left Ventricular Ejection Fraction (LVEF). These parameters represent conventional systolic indices of LV and RV function. However, among these parameters, there are functional measures derived from Speckle Tracking Echocardiography (STE) namely Right Ventricular Global Longitudinal Strain (RVGLS) and Left Ventricular Global Longitudinal Strain (LVGLS) which are more sensitive and superior in detecting dysfunction of ventricle (Egbe et al., 2023; Mihos et al., 2025). This narrative literature review will focus on ventricular functional parameters using STE in patients with Ebstein anomaly.

RESEARCH METHODS

The aim of this narrative literature review is to discuss RV and LV function in patients with Ebstein anomaly using Strain STE. This narrative literature review highlights both ventricular functional parameters in assesing the dysfunction of the ventricles on Ebstein anomaly.

Literature Search Strategy

This narrative literature search was conducted using studies published between January 2019 and November 2025 written in English. The literature search was conducted using Pubmed and Scopus. The search strategy focused on keywords such as “Ebstein Anomaly”, “RVGLS”, “LVGLS”, “right ventricular dysfunction”, “left ventricular dysfunction”, and “Speckle Tracking Echocardiography”. The Boolean keywords used were (“Ebstein anomaly” OR “Ebstein’s anomaly”) AND (“strain” OR “speckle tracking” OR “RVGLS” OR “LVGLS”).

Inclusion and Exclusion Criteria

Inclusion Criteria:

- Studies discussing RV and/or LV function in patients with Ebstein anomaly.
- Studies that utilized echocardiographic assessment with RVGLS and/or LVGLS

parameters in Ebstein anomaly.

- Retrospective study of Ebstein anomaly patients.

Exclusion Criteria:

- Studies that did not include any echocardiographic parameters of STE in the evaluation of Ebstein anomaly.
- Studies that do not involve Ebstein anomaly.
- Studies that examine other congenital heart diseases.
- Studies that used Cardiovascular Magnetic Resonance-feature Tracking (CMR-FT) instead of STE.

Data Extraction and Analysis

Data from the collected studies will be extracted regarding RV and LV function parameters in patients with Ebstein anomaly. All data were analyzed based on age groups (children, adolescents, adults) and comparisons between pre- and post-Tricuspid Valve Surgery (TVR). For RV function, RVGLS was extracted as the primary parameter due to its superior diagnostic sensitivity and prognostic value compared with TAPSE and RVFAC, including its ability to detect subclinical dysfunction and postoperative changes after tricuspid valve repair. For LV function, LVGLS was extracted to identify early dysfunction, evaluate long-term outcomes, and distinguish occult from overt LV impairment, particularly in patients with preserved LVEF. Biventricular interaction was assessed through the relationship between RVGLS and LVGLS to evaluate ventricular interdependence. All extracted data were evaluated using descriptive analysis.

RESULTS AND DISCUSSION

1. Parameter of Right Ventricular Function in Ebstein Anomaly

RV dysfunction of Ebstein Anomaly occurs as a result of primary tricuspid valve lesion. A greater degree of tricuspid severity can lead to ventricular dilatation (Fratz et al., 2013). Evaluation of RV can be assessed using Tricuspid Annular Plane Systolic Excursion (TAPSE), RVFAC (Right Ventricular Fractional Area Change), and Right Ventricular Global Longitudinal Strain (RVGLS).

1.1 RVGLS as Predictor and Prognostic Parameter

In adult patients with Ebstein anomaly, RV dysfunction was defined as RVFAC<35%, TAPSE<16mm, and RVGLS≥-18%. TAPSE and RVFAC were available in 630 patients, whereas RVGLS was available only in 620 patients. TAPSE parameter was able to detect 19% of RV dysfunction, whereas RVFAC performed better, detecting 53% of RV dysfunction. On the other hand, the RVGLS parameter derived from Speckle Tracking Echocardiography was able to detect 51% RV dysfunction. RVGLS and RVFAC therefore showed nearly similar detection rates for RV dysfunction in Ebstein anomaly. However, among all these parameters, RVGLS is the most accurate predictor of mortality and provides superior prognostic parameter (Egbe et al., 2023).

1.2 RVGLS for Detection of Subclinical Dysfunction

In pediatric and young adult Ebstein anomaly patients, the mean RVFAC was 33.1±9.5%, and TAPSE was 20±17 mm. These conventional parameters remained within normal ranges in children and young adults with Ebstein Anomaly. In contrast, the mean RVGLS value was -13.3±7.3%, indicating an early decline in RV function. RVGLS reflects longitudinal contraction; therefore, impairment of longitudinal fiber contraction leads to reduced RVGLS. RVFAC represents both longitudinal and circumferential RV function. In this study, RVFAC remained preserved due to compensatory mechanisms through increased circumferential deformation. A decrease in RVFAC indicates global RV dysfunction. RVGLS can detect dysfunction earlier and more subtly compared with RVFAC (Prota et al., 2019).

1.3 RVGLS on Pre- and Post-TVR

In patients with Ebstein anomaly undergoing TVR, the baseline RVGLS before surgery averaged -19.4±5.6%. One year after TVR, RVGLS decreased to -12.9±5.0%, but improved again at years 2 and 5. This pattern indicated that RV dysfunction declines after TVR. An ventricular myopathy of the right ventricle had already developed, leading to chronic structural changes. RVGLS effectively evaluates right ventricle myocardial contractility both before and after TVR (Ricci et al., 2020). Early and more accurate assessment of RV function has the potential to help identify patients at high risk of developing reccurrent severe TR. In postoperative patients with Ebstein anomaly, those with Carpentier type C-D demonstrated lower RVGLS values -10,8±4.4% compared with Carpentier type A-B -17.9±5.4%. However, at the one-year follow-up, this difference was no longer significant. Surgical correction caused

a temporary decline in RV function, which was sensitively detected by RVGLS (Meng et al., 2024).

2. Parameter of Left Ventricular Function in Ebstein Anomaly

LV dysfunction in Ebstein Anomaly does not occur due to primary structural abnormalities of the LV, but rather as a result of ventricular interdependence, secondary cardiomyopathy, and LV remodeling. Consequently, the LV may experience impaired function or dysfunction. LVEF is a conventional parameter commonly used in echocardiographic assessment; however, LVGLS derived from STE is considered a superior parameter (Mihos et al., 2025).

2.1 LVGLS as A Sensitive Parameter for Early LV Dysfunction

In pediatric and young adult Ebstein anomaly patients, LVEF values were preserved across all patient groups; however, LVGLS was already reduced. In the stable-disease group, the mean LVGLS was -17.5±4%, whereas in the progressive-disease group, LVGLS declined more markedly to -14.5±3.9%. The longitudinal subendocardial myocardial fibers are the earliest to be affected, leading to a decrease in LVGLS before LVEF despite preserved LVEF values. This confirms that LVGLS is more sensitive than LVEF in detecting LV dysfunction across all patient groups (Prota et al., 2019).

2.2 LVGLS and Long-Term Clinical Outcomes

In 673 adult patients with Ebstein anomaly, 10% had Low-LVGLS/Low-LVEF, 13% had Low-LVGLS/Normal-LVEF, 1% had Normal-LVGLS/Low-LVEF, and 76% had Normal-LVGLS/Normal-LVEF. The Low-LVGLS/Low-LVEF group represents overt LV systolic dysfunction and demonstrated a hemodynamic profile similar to the Low-LVGLS/Normal-LVEF group, although with more severe disturbances, including reduced GFR, elevated NT-proBNP, decreased RVGLS, and clinical symptoms of heart failure. This study emphasizes that LVGLS can detect progressive systolic dysfunction, characterized by a decline in LVGLS over time despite stable LVEF values. A decrease in LVGLS is sufficient to reflect declining left heart function. Patients with reduced LVGLS were at increased risk of developing end-stage heart failure leading to death or heart transplantation during the 10-year follow-up period (Egbe et al., 2021b).

2.3 LVGLS on Pre- and Post-TVR

In Ebstein anomaly patients undergoing TVR, LVGLS can differentiate two types of LV systolic dysfunction. Occult LV dysfunction (early-stage dysfunction), as measured by LVGLS, carries a mortality risk comparable to overt LV dysfunction (advanced-stage dysfunction), even though both groups still demonstrated normal LVEF values. LVEF was considered less accurate in detecting dysfunction in this study. These findings support LVGLS as the primary parameter for identifying LV dysfunction (Egbe et al., 2021a).

In the group of Ebstein anomaly patients undergoing TVR, the preoperative LVGLS was -15±4.8%, indicating reduced or impaired LVGLS. In this study, LVGLS was found to be lower than RVGLS. During the first year after TVR, LVGLS declined further, but by the second and fifth years, LVGLS values approached the preoperative levels, whereas LVEF remained normal both before and after surgery. Therefore, LVGLS can detect hemodynamic impact and ventricular interaction more effectively than LVEF (Ricci et al., 2020).

3. Biventricular Interaction Using RVGLS and LVGLS

There is an association between RVGLS and LVGLS in Ebstein Anomaly, where worsening RVGLS corresponds to worsening LVGLS. This occurs because both ventricle share the interventricular septum. As the RV undergoes primary lesions and hemodynamic disturbances, a septal shift toward the left ventricle develops, reducing LV filling and consequently impairing longitudinal contraction of the ventricle (Egbe et al., 2021a).

CONCLUSION AND RECOMENDATION

RVGLS and LVGLS are reliable systolic STE parameters and are more effective than conventional systolic parameters. In the baseline evaluation of patients with Ebstein anomaly, in addition to conventional parameters such as TAPSE, RVFAC, and LVEF, routine assessment of RVGLS and LVGLS are recommended to obtain a more sensitive evaluation of ventricular function. RVGLS can predict mortality and has strong prognostic value. It is capable of detecting early RV dysfunction and is highly effective in assessing RV contractility. LVGLS is also highly sensitive in identifying the progression of LV dysfunction thus serving as a more sensitive parameter of disease progression and long term clinical risk. In patients who have undergone TVR, follow-up using strain parameters is crucial for monitoring biventricular function and enabling early detection of dysfunction that may not be apparent through conventional measures. Furthermore, in patients whose symptoms worsen despite

having normal LVEF, TAPSE, and RVFAC parameters, a decline in LVGLS or RVGLS may serve as an indicator of occult ventricular dysfunction, necessitating further evaluation and closer follow-up. Overall, strain based parameters provide a more comprehensive assessment of ventricular function in patients with Ebstein anomaly. These two parameters are expected to complement conventional parameters and are recommended as primary parameters for evaluating RV and LV function in patients with Ebstein anomaly. Future research should aim to establish strain-specific cut-off values for Ebstein anomaly and conduct prospective multicenter studies with vendor-standardized strain acquisition to enhance the accuracy of ventricular function assessment.

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