The role of 3-dimensional echocardiography in evaluating congenital heart diseases

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ABSTRACT

الأهداف: تقييم مدى فاعلية استخدام تخطيط صدى القلب الصدري ثلاثي الأبعاد ذو الوقت الفعلي في توفير معلومات أدق و أوضح في وصف دقيق لأمراض القلب الوراثية مما يوفره استخدام تخطيط صدى القلب الصدري ثنائي الابعاد لوحده في تشخيص امراض القلب الوراثية.

الطريقة: أجريت هذه الدراسة المقطعية في مركز الأمير سلطان لمعالجة امراض و جراحة القلب في الرياض، المملكة العربية السعودية، وذلك خلال الفترة من يناير إلى يونيو 2009م. حيث قمنا بإجراء تخطيط صدى القلب ثنائي الابعاد لحوالي 50 مريض مصاب بتشوهات القلب الوراثية (مجموعة الدراسة) ثم بعد ذلك قمنا بإجراء تخطيط صدى القلب ثلاثي الابعاد لنفس المرضى مع زيادة عشرة دقائق فقط لإجمالي الوقت اللازم لإجراء تخطيط صدى القلب ثنائي الابعاد باستخدام برامج الأشعة الصوتية الحديثة.

النتائج: اشتملت الدرسة على 29 (58%) ذكر و 21 (42%) أنثى تراوحت أعمارهم من شهر حتى 17 عام. كما أظهر تخطيط صدى القلب ثنائي الأبعاد 12 (24%) تشوهات خلقية في الجانب الأيمن، 18 (36%) تشوهات خلقية في الجانب الأيسر من القلب، 12 (24%) تشوهات في الحاجز بين الأذينين والبطينين، وحالة واحدة (2%) تشوه في الحاجز الأبهر الرئوي، 4 (8%) تشوهات قلب وراثية معقدة، و3 (6%) كان فيها حجم البطين الأيسر يبدو صغيراً. وعند مقارنة تخطيط صدى القلب الثنائي الأبعاد أثبت استخدام تخطيط صدى القلب ثلاثي الأبعاد فعاليته في تخطيط صدى القلب في تشخيص تشوهات القلب الوراثية حيث صنف A في 25 حالة، و B في 24 حالة، و C في حالة واحدة.

خاتمة: أظهرت الدراسة مدى أهمية استخدام تقنية تخطيط صدى القلب ثلاثي الأبعاد في توفير معلومات إضافية هامة، دقيقة وتفصيلية لتشخيص تشوهات القلب الخلقية لم تكن توفرها استخدام تقنية تخطيط صدى القلب ثنائي الأبعاد لوحدها.

Objectives: To assess the feasibility and efficacy of the real-time transthoracic 3-dimensional echocardiography (RTT-3DE) technique in providing more detailed information compared to 2-dimensional echocardiography (2DE) in patients with structural heart disease, and to explore its application in routine clinical use.

Methods: This cross sectional study was carried out at the Prince Sultan Cardiac Center, Riyadh, Kingdom of Saudi Arabia from January to June 2009. Patients with congenital heart disease (CHD) were evaluated by conventional 2DE followed by RTT-3DE using dedicated software and a standard protocol. The 3DE studies were graded as: A - new finding not on 2DE; B - useful anatomic perspective; C - equivalent to 2DE; or D - missed 2DE findings.

Results: Fifty patients, 29 (58%) males and 21 (42%) females with age range from one month to 17 years compose the study group. The 2DE showed 12 (24%) right heart and 18 (36%) left heart lesions, 12 (24%) septal defects, one (2%) aortopulmonary window, 4 (8%) complex CHD, and 3 (6%) borderline left ventricular volume. When compared with the 2DE studies, 3DE studies were graded A in 25, B in 24, and C in one case.

Conclusion: The results in this study show that the RTT-3DE technique is feasible and very effective in providing more detailed information compared with 2DE in structural heart disease evaluation.

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Two-dimensional transthoracic echocardiography (2D-TTE) is the most common diagnostic study used in pediatric cardiology. This technique requires excessive mental exercise to perceive the actually obtained 2-dimensional echocardiography (2DE) pictures of a 3-dimensional (3D) structure, that is the heart, into an imaginary 3-dimension reconstruction.[1,2] Real-time transthoracic 3D echocardiography (RTT-3DE) has eliminated the need for this imaginary reconstruction, and permits a real-time assessment of cardiac and intracardiac anatomical complexities and physiological details.[3] The early 1990's was the era of primitive versions of RTT-3DE, but due to several limitations of long post processing time for offline reconstruction, analysis and sub-optimal image quality it could not gain much popularity in the clinical field,[4] thus rendering its use very limited in daily practice. Since 2002, when it became commercially available, there has been considerable advancement in the production of matrix-array transducers and software of 3DE, which has led to better evaluation of congenital heart disease (CHD) in pediatrics and adult population.[5] Volumetric scanning performed by RTT-3DE is a real-time technique compared to magnetic resonance imaging (MRI) and CT, which requires tremendous off-line reconstruction following 3D data acquisition.[2] This study attempts to report our initial experience with RTT-3DE imaging of CHD, and to validate the potential value of RTT-3DE findings when compared with 2D-TTE examinations, and the user friendly nature of 3DE in daily routine practice at Prince Sultan Cardiac Center (PSCC) in Riyadh, Kingdom of Saudi Arabia (KSA).

Methods. This cross sectional study was carried out at the Prince Sultan Cardiac Center, Riyadh, KSA from January to June 2009. Fifty patients with CHD referred from the Primary Care Clinics of the hospital and periphery were incorporated in this study. They had complete 2D-TTE studies followed by RTT-3DE. The age of the study group ranged from one month to 17 years. Those patients with various forms of CHD diagnosed by 2D-TTE, and had clear 2 DE images were included in this study. Exclusion criteria were: adult patient (>18 years), patients with acquired heart diseases and patients with poor 2D-TTE views. The ethical approval was obtained after submitting the proposal to the Institutional Review Board of the Hospital according to Helsinki Declaration. Off-line analysis was carried out using the machine's software (QLAB Advanced Quantification software, Philips Ultrasound, USA). All subjects were examined using IE-33 echo machine (Philips, Bothell, WA, USA) complete 2D-TTE studies were performed by experienced operator in the left lateral and supine positions for all patients followed by RTT-3DE, with additional 10 minutes for overall scanning time (average scanning time: 25-30 minutes for each patient). For patients less than 12 years old and good body built for age, the X7-2 matrix array transducer (frequency; 2-7 MHz) was adequate for 2D-TTE, Doppler, and RTT-3DE acquisition. The X3-1 matrix array transducer (frequency; 1-3 MHz) was used for patients who had relatively poor windows by X7-2 matrix array transducer, particularly patients who are above 12 years old and were overweight for age. The apical, parasternal, supra sternal, and subcostal windows were employed for 2D-TTE and RTT-3DE in all cases. Technical parameters (frequency, focus, gain) were optimized for each individual case to obtain the optimal image quality. In this study, we used 3 main 3DE acquisition methods, the choice of which method to be used depends on the cardiac lesion to be examined, for instance, the first method of RTT-3DE data acquisition used was Live 3DE with Zoom mode (50° x 30° pyramidal volume), which was an excellent tool for valvular pathology (for example; mitral valve prolapse [MVP], or septal defects [such as, atrial septal defect [ASD], or ventricular septal defect [VSD] assessment and delineation). The second 3DE acquisition method used was full volume (FV) acquisition of 4 electrocardiograms (ECG)-triggered sequential volumes in rapid sequence, obtaining (90° x 90° pyramidal volume), which was a good method for complex CHD segmental analysis using multiplanar reformation (MPR) technique and left ventricle (LV) volume quantification. The third 3DE acquisition method used was 3D color Doppler full volume mode (50° x 50° pyramidal volume), which was good for delineating valvular regurgitation mechanisms and severity by measuring vena contracta area. The acquired RTT-3DE data sets were stored in the machine's hard disk as Digital Imaging and Communications in Medicine (DICOM) format. Off-line data analysis was performed using dedicated software analysis available in the machine (including the QLAB Advanced Quantification software, Philips Ultrasound, USA). The reports were prepared solely on the basis of 2D echocardiography findings. The 3D studies were analyzed by a single reviewer with offline slicing at a later date focusing on the area of interest (that is, abnormalities found by 2DE) for example, if the mitral valve (MV) were found to have mitral regurgitation (MR) due to a cleft MV by 2DE, then a 3DE focused study will follow to evaluate the cleft MV in detail demonstrating the MV lesion (cleft) echocardiographic and surgical en face views. Post processing analysis of the acquired 3DE dataset was accomplished by cropping (slicing) using 2 methods;

direct cropping method (the echocardiographic full volume dataset were sectioned to obtain the structure of interest (for example, removal of left atrial (LA) upper part and lower (apical) half of the LV to see MV en face view from LA and LV perspective), and MPR-technique by pressing the MPR button in the machine's screen, displaying the 3 imaging planes (transverse, coronal, and sagittal) on the screen, by doing so any structure of interest available in any one of the 3 planes could be assessed and analyzed separately by moving the color coded bars, and the illustrated segmental changes in the corresponding colored box were observed.[6] A 4-grade system was used to compare 3D studies with conventional 2D studies (**Table 1**).[7]

Results. Fifty patients were enrolled in the current study, 29 (58%) were males and 21 (42%) were females. The patient's age group ranged from one month to 17 years (median: 60 months; range: 1-204 months), and their weights ranged from 4-70 kg (median: 22 kg). The results of the RTT-3DE were reviewed and compared with 2DE findings to define the incremental value of RTT-3DE over 2D-TTE findings. The average time required for 2DE and 3DE data acquisition including patient preparation was 25-30 minutes, and for analysis of the whole 3DE data was 25±5 minutes. The primary diagnoses on the basis of 2D examination are shown in **Table 2**. When compared with the 2D studies, 3D studies were graded A in 25, B in 24, and C in one case (**Table 3** & **Table 4**). Of the 25 cases graded as A, 9 (36%) had left heart lesions, 10 (40%) septal defects, 4 (16%) complex CHD, one (4%) aortopulmonary (AP) window, and one (4%) had right heart problem. In grade B category, 10 (41.7%) had right heart problems, while 9 (37.5%) with left heart diseases yielded useful anatomic perspectives. Two patients with ASD secundum, pre- and post device closure (8.3%), and 3 (12.5%) had precise estimation of LV volume without geometric assumption as LV volume quantification by 2D-TTE. In one patient with Tetralogy of Fallot (TOF), RTT-3DE did not add much to 2D-TTE for disconnected left pulmonary artery (LPA) detection, and this patient was graded in C category. In our series, no patient was labeled as grade D.

The shapes of atrial (2 patients) and ventricular septal defects in 6 different patients were demonstrated, and their varied nature was evident on 3D imaging, and the defect could be displayed either from the right or left atrial, or ventricular perspectives (**Figure 1A** & **Figure 1B**). Another example of the unique anatomic perspective obtained by 3DE was in 4 patients with atrioventricular septal defects (AVSD) where RTT-3DE clearly described the en face views of the ASD and VSD

Table 1 - Grading system to compare 3-dimensional (3D) with 2-dimensional (2D) echocardiography.[7]

A	3D echocardiography provides new findings not detected by 2D echocardiography
B	3D echocardiography provides useful and unique anatomic perspective but did not yield new findings
C	3D echocardiography provides equivalent diagnostic information to 2D echocardiography
D	3D echocardiography misses findings detected by 2D echocardiography

Table 2 - Primary 2-dimensional echocardiographic diagnoses in 50 CHD patients included in a study at the Prince Sultan Cardiac Center, Riyadh, Kingdom of Saudi Arabia.

Diagnosis	n
Right heart problems	12
Tricuspid valve abnormalities	8
Double outlet right ventricle	3
Tetralogy of Fallot	1
Left heart problems	18
Mitral valve abnormalities	10
Aortic valve abnormalities	6
Left ventricular outflow tract obstruction	2
Septal defects	12
Ventricular septal defects	6
Atrial septal defects	2
Atrioventricular septal defects	4
Aortopulmonary window	1
Complex CHD	4
Border line LV volume	3

CHD - congenital heart disease, LV - left ventricle

Table 3 - New findings visualized by 3-dimensional echocardiography (Grade A) in patients included in a study at the Prince Sultan Cardiac Center, Riyadh, Kingdom of Saudi Arabia (N=25).

Findings	n
MPR detected trivial TV prolapse	1
Minimal degree of MV prolapse detected	3
Multiple shapes of vena contract were identified in MR (which was always assumed to be circular by 2D-TTE)	1
Both echocardiographic and en face surgical views of cleft MV seen	2
Direct cropping clearly visualized the edges and stitched lines of MV ring repair	1
Dilated aortic root with commissural fusion in Marfan's syndrome	1
Perforation of aortic valve (right coronary cusp perforation)	1
Shape and location of ventricular septal defect in the interventricular septum	6
Shapes of atrioventricular septal defects and bridging leaflets	4
Clear en face view of aorto-pulmonary window	1
One window of 4 chamber view dataset is needed for complex CHD segmental analysis by MPR method	4

MPR - multi planar reformation, TV - tricuspid valve, MV - mitral valve, MR - mitral regurgitation, 2D-TTE - two dimensional transthoracic echocardiography, CHD - congenital heart disease

Table 4 - Useful anatomic perspectives provided by 3-dimensional echocardiography (Grade B) in patients included in a study at the Prince Sultan Cardiac Center, Riyadh, Kingdom of Saudi Arabia (N=24).

Findings	n
Visualization of the prolapsed scallops from right atrium and right ventricle perspectives in tricuspid valve (TV) prolapse patient	1
Direct cropping delineated clear tricuspid regurgitation (TR) mechanism of tethered TV leaflet in Ebstein malformation	3
Multiplanar reformation (MPR) gave precise measurements of vena contracta in gradation of TR severity	2
Clear en face views of TV morphology in hypoplastic left heart syndrome	1
In 3 double outlet right ventricle (DORV) patients, MPR proved helpful for cardiologist and cardiac surgeon in deciding:	
First patient (DORV with subaortic ventricular septal defect [VSD]): able to tunnel VSD to aorta	1
Second patient (DORV with malposed great arteries): able to tunnel VSD to pulmonary artery (followed by switch operation)	1
Third patient DORV with non committed VSD: remote VSD (not able to tunnel VSD to aorta)	1
MPR planimetry revealed precise measurements of mitral valve stenosis	2
Direct cropping revealed leaflets scallops and commissural fusion in mitral valve stenosis	1
MPR planimetry revealed precise measurements of aortic valve stenosis	3
Precise gradation of aortic regurgitation severity by MPR technique	1
Mass in left ventricular outflow tract (subaortic membrane, fibro muscular tunnel)	2
En face views of the atrial septal defect from both atria and its relation to surrounding anatomical tissue	1
Precise location of post atrial septal defect device with delineation of surrounding structures	1
Actual estimation of left ventricle volume in borderline left ventricle size	3

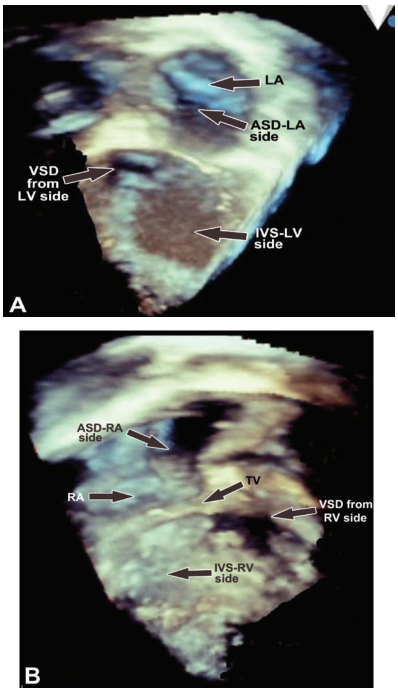

Figure 1 - Reconstruction was carried out to visualize the en face view of the atrial septal defect (ASD) and ventricular septal defect (VSD) as it would be observed: **A**) from the left atrial (LA) and left ventricle (LV) perspectives; and **B**) the right atrial (RA) and right ventricle (RV) perspectives, and its relationship with the adjacent cardiac structures. IVS - interventricular septum

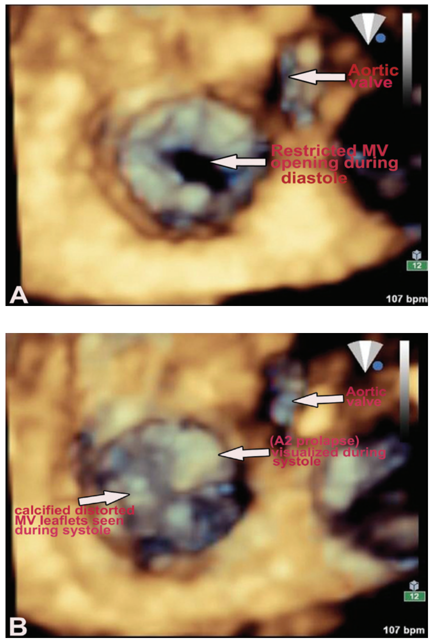

Figure 2 - Mitral valve (MV) stenosis en face views visualized by direct cropping method: **A**) surgical view during diastole; and **B**) surgical view during systole, where calcified distorted MV leaflets, in addition to prolapsed second scallop of anterior MV leaflet (A2-prolapse) visualized during systole and restricted MV opening observed during diastole.

components of AVSD from both the right and left atrial and ventricular perspectives, whereas 2DE correctly identified the defects, but was unable to provide detailed information on the morphology.

The mechanism of AR (perforation of the right coronary cusp) was clearly demonstrated by direct cropping method in one patient. In 2 patients with MV cleft, the depth of the mitral leaflet cleft was clearly demonstrated by RTT-3DE, illustrating both echocardiographic and surgical views of the MV cleft (**Table 3**). In contrast, only the 2D slice but not the interface of the cleft could be visualized on conventional 2D echocardiography. In one patient with MR, varied geometric shape of vena contracta was observed, which was assumed to be circular only by 2D-TTE studies. Slicing the upper half of LA and apical part of LV frequently provided a good en face view of the MV from the atrial and ventricular perspectives, such that the prolapsed scallops of both the anterior and posterior leaflets were visualized. The additional value of RTT-3DE on top of 2D-TTE in demonstrating AP window in 1 patient, and complex CHD segmental analysis using MPR technique were illustrated in 4 patients, 2 patients aged 2 and 4 years old were diagnosed by 2DE to have congenitally corrected transposition of the great arteries (CCTGA), the third 3-year-old patient had DORV, malposed great arteries with subpulmonic VSD, and the fourth 2½-year-old patient had pulmonary atresia intact interventricular septum (PAIVS), in all these 4 patients with complex CHD the diagnoses were made correctly by 2DE but with the use of several sweeps and windows compared with the accurate diagnoses obtained by RTT-3DE using only one view (4 chambers, full volume acquisition) with complete analysis by 3DE-MPR technique, which is adventitial compared to 2DE since less time is required by 3DE single 4 chamber view compared to the time required to obtain several windows and cuts by 2DE to reach to the similar findings as by 3DE, a fact which is extremely helpful for screening unwell young pediatric patients, in whom sedation cannot be administered, and less time is required to reach the complex CHD segmental analysis and correct diagnosis by 3DE.

Useful perspectives of cardiac anatomy (grade B) were provided by 3DE in 24 patients (**Table 4**). The AS and AR severity were precisely measured using MPR technique. Mitral valve stenosis was visualized by direct cropping method (**Figures 2A & 2B**) where calcified distorted MV leaflets were observed, in addition to prolapsed A2 scallop. In 3 patients with Ebstein's anomaly, the exact mechanism of TR and tethered leaflets of TV were visualized. Proper evaluation of the LV size using advanced 3DE quantification Q Lab software semi-automatically detects and traces the endocardial border of the LV during the cardiac cycle, with a possibility to perform manual corrections as needed.[8] In 3 patients with borderline LV volume, it was significantly helpful for the physicians to decide which patients should go for biventricular type of surgical repair (patients diagnosed by RTT-3DE to have adequate LV size (LV volume of LVEDV/BSA of 15-20 mL/m^2)[9] as in patient 1, 3 were candidates for this kind of repair), and in patient 2, RTT-3DE declared the LV volume small (LV volume of less than 15 mL/m^2) and inadequate for biventricular repair.

Discussion. This study reports on a local experience in KSA of CHD evaluation by RTT-3DE. This current study proved the additional value of RTT-3DE over 2D-TTE in delineating more detailed anatomical structures not well perceived by 2D-TTE. Takahashi et al,[10] Seliem et al,[11] and this present study clearly emphasized that RTT-3DE is complementary, but not a replacement to 2D-TTE. This study demonstrated the additional value of RTT-3DE in providing more anatomical details of various cardiac lesions including, right and left heart problems and septal defects when compared to 2D-TTE studies of similar lesions, thus reinforcing the findings of some other studies.[12,13] Several reports described previously under-pinned the added strength of RTT-3DE in evaluating valvular heart lesions in adults.[14-16] The present report is consistent with the few reported experiences in demonstrating the additional value of RTT-3DE over 2D-TTE in evaluating cardiac valvular lesions in pediatrics.[17-19] Direct cropping methods delineated the TV malformation in Ebstein anomaly clearly in our 3 patients while using RTT-3DE. Similar findings have been reported by Vettukattil et al as well.[20] The RTT-3DE - MPR technique turned out to be very helpful in grading the severity of TR in 2 patients enrolled in this study. Velayudhan et al[21] has reported similar experience for RTT-3DE, however its implementation as the investigation of choice requires more validated studies to be carried out.

This report is in agreement with Khanna et al's[22] study in confirmation of RTT-3DE supremacy over 2D-TTE in quantification of MR severity. The RTT-3DE provides unique information regarding direction and extent of jet, which is useful in eccentric jets but requires more studies to confirm the usefulness of RTT-3DE estimation of MR severity in pediatric population. This study has reaffirmed the adjuvant role of RTT-3DE in evaluation of AoV stenosis and regurgitation, which is consistent with published literature.[23,24]

In this study, RTT-3DE was found advantageous in providing unique en face views of the ASD, VSD, and AVSD from right atrial (RA), LA, right ventricular (RV) and LV sides with clear delineation of the defects shape, size, and number. It was found that 3DE provides incremental value for diagnosis, assessment, and interventional guidance during closure of secundum ASDs, as also narrated by Ge et al.[25]

The results of this study are in agreement with Kutty and Smallhorn's[26] conclusion that the real strength of RTT-3DE in AVSD evaluation is the ability to identify the left AV valve complexity and left ventricular out flow tract (LVOT) abnormalities associated with the defect. These 3D images have been useful for surgeons in analyzing the congenital heart lesions without imaginary reconstruction of the anatomical structures and deciding the type of intervention before opening the heart. The RTT-3DE in assessment of ventricular volume does not have any assumption to calculate LV volume compared to quantification with 2D-TTE, which requires several anatomical assumptions of true myocardium inclusion.[27] As previously described,[28-30] this study demonstrated the effectiveness of LV volume assessment by RTT-3DE.

This study pointed out that 2DE views taken by X7-2 matrix array transducer were of sub-optimal quality as compared to views obtained by dedicated 2DE transducers. This demands the application of separate dedicated 2DE and RTT-3DE transducers which will certainly increase the acquisition time, but we think that with the next generation of fully matrix arrayed transducer, the RTT-3DE transducers will be able to obtain higher quality 2D-TTE images like dedicated 2DE transducer, in addition to their original function of 3DE images acquisition. Similar findings were reported by Bhan et al.[31] Although this study illustrated the usefulness of RTT-3DE over 2D-TTE in evaluating various forms of CHD, in one of our patients (TOF with disconnected LPA), RTT-3DE did not add significant additional information over 2D-TTE. Lang et al[32] has claimed the useful application of RTT-3DE in adult echocardiographic studies particularly in LV mass, volumes, and ejection fraction estimation, and evaluation of valvular heart diseases especially mitral valve evaluation in the operating room before repair. They have also described other striking usefulness of RTT-3DE in CHD evaluation, but at the same time they still think that there are some unresolved issues regarding the utility of RTT-3DE in routine clinical use as the temporal and spatial resolution limitations, image acquisition, processing, and storage limitations, and cost effectiveness. On the other hand, Bhan et al[31] clearly stated that infrequent users of RTT-3DE continued to have some confusion regarding its exact role, usefulness, and drawbacks. Apparently, the debate for the benefits and limitations of routine clinical use of RTT-3DE may continue, however, evidence-based research substantially supports the complementary usefulness of RTT-3DE in addition to 2D-TTE in routine practice of CHD evaluation.

Limitations of RTT-3DE. During interventricular or inter-atrial communications assessments by RTT-3DE, echo dropout might develop but by implementing dedicated protocols and procedure techniques along with rigorous training of echocardiographers, this shortcoming can be controlled. Arrhythmic patients, breath holding, and inability to stay still in pediatric population are sources of stitch artifact during full volume and 3DE color flow Doppler mode acquisition, which requires consecutive 4-7 ECG triggered cycle. The newer generation of single beat acquisition instead of 4-7 consecutive ECG- triggered full volume acquisition is now available, and hopefully will overcome this problem.

In conclusion, the RTT-3DE is an important emerging diagnostic tool for real-time CHD evaluation. The current study reported that, it can provide additional information regarding the surface anatomy of cardiac valves, superior evaluation of inter-atrial and inter-ventricular septal en face views, and a better analysis of complex morphology of CHD lesions. This technique will eliminate the imaginary anatomical and geometrical assumptions in LV volume and size assessment. Any further improvement in the learning curve and RTT-3DE experience of the echocardiographers together with the machine's software advancement will definitely result in extensive improvement in the diagnosis, deciding best interventional modalities, and finally improve the prognosis and outcomes in CHD patients.

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