REVIEW

EDUCATIONAL SERIES IN CONGENITAL HEART DISEASE: Three-dimensional echocardiography in congenital heart disease

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Abstract

Three-dimensional echocardiography is a valuable tool for the assessment of cardiac function where it permits calculation of chamber volume and function. The anatomy of valvar and septal structures can be presented in unique and intuitive ways to enhance surgical planning. Guidance of interventional procedures using the technique has now become established in many clinical settings. Enhancements of image processing to include intracavity flow, image fusion and true 3D displays look set to further improve the contribution of this modality to care of the patient with congenital heart disease.

Introduction

Three-dimensional echocardiography (3DE) has become a complementary echocardiographic modality in the management of patients with congenital heart disease. 3DE is used frequently in children, but also its role in the long-term care of adult congenital heart disease (ACHD) is well established. The technique has been applied to assessment of cardiac anatomy and quantification of cardiac function as well as peri-operative and interventional guidance. A recent expert consensus document has reviewed the applications in clinical practice for the patient with congenital heart disease (1) and the core concepts of the 3D approach have been described previously (2). This review will focus on practical aspects of the technique, current areas of interest and advances.

Data acquisition modes

Good spatial and temporal resolution in 3DE is a priority for imaging of CHD, particularly valve pathology and complex lesions. The matrix transducer has different modalities of data acquisition whose use is dictated by the clinical question (Fig. 1). Different ultrasound manufacturers will have variable terminology for the different modalities, but there are a number of common themes. At some point, it is hoped that terminology may converge so that users have a ‘common language’ to describe different aspects of the technique.

Imaging of orthogonal planes

Most 3D matrix transducers permit the simultaneous display of orthogonal planes so that two or three distinct cross-sectional planes can be viewed simultaneously. Although not truly ‘3D’, this modality makes use of the matrix probe design so that the scanning plane can be altered electronically rather than by rotation of the ultrasound probe. As an example, this technique can be used for the visualization of atrial septal defects (3).
This modality displays in real-time, a 3D pyramidal volume with a limited sector size. Rotating this 3D image quickly checks and allows optimization of general 3D image settings.

3D zoom

Zoom displays in real-time, a magnified subsection of 3D pyramidal volume that is centered to a specific region of interest for example the mitral valve (Fig. 2).

Real-time 3D full-volume

Full-volume 3D data set is an ECG-gated acquisition of a large 3D pyramidal volume. Individual wedge-shaped subvolumes obtained over consecutive heartbeats (R wave of ECG) are stitched together and synchronized to the same cardiac cycle. Current ultrasound machine software displays the full-volume in real-time with the option to determine volume size (elevational and lateral controls) and volume rate (number of heart beats) independently.

3D color Doppler

Color Doppler full-volume 3D data set is a gated acquisition of a small 3D pyramidal volume with superimposed 3D representation of color Doppler flow. The color flow can be displayed in real time.

For all modalities, the echocardiographer needs to optimize the temporal and spatial resolution by adjustment of the size of the region of interest and the number of beats used to acquire the volume. Image gain control has a critical role in reducing intracavity noise so that far field structures can be visualized. All manufacturers provide different rendering modalities which include color coding to improve depth perception and adjustment of smoothing and dynamic range.

Assessment of cardiac function using 3D echocardiography

All 3D echocardiographic modalities used to assess cardiac function depend on the detection of the endocardial and/or epicardial border. Thus, their accuracy depends on incorporation of the entirety of the chamber in the echocardiographic volume and high quality imaging. This may be challenging in the context of dilated ventricles for example dilated cardiomyopathy or if sonographic views are challenging for example anterior position of the right ventricle.

3D echocardiographic assessment of the LV

Chamber quantification is crucial for the prognostication, guiding therapy and follow-up in patients with congenital heart disease. Today, real-time 3DE is a valuable tool for the assessment of LV volumes and LV ejection fraction (LVEF). Multiple studies have shown that 3DE is more accurate and reproducible than 2DE, because direct measurement of volumes can be achieved without the need for geometrical assumptions about cavity shape and limitations associated with foreshortened views (1, 4). In the patient with congenital heart disease, LV geometry is often distorted due to the malformation itself or as a result of a dilated and/or high-pressure right ventricle. In this regard, 3D echocardiography has an important contribution to the assessment of LV volumes, function and LV mass.
Analysis of LV volumes and function

The 3D echocardiographic data set is obtained from an apical or subcostal position or a modified transducer position to ensure the whole LV is captured in the 3D data set. Normally the 3D data set will comprise the entire ventricle, except in dilated left ventricles or in adults with functionally single ventricle circulation. To assess global and regional LV function, surface rendering is used and most vendors offer software packages for online and offline quantification of LV volume and function. The endocardial border tracking algorithms used for calculating a 3DE LV volume represents a wide spectrum ranging from fully automated to manual-based algorithms. The variability of the LV measurements with 3D echocardiography is dependent on the image quality and operator experience. In congenital heart disease, if the LV is abnormally shaped, fully automated border detection seems to be less reliable than semi-automated methods with the potential for manual correction/override. This implies that functional analysis with the 3D echocardiographic software needs to be used with caution in patients with abnormal anatomy. Use of a manual method of disks has provided reasonable correlation with MRI estimation of LV volumes and mass in patients with congenital heart disease but with lower estimated ejection fraction using echocardiography (5), and more manual border tracking throughout the cardiac cycle is required. When LV volumes measured by 3DE are compared to MRI, there is a bias for echocardiography to produce lower volumes than those from MRI. The reader is referred to the recent consensus document for further information (1).

Analysis of LV mass

3D echocardiography has been used to assess LV mass. However, accurate assessment is challenging, especially in large and abnormally shaped left ventricles as in congenital heart disease. Reports on LV mass assessment in children are very limited (5, 7, 8). Inaccurate measurement of LV mass can occur for a number of reasons; tracing difficulties of endocardium and epicardium might explain measurements differences, but also inadequate visualizing of the apex and poor image quality with lower echogenicity, hampers accurate calculation of LV mass. Therefore, the applicability of 3D echo for LV mass calculation for use in clinical practice in patients with congenital heart disease remains to be established.

Analysis of LV dyssynchrony

3D echocardiography can capture the entire LV and offers the opportunity to assess global and regional LV function accurately and reproducibly and to assess LV intra-ventricular synchrony. 3D echocardiography has been used to assess intra-ventricular dyssynchrony and is expressed as the standard deviation of the time taken for segments to reach their minimum systolic volume, indexed to the cardiac cycle length (systolic dyssynchrony index (SDI)). Normal data in children and adolescents are available (9); however, current software packages define abnormal wall motion with respect to the central LV axis. This is a limitation for some CHD patients with an LV of unusual shape. Furthermore, analysis of timing of minimum systolic volume of any given segment is particularly challenging when ventricular function is poor. Therefore, caution is required when using 3DE as a modality to quantify LV dyssynchrony in CHD and especially if overall ventricular function is poor. Further work in this area is necessary to establish the prognostic value of such measurements in the patient with CHD.

3D speckle tracking of the left ventricle

3D echocardiography has theoretical advantages compared to 2D techniques for the assessment of myocardial deformation. 2D speckle techniques can only follow unique kernels through the cardiac cycle if these remain within plane. In contrast, 3D techniques can potentially allow for through plane motion and also measure rotation of the myocardium to facilitate measurement of twist and torsion. There are limited pediatric data of the application of this technique (10), which is restricted to the morphologically normal LV and the challenge of abnormal ventricular shape remains an issue for this modality. There are scant data on application in adult patients with congenital heart disease (11, 12).

Assessment of the right ventricle

The right ventricle (RV) is frequently involved in congenital heart disease and measurement of RV
volume and function are increasingly important for prognosis and clinical decision making. Conventional 2D echocardiographic assessment of RV volume and function is hampered by the complex geometry and retrosternal position. In congenital heart disease, the RV is often volume and/or pressure overloaded resulting in a dilated RV with abnormal shape. There may be a systemic RV in the context of a biventricular circulation for example, congenitally corrected transposition of the great arteries (TGA) or in the context of a functionally single ventricle circulation for example hypoplastic left heart (HLH). In such patients, ventricular dysfunction and heart failure may occur. Accurate quantification by 3D echocardiography may be able to identify ventricular dysfunction before overt clinical symptoms. At present, cardiac magnetic resonance has become the imaging modality of choice for assessment of RV volumes and function, because visualization is not constrained by acoustic windows and the analysis makes few assumptions about the shape of the heart chambers. However, compared with TTE, CMR is more expensive, is not portable and requires sedation or anesthesia in young patients. In addition, some patients will have prosthetic valves, pacemakers or implantable defibrillators which preclude the use of CMR. Several studies have demonstrated the ability of 3D echocardiography to assess RV volumes and function in children and adults. Also, the accuracy of 3D echocardiography for RV assessment in congenital heart disease, for children and adults, has been validated with magnetic resonance imaging. At this time, 3D echocardiography uses semi-automated border detection of a volume of data, usually acquired over several cardiac cycles. Images for RV analysis are preferably acquired by transthoracic echocardiography, but can also be acquired with transoesophageal echocardiography. Limitations include poor acoustic windows and therefore limited feasibility. Direct comparison of echocardiographic and CMR volumes have shown systematic underestimation of RV volumes by echocardiography compared to CMR. The extent of agreement between echocardiography and MRI also varies according to the specific lesion under consideration and between publications (1). Currently, echo and MRI-derived measurements of RV volume cannot be used interchangeably. Very recently, a large series in children and adolescents has shown a much closer agreement than previously described using newer echocardiographic software and has provided normal ranges in younger patients based on a far larger population than previously reported (13). Other recent work has co-registered MRI and 3D echocardiographic images in HLH to locate where differences between the techniques are noted (14). That publication reported that most of the difference in CMR and 3D echocardiographic volumes related to the endocardial border in the ventricular apex.

### Echocardiographic techniques for the analysis of RV volume and function

#### Semi-automated border detection

This is the most common 3DE method to assess RV volumes and EF. A full-volume 3D data set is acquired and segmented into four-chamber, sagittal and coronal views. The RV end-diastolic and end-systolic contours are manually drawn in each view to construct a dynamic polyhedron model of the RV (Fig. 3).

#### Knowledge-based 3D reconstruction

Knowledge-based 3D reconstruction evaluates 3D RV volumes from a series of 2DE images localized using a magnetic tracking system. The RV anatomic landmarks are identified on the images, which are processed over the Internet using a reference lesion-specific MRI database. This technique has been validated against MRI in children after TOF repair (15). Initial reports suggested that bias, intra- and interobserver reliability were better than semi-automated techniques and had a closer agreement with MRI. The limitations of the knowledge-based technique include the necessity for a tracked ultrasound transducer and for the patient to remain still throughout the study. Thus, at present, this technique has not found widespread application and tends to be limited to a research setting.

#### Manual planimetry techniques

Some software packages permit planimetry of the ventricle by manually tracing the contour of the endocardial border in a ‘slice-by-slice’ fashion (16). This is analogous to the approach used in CMR. This technique is quite time consuming and has not been adopted widely in regular clinical practice.

### Limitations of 3D echocardiography for assessment of RV function

3D echocardiography still has shortcomings in terms of spatial and temporal resolution and the requirement to obtain high-quality datasets over multiple cardiac cycles. The extent of agreement between echocardiography and MRI also varies according to the specific lesion under consideration and between publications (1). Currently, echo and MRI-derived measurements of RV volume cannot be used interchangeably. Very recently, a large series in children and adolescents has shown a much closer agreement than previously described using newer echocardiographic software and has provided normal ranges in younger patients based on a far larger population than previously reported (13). Other recent work has co-registered MRI and 3D echocardiographic images in HLH to locate where differences between the techniques are noted (14). That publication reported that most of the difference in CMR and 3D echocardiographic volumes related to the endocardial border in the ventricular apex.

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### Limitations of 3D echocardiography for assessment of RV function

3D echocardiography still has shortcomings in terms of spatial and temporal resolution and the requirement to obtain high-quality datasets over multiple cardiac cycles.
The feasibility of 3D echocardiographic RV volumes measurements in daily clinical practice is around 60%, even in experienced hands. However, when feasible, the interobserver and interstudy reproducibility is good for RV volume and moderate for RV ejection fraction. Also, there are limited normative data available, with studies using different methods and small numbers of subjects. It should be taken into account that there is a systematic underestimation of RV with 3D echocardiographic compared to CMR.

Functionally single ventricle circulation

In patients with a single ventricle, ventricular dysfunction remains one of the most important long-term complications. In children, the subcostal position can be used to capture the entire ventricle in the 3D data set. However, from this echo window the heart is in the far field, which may influence frame rate and spatial resolution. Aside from the technical considerations, ventricular morphology in the functionally single ventricle circulation may be far removed from ‘normal RV’ or ‘normal LV’, which limits the use of semi-automated software algorithms (16). The enlarged dominant ventricle in these hearts, especially in adults with single ventricle, makes it difficult to incorporate this whole ventricle in a single data set with acceptable resolution and access to visualize all necessary borders.

Three-dimensional echocardiographic assessment of cardiac morphology

The application of 3D echocardiography to the assessment of cardiac morphology can be broadly subdivided into assessment of heart valves, visualization of septal structures and interrogation of more complex disease to assist surgical planning (1). The technique has been widely applied before procedures but also during either catheter or surgical procedures, which has been facilitated by the development of transoesophageal echocardiography probes, which have 3D functionality. Each of these applications will be addressed in turn.

Heart valves

3D echocardiographic techniques have been widely used to assess heart valves for a number of reasons including the ability to display en face views of such valves with

(A) Shows the contours of the right ventricle in the four-chamber, sagittal and coronal planes which are used to construct the 3D volume which is shown in (B) along with the rendered 3D image of the right ventricle.
a depth of field in a manner which is not technically feasible using standard cross-sectional techniques. In congenital heart disease, application of 3D echo for the interrogation of the mitral valve, tricuspid valve and atrioventricular septal defects are some of the most common applications of the modality (17, 18). Current software packages permit precise alignment to the plane of the valve and measurement of both rendered 3D images and multiplanar reformatted images. The ability to rapidly move between different projections for example display of the mitral valve from either the atrial or ventricular aspect makes the technique particularly adaptable to provide comprehensive visualization of the valve for surgery (19). Imaging of valves and chordal support apparatus in non-standard planes, which are user defined is another major advantage of the technique (Fig. 4 and Videos 1, 2). There has been a major drive to automate quantification of size and function of cardiac valves including the mitral and aortic valves to improve workflow and derive maximum information about valvar function, including leaflet area, tenting height and volume and accurate localization of abnormal valve leaflets. However, to date, most of these innovations have made assumptions about valvar structure for example the mitral valve is bileaflet, which may not be the case for the patient with congenital heart disease. Hence, further work and innovation will be necessary to bring such automation to a wide spectrum of congenital heart defects. In a research setting, individual patient valves can be modeled to produce a ‘patient-specific’ view of abnormal valves (Fig. 5). Patients with CHD are prone to endocarditis and 3D echocardiography is an excellent modality for visualization of vegetations or damaged heart valves cause by infective endocarditis.
Video 1

Video 2

Septal structures
Beyond visualization of heart valves, 3D echocardiography is the modality of choice to image septal abnormalities such as ventricular and atrial septal defects of different types. This can be done by either transthoracic or transoesophageal echocardiography (Figs 6, 7 and Videos 3, 4, 5, 6). The only current technical limitation is the size of 3D TOE probes, which cannot be used in patients below about 20–25 kg. Examples of atrial and ventricular septal defects visualized by 3D echocardiography are shown in Figs 6 and 7. Particular advantages of the 3D approach is that defects of unusual shape, location or multiple defects can be visualized clearly. The relationship of defects to adjacent anatomic structures is intuitive and can assist both the surgeon and interventionist. The technique can be applied either before or during the procedure. This is discussed in more detail below.

Video 3

Video 4

Figure 7
Transthoracic 3D echocardiogram of a perimembranous ventricular septal defect projected from the right ventricular aspect (A) and left ventricular aspect (B). (A) This view shows the relationship of the VSD to the tricuspid valve and right ventricular outflow tract. (B) This view, from the left ventricular aspect, shows the relationship of the VSD to the aortic valve. LV, left ventricle; RA, right atrium; RV, right ventricle; RVOT, right ventricular outflow tract; TV, tricuspid valve; VSD, ventricular septal defect.

Figure 8
Transthoracic 3D echocardiographic interrogation of a patient with usual atrial arrangement, discordant atroventricular connections and double outlet right ventricle. This was done to plan surgery, including feasibility of routing of blood flow from the morphologic left ventricle to the aorta. (A) View from the ventricular apex. (B) Multiplanar reformatted image using cropping tools. The plane of visualization is from the red dotted line into the white box. (C) 3D rendered image of the projection defined in (B). This shows the position of the tricuspid valve which overlies a large ventricular septal defect (outlined by asterisks) and the location of the aorta and pulmonary artery in relation to the ventricular septal defect. Ao, aorta; LA, left atrium; mLV, morphologic left ventricle; mRV, morphologic right ventricle; MV, mitral valve; PA, pulmonary artery; TV, tricuspid valve; VSD, ventricular septal defect.

Video 6

Complex congenital heart disease
In addition to use of 3D echocardiography to visualize valves and septal structures, there are more complex situations where 3D echocardiography can assist planning (20). This approach utilizes the depth of field of 3D echocardiography to show the orientation of different parts of the cardiac anatomy, for example, the relative position of atrioventricular valves, septal defects and the outflow tracts. In these authors’ practice, this approach can be assistance for lesions such as double outlet RV, transposition of TGA with associated lesions and discordant atrioventricular connections. An example of the benefits of this type of approach is shown below (Fig. 8 and Videos 7, 8). An advantage of the use 3D echocardiography for this type of assessment is that the real-time motion of the atrioventricular valves is appreciated and the changing size of septal defects through the cardiac cycle. In practice, the information from 3D echocardiography is frequently integrated with other modalities such as CT, magnetic resonance imaging or 3D printed models. CT and MRI are unconstrained by acoustic windows and build up the ‘full picture’ along with the dynamic 3D echocardiographic data.

Video 7
Transthoracic 3D echocardiographic view of a patient with discordant atrioventricular connections. The tricuspid valve (left side of patient) and mitral valve (right side of patient) can be visualized as well as the aorta which arises from the RV. View Video 7 at http://movie-usa.glencoesoftware.com/video/10.1530/ERP-18-0074/video-7.

Figure 9
Transoesophageal 3D echocardiography during occlusion of atrial septal defect. (A) Transoesophageal 3D echocardiogram from a right atrial view showing a catheter coursing superiorly, adjacent to a large atrial septal defect. The depth of field of the 3D technique permits visualization of both defect and catheter. (B) Left atrial view of ASD occlusion device and delivery catheter. LA, left atrium; RA, right atrium.

Figure 10
3D printed model of double outlet right ventricle in an infant. This model was produced from cardiac MRI and was created to assist surgical planning, in particular with respect to routing of blood from the left ventricle to the aorta. The model is viewed from the right, with the right ventricular free wall reflected away. The proposed route is shown by the orange tubing.
Video 8

Figure 11
This image shown blood speckle tracking through a ventricular septal defect. The direction and velocity of flow is indicated by the direction and length of the arrows respectively. Reprinted from Ultrasound in Medicine and Biology, Vol 40, Fadnes S, Nyrnes SA, Torp H & Lovstakken L, Shunt flow evaluation in congenital heart disease based on two-dimensional speckle tracking, Pages 2379–2391, Copyright (2014), with permission from Elsevier (34).

Guidance of catheter interventions
3D echocardiography is well established for the guidance of catheter interventions, as it can project defects in real-time, as well as high-quality imaging of catheter delivery systems and the devices themselves (1) (Fig. 9). Thus, the technique has found application in transcatheter closure of atrial septal defects and ventricular septal defects in particular as well as a range of other situations such as imaging of coronary artery fistulas, closure of baffle leaks and fenestrations (21, 22, 23). Although attempts are made to preserve native valves, particularly in younger patients, 3D echocardiography provides excellent imaging of prosthetic valves and their complications, including guidance of closure of paravalvar leaks. In our usual practice, in patients large enough to take the 3D TOE probe (typically >25 kg), we would image any defect planned for closure using either a full-volume or live 3D modalities in advance of the closure procedure. Live 3D guidance of ASD closure is particularly helpful if there are multiple defects, fenestrated defect or if more than once device is being deployed.

New developments in the application of 3D echocardiography
Image fusion
Although 3D echocardiography has major advantages for the real-time imaging of dynamic structures such as the atrioventricular valves, there is an inherent constraint on both field of view and sonographic windows. Both CT and MRI provide superb high-resolution images which are unconstrained by acoustic access. Thus, there is increasing interest in image fusion to play to the strengths of each
technique to provide the maximum amount of diagnostic information and to assist surgical and catheter procedures (24, 25, 26). In the cardiac catheterization laboratory, angiographic information and echocardiographic visualization can be combined to assist in navigation through the catheter procedure (27).

3D printing from echocardiography

3D printing has become increasingly accessible due to technological advances and decreased cost of printing from imaging data formats. There is clear evidence that printing of 3D models can impact on surgical decision making in patients with congenital heart disease (28). In most cases, 3D printing has been based on either CT or CMR data but printing from 3D echo has now been described (29). Such printing is being extended to features such as atrioventricular valves which are difficult to image by other modalities (30). This permits printing of a ‘patient-specific’ valve but only at a single point in the cardiac cycle due to the static nature of the 3D print (Fig. 10).

Intracavity flow

Most work on 3D echocardiography has focused on the myocardium or on cardiac valves but the technique can be applied to tracking of blood flow within the chambers of the heart and through cardiac defects (Fig. 11). This has the potential to map blood flow vortices in three dimensions (31, 32, 33) (Fig. 12 and Video 9). Currently, this type of approach is confined to a research setting but has potential clinical application in measurement of kinetic energy of blood flow, energy loss within ventricles and energy efficiency. A 2D application of tracking intracardiac flow is now commercially available which tracks the blood speckle pattern at high frame rates (34) (Video 10). The place of such techniques in clinical practice for the management of the congenital heart disease patient remains to be established. Preliminary work suggests that intracavity flow patterns may be of importance with respect to kinetic energy of blood flow and ventricular efficiency in selected groups with congenital heart disease (35).

Video 9


Video 10

**Image display**

One of the criticisms of current 3D imaging techniques is that they remain displayed on a ‘flat screen’ interface. Manufacturers have used a variety of shading, color coding and illumination methods to enhance depth perception of the rendered 3D images. The ability to tilt and rotate images also enhances the interface with the echocardiographer. There are a number of emerging holographic, augmented and virtual reality approaches which are likely to emerge to enhance the user interface and to assist understanding of the 3D anatomy even further (Fig. 13 and Video 11) (36). This may assist not only surgeons or cardiologists but also patients’ understanding of their condition.

**Video 11**

Augmented reality cardiac imaging of a normal heart showing a ‘free floating’ heart which can be interrogated and measured. Video courtesy of the ‘3D Heart Project’, a collaboration between Guy’s and St Thomas’ NHS Trust and King’s College London. View Video 11 at http://movie-usa.glencoesoftware.com/video/10.1530/ERP-18-0074/video-11.

**Declaration of interest**

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