

REVIEW

Three-dimensional echocardiography of congenital abnormalities of the left atrioventricular valve

Kathryn Rice MBChB and John Simpson MD FRCP FESC

Department of Congenital Heart Disease, Evelina London Children's Hospital, Guy's and St Thomas NHS Foundation Trust, Westminster Bridge Road, London SE1 7EH, UK

Correspondence should be addressed to J Simpson
Email
John.Simpson@gstt.nhs.uk

Abstract

Congenital abnormalities of the left atrioventricular (AV) valve are a significant diagnostic challenge. Traditionally, reliance has been placed on two-dimensional echocardiographic (2DE) imaging to guide recognition of the specific morphological features. Real-time 3DE can provide unique views of the left AV valve with the potential to improve understanding of valve morphology and function to facilitate surgical planning. This review illustrates the features of congenital abnormalities of the left AV valve assessed by 3DE. The similarities and differences in morphology between different lesions are described, both with respect to the valve itself and supporting chordal apparatus. The potential advantages as well as limitations of this technique in clinical practice are outlined.

Key Words

- ▶ mitral valve
- ▶ left atrioventricular valve
- ▶ three-dimensional echocardiography
- ▶ congenital heart disease
- ▶ atrioventricular septal defect
- ▶ congenital mitral valve lesions

Introduction

Clinically significant congenital mitral valve lesions have been described in 0.49% of new patients presenting to a tertiary centre (1). This probably underestimates their true prevalence due to associated abnormalities dominating the clinical presentation. Cross-sectional echocardiography has a firmly established role in the evaluation of congenital mitral valve disease (1, 2, 3). Standard views include parasternal long- and short-axis projections, supplemented by 'sweeps' of the sonographic plane through the valve and chordal support apparatus. Real-time three-dimensional echocardiography (RT3DE) has some major advantages for the visualisation of atrioventricular (AV) valves because it provides a depth of field, and the ability to post process images to show unique projections, such as en face views of AV valves, which cannot be achieved using cross-sectional techniques alone (4, 5). The anatomy and cross-sectional echocardiographic features of congenital mitral valve abnormalities have been described previously (1, 6, 7).

The aim of this review is to present the RT3DE appearances of the normal mitral valve and a range of congenital abnormalities of the left AV valve with emphasis on the novel and additional information which may be provided by RT3DE.

Institutional practice and methods of acquisition

This study was undertaken at the Evelina London Children's Hospital, a designated tertiary referral centre for congenital heart disease. RT3DE is used in all cases before surgical repair of an AV valve. The choice of transthoracic vs transoesophageal RT3DE was determined by the adequacy of transthoracic acoustic windows and the adequate size of the patient to accommodate a RT3DE transoesophageal echocardiography probe (≥ 25 kg). This review uses an anonymised selection of images,

which have been obtained for clinical reasons; publication of which is supported by institutional policy. Trans-thoracic RT3DE acquisitions for the left AV valve were obtained using a modified parasternal long-axis view, parasternal short-axis view or less commonly an apical four-chamber view. Images were obtained either using the live 3D modality or by acquisition of a sonographic volume over four to six cardiac cycles, incorporating the valve and chordal support apparatus. Transoesophageal RT3DE was only used in patients with a weight ≥ 25 kg due to the size of the probe. Acquisitions were from a four-chamber view at a mid-oesophageal level at 0–30°. Other views also used include a mid-oesophageal level at ~ 120 –130° to show the mitral valve, left ventricle and left ventricular outflow tract (LVOT), or a transgastric view at 0–30° to project the short axis of the left AV valve. The RT3DE volumes were acquired on a Philips IE33 system using the Philips X3-1, X5-1 or X7-2 transthoracic ultrasound probes appropriate for patient age and size. The X7-2t transoesophageal ultrasound probe was used for patients ≥ 25 kg. Post-acquisition analysis was via Q Lab Software V9 (Philips Medical Systems, Andover, MA, USA). Details of image acquisition and post-processing techniques have been published previously (8, 9, 10, 11, 12). Images of AV valves are presented in an anatomical orientation as described previously (13, 14). Such orientation is achieved in post processing, independent of the imaging projection used to acquire the sonographic volume. Diagrams of the mitral valve were produced using the Heartworks echocardiographic simulator (Inventive Medical, London, UK).

The normal mitral valve

The mitral valve is a complex composed of the annulus, leaflets, tendinous chords and papillary muscles. It occupies the left ventricle inlet extending from the AV junction to the border of the apical region, with the apical junction demarcated by the mitral papillary muscle insertion (6). The annulus undulates to form a saddle shape, with a more D shaped than circular profile. The anterior (aortic) leaflet of the mitral valve is in fibrous continuity with the aortic valve anterosuperiorly. Thus, blood flows into the heart over the atrial aspect of the anterior mitral valve leaflet and out of the heart into the aorta over the ventricular aspect of this leaflet. The annulus support for the posterior (mural) leaflet is less rigid than the anterior annulus (15). The mitral valve sits obliquely within the left ventricle with the papillary muscles in superolateral and inferomedial position.

These papillary muscles have also been termed anterolateral and posteromedial respectively, reflecting their position when the heart is viewed whilst placed on its apex, though not when it is in an anatomically correct position (14). The rounded anterior leaflet has two demarcated areas, the first a substantial planar region, free of chordal attachments called the smooth zone. Either side of this and nearer to the leaflet edge is where layered attachments of tendinous chords occur. This leaflet region is nodular and irregular. It is called the rough zone (16). The posterior leaflet is more variable in its anatomy. It is shallower than the anterior leaflet and forms two-thirds of the valve circumference (6). It contains at least three indentations along its free edge dividing the leaflet into ‘scallops’ that are best seen when the valve is closed (15). The posterior leaflet also has smooth and rough zones demarcated by the same anatomical characteristics, but in addition has a more proximal leaflet area, the basal zone, which is where the tertiary chords attach. There is a single line of apposition between the two leaflets along the rough zone, with this closure line falling short of the annulus (15). The primary chords attach along the free edge of both leaflets. Their role is in prevention of leaflet prolapse (17). Secondary chords of the anterior leaflet extend from the ventricular under-surface of the rough zone to the most proximally placed papillary muscle below. Tertiary chordal attachments arise directly from small muscle bellies on the wall of the ventricle to the ventricular surface of the posterior leaflet only (16). Both secondary and tertiary chordal attachments have a haemodynamic role during ventricular deformation (15, 17). The main papillary muscles arise from the left ventricle directly below the two ends of the single zone of mitral valve leaflet apposition (15). Whilst described as two distinct papillary muscles, they can be formed of two muscle clusters (16).

The RT3DE appearances of the normal mitral valve are shown in Fig. 1. The three viewing aspects that provide the most information include rendered views of the mitral valve from an atrial projection as shown in Fig. 1A and Video 1, from a ventricular projection as seen in Fig. 1B and Video 2 ‘chordal’ projection showing the chordal support apparatus of the mitral valve to both its medial and lateral margins is shown in Fig. 1C.

Congenital mitral valve disease

Parachute mitral valve

Figures 2A, B and Video 3 show the defining features of the parachute mitral valve, namely insertion of the chordae

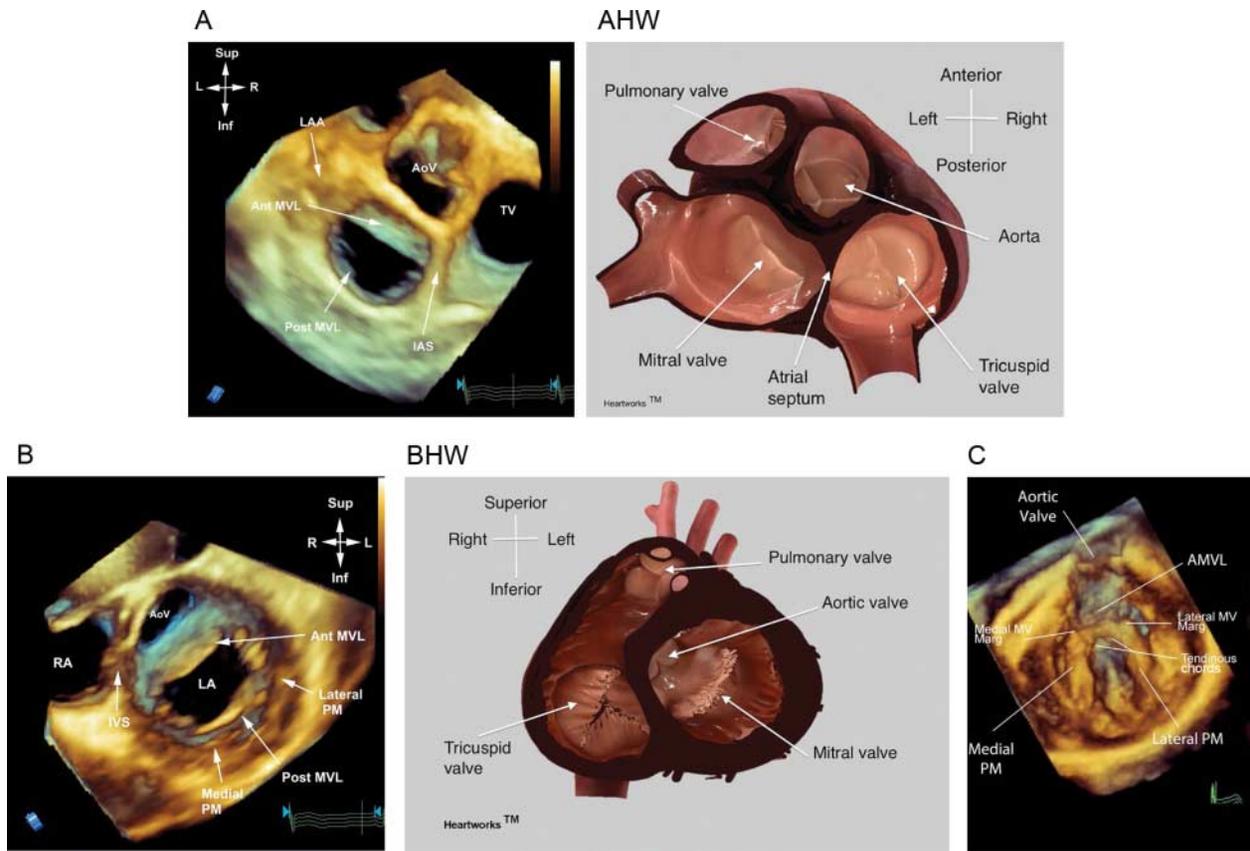


Figure 1

The normal mitral valve is presented using Heartworks and RT3DE transthoracic rendered image views. (A and AHW) An en face view of the mitral valve as viewed from the left atrium and in an anatomic orientation. The aorta is positioned at ~2 o'clock using a clock face analogy. This represents a true superior/inferior, left/right spatial position. This demonstrates the bileaflet nature of the mitral valve. The posterior mitral valve leaflet has a crescentic shape. The anterior mitral valve leaflet is adjacent to the aortic valve and these structures are in fibrous continuity. (B and BHW) An en face anatomically orientated view from the ventricular aspect. The most inferior structure is the left ventricle inferior wall, which corresponds to the inferomedial mitral valve papillary muscle. The left ventricular outflow tract (LVOT) lies at ~10 o'clock using a clock face analogy. This demonstrates the mitral valve orientation within the left ventricle, its relationship to the IVS and LVOT, and the bileaflet nature of the mitral valve. The curved nature of the zone of coaptation can be appreciated. This projection can permit visualisation of the subleaflet apparatus depending on the cropping proximity to the leaflet edge during

postprocessing. (C) A 'chordal projection' which is unique to RT3DE and cannot be achieved using cross-sectional echocardiography. During postprocessing, the cut plane is placed parallel to the plane of the anterior mitral valve leaflet. This permits visualisation of the anterior mitral valve leaflet (AMVL) and the aortic valve, but its primary purpose is to demonstrate the anatomy of the lateral and medial papillary muscles and chordal attachments to the mitral valve. In this example, the length of the lateral and medial papillary muscle can be visualised in its entirety and it is clear that there are two papillary muscles. This view is not strictly in anatomical orientation due to its unique perspective. This view is valuable to show the primary and secondary chords, two papillary muscles, and their position within the left ventricle. Sup, superior; Inf, inferior; L, left; R, right; Ant MVL, anterior mitral valve leaflet; Post MVL, posterior mitral valve leaflet; PM, papillary muscle; LA, left atrium; LAA, left atrial appendage; RA, right atrium; LV, left ventricle; IAS, interatrial septum; IVS, interventricular septum; AoV, aorta valve; TV, tricuspid valve; Lat MV Marg, lateral mitral valve margin; Med MV Marg, medial mitral valve margin.

tendinae from the medial and lateral aspects of the mitral valve into a single papillary muscle (1, 18). Stenosis is the common functional outcome, due to a variable combination of underdeveloped chordae limiting leaflet movement, along with significantly reduced interchordal spaces limiting secondary orifice function (18, 19, 20, 21). This lesion is usually found in association with other anatomical abnormalities representing multilevel left heart obstruction (6, 18, 20).

2DE infers a parachute mitral valve through visualisation of a single papillary muscle on a parasternal short-axis view. Colour and pulse wave Doppler on 2DE demonstrates the functional consequence. 2DE cannot give the same information as RT3DE on the chordal apparatus, and its relationship to the papillary muscle or the reduced interchordal spaces. The minimal effective orifice area is also more precisely localised on RT3DE. RT3DE transthoracic acquisition is best from a parasternal

Video 1

RT3DE transthoracic atrial en face normal mitral valve. Download Video 1 via <http://dx.doi.org/10.1530/ERP-15-0003-v1>.

Video 2

RT3DE transthoracic ventricular en face normal mitral valve. Download Video 2 via <http://dx.doi.org/10.1530/ERP-15-0003-v2>.

long- or short-axis or an apical four-chamber view. Transoesophageal acquisition is best from four-chamber mid-oesophageal view at 0° and ~120°. Figure 2A shows the cropping plane to best show the subvalvar apparatus from a parasternal long-axis acquisition. In Fig. 2B and Video 3, the entirety of the subleaflet apparatus can be identified including short and thickened chords attaching both leaflets of the mitral valve to a single papillary muscle.

Mitral valve arcade

Arcade mitral valve features are demonstrated in Fig. 3A, B and C and corresponding figures and Videos 4 and 5. The hallmark of this lesion is a bridge of fibrous tissue, which arches between both papillary muscles, along the line of closure of the mitral leaflets (22, 23). This 'bridge' may directly attach to the free leaflet edge, or be separated from the leaflet by short, thickened chordae. These short thickened chords are attached to papillary muscles, which are typically elongated. The predominant functional consequence is valve regurgitation, but it may be mixed with stenosis (19, 22, 23). The mechanism of this is thought to be abnormal leaflet excursion and underdeveloped commissures (19, 22). The subvalvar abnormalities result in degrees of obliteration of interchordal spaces, causing the stream to eccentrically divert through remaining spaces. This is visualised as colour inflow 'splaying' on 2DE imaging (6, 23). The features of an arcade mitral valve are again alluded to, but not fully represented on 2DE due to the lack of depth of field. The best views for RT3DE acquisition are the same as for a parachute mitral valve. Chordal projections are shown in Fig. 3B and C modified with the anterior leaflet cropped away. They demonstrate the extent of the papillary muscle extension, the absence of chordae and loss of interchordal spaces that cannot be projected on a 2DE image. The abnormal leaflet excursion is demonstrated in Videos 4 and 5.

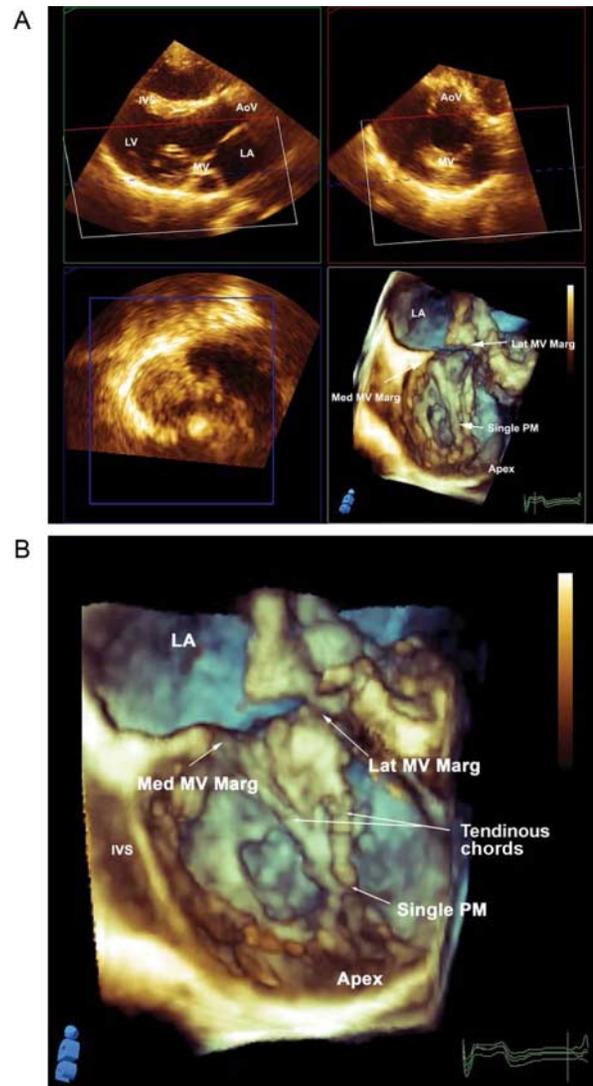


Figure 2

(A) The cropping orientation which is used to visualise the chordal support of the mitral valve. The valve is viewed as if from the red dotted line looking into the box defined by the white margins. This example shows that both the lateral and medial aspects of the mitral valve have chordal attachments, which insert into a single papillary muscle, which is the defining feature of the parachute mitral valve. (B) An enlarged rendered image of the parachute mitral valve. The depth of field and unique cropping plane permit this to be visualised in a single projection in contrast to cross-sectional echocardiography. LA, left atrium; MV, mitral valve; Lat MV Marg, lateral mitral valve margin; Med MV Marg, medial mitral valve margin; IVS, interventricular septum; single PM, single papillary muscle; LV, left ventricle; AoV, aortic valve.

Video 3

RT3DE transthoracic chordal view of a parachute mitral valve. Download Video 3 via <http://dx.doi.org/10.1530/ERP-15-0003-v3>.

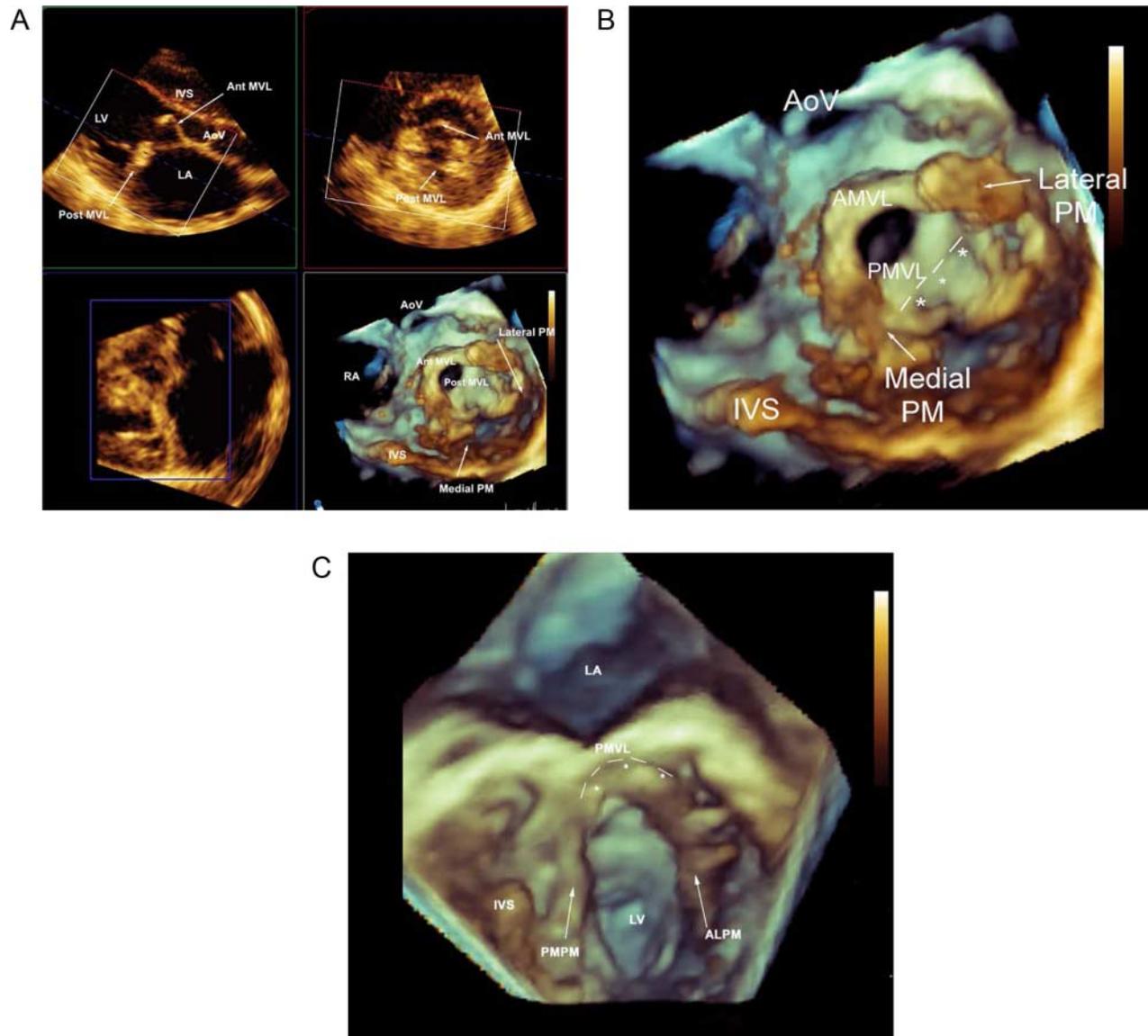


Figure 3

A chordal projection of a mitral valve arcade. (A) The whole mitral valve apparatus, including anterior and posterior mitral valve leaflets. The thickened nature of the posterior mitral valve leaflet and eccentric motion of the anterior mitral valve leaflet is best appreciated in the upper left pane. (B) An enlarged rendered RT3DE chordal view tilted slightly upwards to show that the papillary muscle attachment is direct into the posterior mitral valve leaflet. The symbol (*) indicates the posterior leaflet mitral valve arcade bridge. (C) Cropping away the anterior mitral valve leaflet

shows the anatomy more clearly. This shows the direct papillary muscle attachment to the posterior leaflet tip as a 'bridge' with no chordal presence and totally obliterated interchordal channels. LA, left atrium; LV, left ventricle; LVOT, left ventricular outflow tract; IVS, interventricular septum; Ant MVL, anterior mitral valve leaflet; Post MVL, posterior mitral valve leaflet; (P)M PM, medial papillary muscle; (A)L PM, lateral papillary muscle; RA, right atrium.

Isolated cleft mitral valve

A typical anterior cleft mitral valve is shown in Fig. 4A and B (Videos 6 and 7). A true cleft in the mitral valve is morphologically distinct from the anatomy of the left AV valve in the context of an atrioventricular septal defect (AVSD), even if the term 'cleft' has been loosely applied to

the zone of apposition between the superior and inferior bridging leaflets in the context of an AVSD. Major differences between AVSD and true cleft in the mitral valve are summarised in Table 1 (24, 25, 26). Chordal attachment of the anterior mitral valve leaflet to the septum is frequent with a true cleft in the mitral valve and

Video 4

RT3DE tilted transthoracic chordal view of an arcade mitral valve. Download Video 4 via <http://dx.doi.org/10.1530/ERP-15-0003-v4>.

Video 5

RT3DE transthoracic chordal view of an arcade mitral valve posterior apparatus. Download Video 5 via <http://dx.doi.org/10.1530/ERP-15-0003-v5>.

can lead to LVOT obstruction (26, 27). Cleft mitral valves can be isolated or associated with other cardiac abnormalities. Where ventricular septal defects (VSD) are present

Video 6

RT3DE transoesophageal atrial en face view of a cleft mitral valve. Download Video 6 via <http://dx.doi.org/10.1530/ERP-15-0003-v6>.

Video 7

RT3DE transoesophageal ventricular en face view of a cleft mitral valve. Download Video 7 via <http://dx.doi.org/10.1530/ERP-15-0003-v7>.

in association with a true cleft of the mitral valve, differentiation from an AVSD is important as the surgical intervention timing, repair technique and longer-term outcome may differ between these two morphologies (26, 27, 28). The vast majority of mitral valve cleft affect the anterior leaflet and congenital posterior mitral leaflet clefts are very rare (0.11%). They may have counter-clockwise papillary muscle rotation, are strongly associated with other mitral valve abnormalities and the majority are positioned in the mid P2 scallop (29, 30).

Isolated mitral valve clefts can be missed on 2DE even on parasternal short-axis view, which is the most helpful view for this lesion. They are even harder to detect on 2DE, where the cleft does not extend all the way to the valve annulus. The best transthoracic acquisition views for RT3DE of cleft mitral valves include the parasternal short-axis or, in infants, the short-axis subcostal view. On transoesophageal imaging, the four-chamber mid-oesophageal view at 0° is usually optimal. Other views include the ~120° view from which Fig. 4 and Videos 6 and 7 were acquired and transgastric views at 0°. Figure 4 and Videos 6 and 7 show the cleft orientation towards the LVOT and that the cleft does not extend to the full length of the anterior mitral valve leaflet. The posterior leaflet of the mitral valve has a more normal morphology than the mural leaflet of the AVSD.

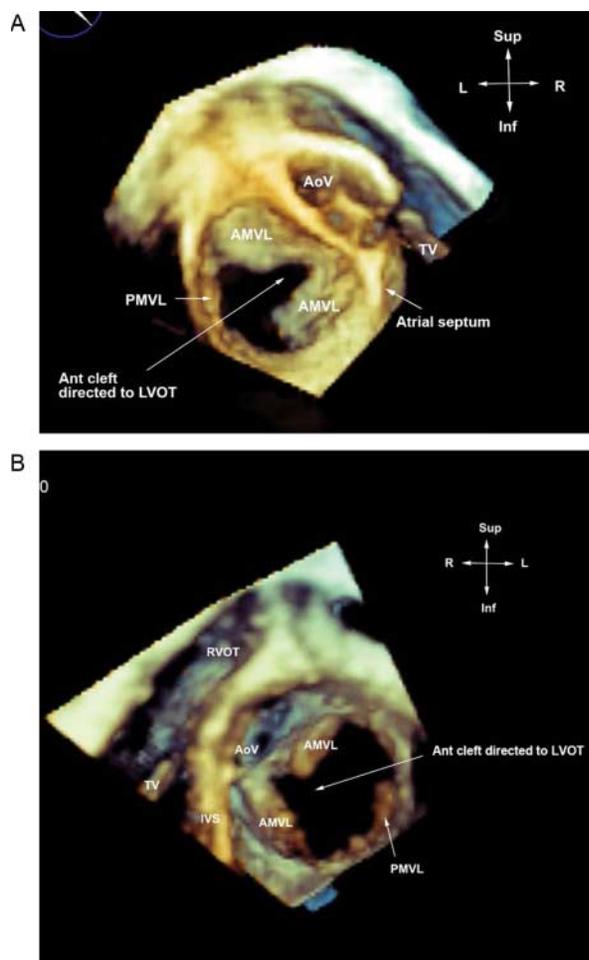


Figure 4

(A) A cleft mitral valve viewed from an atrial perspective and (B) ventricular perspective on transoesophageal echocardiogram. This orientation of the anterior cleft is towards the LVOT. The posterior (mural) leaflet is of near normal size. The cleft does not extend to the full length of the valve leaflet. Note that the crescentic shape of the posterior mitral valve leaflet is retained. AMVL, anterior mitral valve leaflet; PMVL, posterior mitral valve leaflet; AoV, aortic valve; TV, tricuspid valve; Sup, superior; Inf, inferior; L, left; R, right; Ant, anterior.

Double-orifice mitral valve

An example of a double-orifice mitral valve (DOMV) is represented in both Fig. 5 and Video 8. This abnormality is characterised by a single fibrous annulus with two orifices that open into the left ventricle. DOMV forms around 1% of congenital heart disease and is predominantly associated with other cardiac defects (31). As Fig. 5 reflects, DOMV has been occasionally found incidentally in cases of intrinsic cardiomyopathy (32). DOMV manifest with regurgitation in 43% of cases, normal function in 37%, with stenosis less likely (31).

Table 1 Comparison of morphological features of an anterior cleft mitral valve vs an AVSD left AV valve.

AV valve morphology	Offset of AV valves	Orientation of 'cleft'	Bridging of valvar leaflets	Posterior leaflet	LV papillary muscle position
Anterior cleft mitral valve	Yes, usually	Towards LVOT (predominantly)	No	Normal usually	Typically normal
AVSD left AV valve	No (common AV junction)	Towards ventricular septum	Yes	Mural leaflet smaller, triangular	Rotated counter-clockwise

2DE parasternal and subcostal short-axis views have defined this lesion into three types (33). The first is 'complete bridge' type where the orifices are visible fully throughout on sweeping through annulus to leaflet tip. These orifices are usually circular and relatively equal size with a complete tissue bridge between them (Fig. 5). The second is 'incomplete bridge type' with the tissue connection between the anterior and posterior leaflets (and second orifice) visualised distally only at the leaflet edges and not at midleaflet or annulus level. This type is thus only seen in a more apically directed view and can be easily missed on 2DE imaging. A further 'hole' type is predominantly seen in AVSD settings, therefore of left

AV valve type. The second orifice is usually small and can be only visualised at a mid-leaflet level tucked close to the commissure. RT3DE in this setting provides information about the relationship of the orifices, their anatomical character and function (34). It also provides better assessment of the subleaflet apparatus, which is frequently abnormal (35).

Non-mitral left AV valve lesions

Atrioventricular septal defect

A typical unrepaired AVSD is shown via a RT3DE transthoracic view from an atrial and ventricular aspect in Fig. 6A and B (Videos 9 and 10). The hallmark of an AVSD is a common AV junction with a common AV valve. In this lesion, the left AV valve is formed by superior and inferior bridging leaflets, which 'bridge' between the left and right ventricles. The third leaflet of the left AV valve is the mural leaflet, which is smaller and has a more triangular shape than the normal posterior mitral valve leaflet. This group of lesions accounts for ~3% of congenital heart disease (36). A broad morphological spectrum exists, with variability of the relationship and size of the leaflets and the interaction of the bridging leaflets with the atrial and ventricular septum (37). In most cases, there are communications between both the ventricle and atrium, when the lesion is termed 'complete'. The term 'partial' AVSD is commonly applied to the situation where there is no communication between the ventricles. The common AV junction means that the aorta cannot assume its normal position wedged between the AV valves and the aorta is displaced or 'unwedged' anteriorly. This is due to the intrinsic AV septation abnormality, which results in

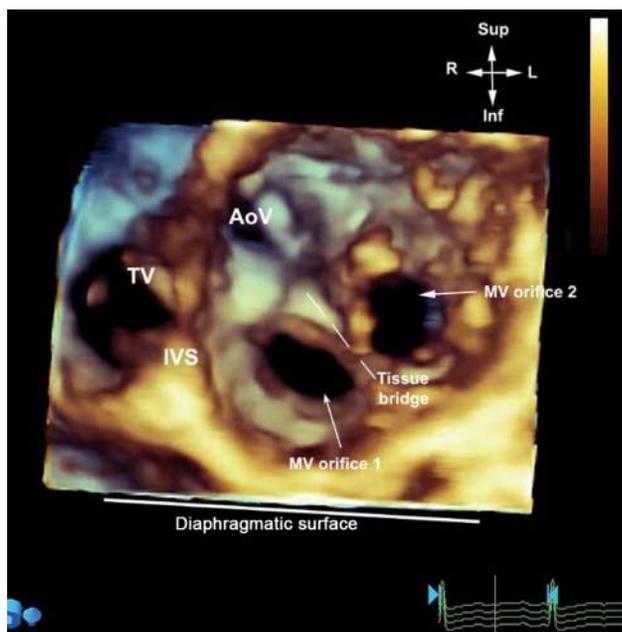


Figure 5

This is a transthoracic apical four-chamber acquisition showing a complete bridge type double-orifice mitral valve from a ventricular aspect showing the two orifices of equal size, both of mitral morphology, with a constant bridge of tissue between the orifices. MV orifice 1, mitral valve orifice 1; MV orifice 2, mitral valve orifice 2; AoV, aortic valve; TV, tricuspid valve; IVS, interventricular septum; Sup, superior; Inf, inferior; L, left; R, right.

Video 8

RT3DE transthoracic ventricular en face view of a double-orifice mitral valve. Download Video 8 via <http://dx.doi.org/10.1530/ERP-15-0003-v8>.

ventricular inlet–outlet disproportion, variability of the size and relationship of the AV valve leaflets and their attachments, and an altered position of the AV node (38). AVSD left AV valve morphology is distinct from mitral

Video 9

RT3DE transoesophageal atrial en face view of an unrepaired partial AVSD. Download Video 9 via <http://dx.doi.org/10.1530/ERP-15-0003-v9>.

Video 10

RT3DE transoesophageal ventricular en face view of an unrepaired partial AVSD. Download Video 10 via <http://dx.doi.org/10.1530/ERP-15-0003-v10>.

Video 11

RT3DE transoesophageal ventricular en face view of a previously repaired AVSD left AV valve with incomplete closure of bridging leaflet interface. Download Video 11 via <http://dx.doi.org/10.1530/ERP-15-0003-v11>.

due to the common AV junction. There is clockwise rotation of the papillary muscles (26) and an altered inlet-to-outlet ratio of to the left ventricle.

2DE demonstrates the loss of normal AV valve offset, suggests a trileaflet left AV valve and shows a zone of apposition directed towards the inlet septum. Sometimes erroneously called a ‘cleft’, this is the interface between the superior and inferior bridging leaflets. RT3DE can outline the left AV valve anatomy in greater detail, which is particularly valuable where an AV valve repair is required during a primary AVSD repair (39). Shunts at atrial and ventricular level can be obvious on 2DE imaging, but where shunts are smaller, or with complex AV valvar attachments, 2DE assessment be challenging. RT3DE may provide more information in this context. The best views for acquisition are parasternal long and short axis, apical four-chamber and in smaller children subcostal. The predominant transoesophageal view is a four-chamber mid-oesophageal view at 0°. Occasionally,

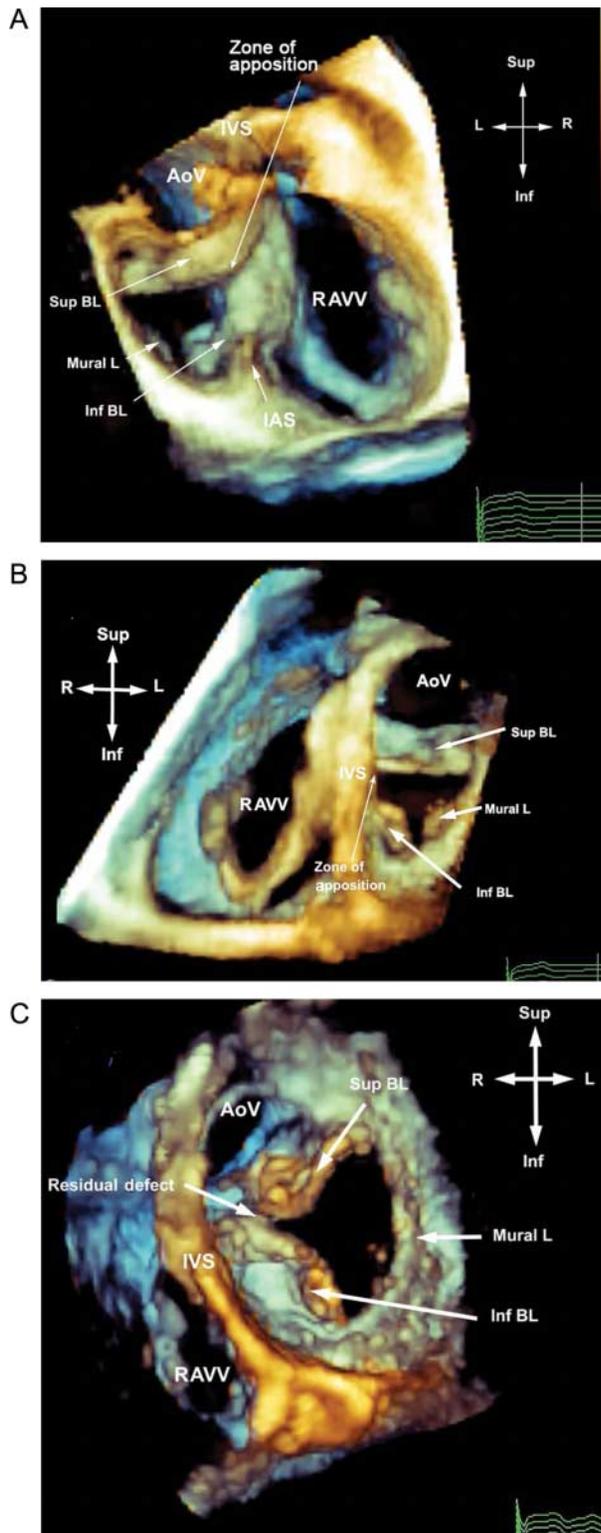


Figure 6

(A and B) An unrepaired partial AVSD via transoesophageal echocardiography is shown from an atrial (A) and ventricular (B) aspect. The interface of the superior and inferior bridging leaflets is seen orientated towards the IVS along with the un-wedged anterior aortic position and a left AV valve small triangular mural leaflet. The bridging of the superior leaflet across the ventricular septum is clearly visualised in A. (C) A live 3D acquisition of a previously repaired partial AVSD with a residual defect between the superior and inferior bridging leaflet contributing to significant left AV valve regurgitation. Sup BL, superior bridging leaflet; Inf BL, inferior bridging leaflet; Mural L, mural leaflet; AoV, aortic valve; RAVV, right AV valve; IVS, interatrial septum; IVS, interventricular septum; Sup, superior; Inf, inferior; L, left; R, right.

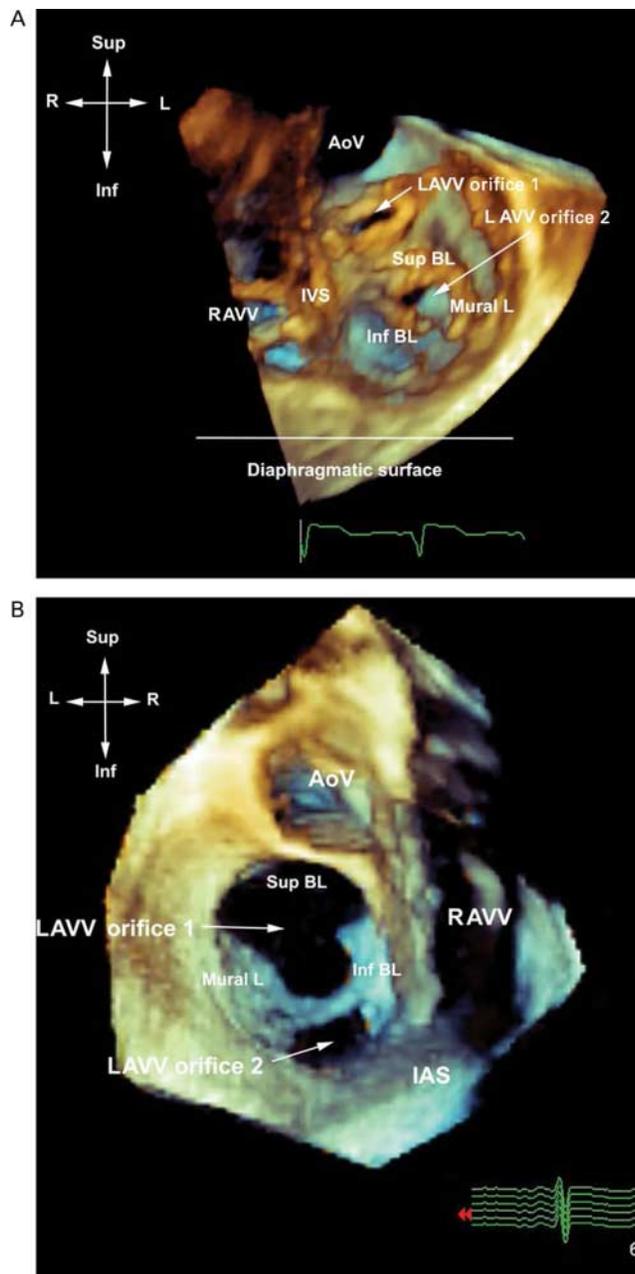


Figure 7
(A) A live RT3DE transthoracic parasternal short-axis view acquisition of double-orifice left AV valve in a patient with partial AVSD. The two orifices are in a superior/inferior orientation. Both are of similar size. (B) A transoesophageal echocardiogram acquisition of a double-orifice left AV valve from the atrial aspect in the context of an AV septal defect. This is of 'hole' type with the second orifice close to the interface of the superior and inferior bridging leaflets and only shown at the level of the leaflet tips. AV, atrioventricular; LAVV orifice 1, left AV valve orifice 1; LAVV orifice 2, left AV valve orifice 2; Sup BL, superior bridging leaflet; Inf BL, inferior bridging leaflet; Mural L, mural leaflet; AoV, aortic valve; RAVV, right AV valve; IVS, interventricular septum; IAS, interatrial septum; Sup, superior; Inf, inferior; L, left; R, right.

Video 12

RT3DE transthoracic ventricular en face view of a partial AVSD with equal orifice double-orifice left AV valve. Download Video 12 via <http://dx.doi.org/10.1530/ERP-15-0003-v12>.

Video 13

RT3DE transoesophageal atrial en face view of a 'hole' type orifice double-orifice left AV valve in a partial AVSD. Download Video 13 via <http://dx.doi.org/10.1530/ERP-15-0003-v13>.

the $\sim 120^\circ$ view of the mitral valve with the LVOT in view can also be helpful for acquisition.

Left AV valve regurgitation remains the most common aetiology (3–10% of cases) for late surgical reintervention after primary AVSD repair and carries important morbidity and mortality (40, 41, 42). Establishing the mechanism of left AV valve regurgitation can be enhanced using RT3DE. Figure 6C and Video 11 show incomplete closure of the superior and inferior bridging leaflet interface after a previous partial AVSD repair.

Double-orifice left AV valve

Figure 7 and Videos 12 and 13 show a double-orifice left AV valve (DOLAVV) in two patients with partial AVSDs. Figure 7A reflects a complete tissue bridge between the two orifices that are of similar size. Figure 7B demonstrates a RT3DE 'hole' type of DOLAVV (33). On 2DE short-axis views, the additional smaller orifice is only visible at mid-leaflet level postero-inferiorly near to the interface of the bridging leaflets. Viewing at an annular level, 2DE only demonstrates the superior–inferior bridging leaflet zone of apposition. DOLAVV can be acquired post surgical repair as a complication or an intentional reparative technique (43).

Other left sided non-mitral valves

Figure 8 and Video 14 shows a left-sided tricuspid valve on transthoracic echocardiography from the ventricular aspect. With usual atrial arrangement and discordant AV connections, the left-sided AV valve is the tricuspid valve connecting to a systemic right ventricle. Salient distinguishing features of a tricuspid valve are the leaflet configuration of an anterior superior, an inferior and a septal leaflet with attachments of subvalvar apparatus to the septum as well as the free wall of the ventricle. An increased association of Ebstein's anomaly of the

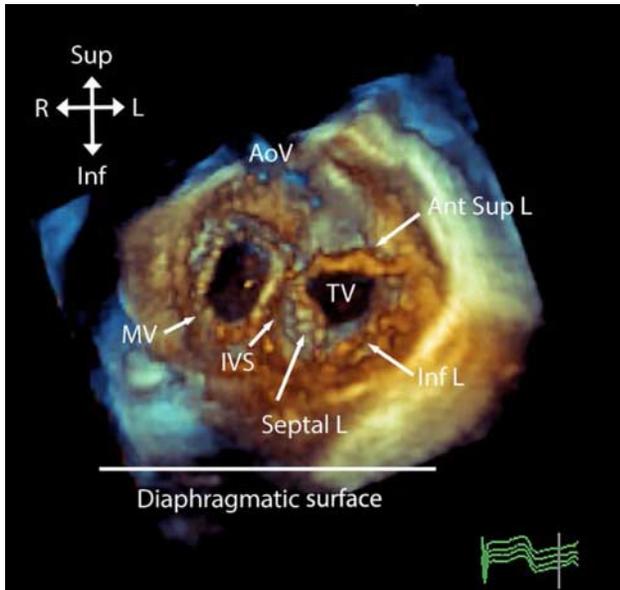


Figure 8

A transthoracic view of a left-sided tricuspid valve in the setting of discordant AV connections. This shows that the left AV valve has the characteristic trileaflet arrangement and septal attachment of a tricuspid valve and that the right AV valve is of mitral type. AV, atrioventricular; Ant Sup L, anterior superior leaflet; Inf L, inferior leaflet; Septal L, septal leaflet; MV, mitral valve; IVS, interventricular septum; AoV, aortic valve; TV, tricuspid valve; Sup, superior; Inf, inferior; L, left; R, right.

Video 14

RT3DE transthoracic ventricular en face view of a left-sided tricuspid valve in atrial situs with AV and ventriculoarterial discordant connections. Download Video 14 via <http://dx.doi.org/10.1530/ERP-15-0003-v14>.

tricuspid valve, VSD, pulmonary stenosis and complete heart block exists in settings of combined AV and ventriculoarterial discordant connections. 2DE is adequate for distinguishing ventricular morphology including the AV valve in simple cases, but becomes more difficult in complex settings particularly where there is ventricular rotation. In addition, the morphological mitral valve may not always be normal with discordant AV connections. This prompts closer assessment of the mitral valve complex, which may be enhanced by RT3DE (44). In Fig. 8, RT3DE clearly demonstrates the trileaflet configuration and septal attachment of the tricuspid valve on the left and the bileaflet mitral valve on the right. It also shows the relationship of the AV valves with a VSD in this example.

Discussion and conclusions

1. This review emphasises the wide range of congenital abnormalities of the left AV valve, which may be encountered in clinical practice.
2. RT3DE may be a useful complementary technique to standard 2D and colour flow-Doppler assessment of such valves. RT3DE can be achieved in a real clinical workflow scenario.
3. 2D echo still provides the highest temporal and spatial resolution, which is important for fast-moving structures such as valves and chordal support apparatus.
4. RT3DE provides a depth of field so that
 - i) The 'big picture' anatomy may be appreciated.
 - ii) The full depth of leaflet morphology may be appreciated.
 - iii) Chords and papillary muscles can be visualised along their entire length.
 - iv) The true effective orifice can be appreciated via the ability to crop valves deep into the ventricle.
5. Although not the subject of this review, areas of mitral valve regurgitation may be identified by RT3DE and the mechanism of regurgitation fully appreciated.

Declaration of interest

The authors declare that there is no conflict of interest that could be perceived as prejudicing the impartiality of this review.

Funding

This review did not receive any specific grant from any funding agency in the public, commercial or not-for-profit sector.

References

- 1 Banerjee A, Kohl T & Silverman NH 1995 Echocardiographic evaluation of congenital mitral valve anomalies in children. *American Journal of Cardiology* **76** 1284–1291. (doi:10.1016/S0002-9149(99)80357-9)
- 2 Grenadier E, Sahn DJ, Valdes-Cruz LM, Allen HD, Oliveira Lima C & Goldberg SJ 1984 Two-dimensional echo Doppler study of congenital disorders of the mitral valve. *American Heart Journal* **107** 319–325. (doi:10.1016/0002-8703(84)90381-8)
- 3 Gilbert BW, Schatz RA, VonRamm OT, Behar VS & Kisslo JA 1976 Mitral valve prolapse. Two-dimensional echocardiographic and angiographic correlation. *Circulation* **54** 716–723. (doi:10.1161/01.CIR.54.5.716)
- 4 Lang RM, Mor-Avi V, Sugeng L, Nieman PS & Sahn DJ 2006 Three-dimensional echocardiography: the benefits of the additional dimension. *Journal of the American College of Cardiology* **48** 2053–2069. (doi:10.1016/j.jacc.2006.07.047)

- 5 Mor-Avi V, Sugeng L & Lang RM 2009 Real-time 3-dimensional echocardiography: an integral component of the routine echocardiographic examination in adult patients? *Circulation* **119** 314–329. (doi:10.1161/CIRCULATIONAHA.107.751354)
- 6 Asante-Korang A, O'Leary PW & Anderson RH 2006 Anatomy and echocardiography of the normal and abnormal mitral valve. *Cardiology in the Young* **16** (S3) 27–34. (doi:10.1017/S1047951106000734)
- 7 Séguéla P-E, Houyel L & Acar P 2011 Congenital malformations of the mitral valve. *Archives of Cardiovascular Diseases* **104** 465–479. (doi:10.1016/j.avcd.201.06.004)
- 8 Simpson JM 2008 Real-time three-dimensional echocardiography of congenital heart disease using a high frequency paediatric matrix transducer. *European Journal of Echocardiography* **9** 222–224. (doi:10.1016/j.euje.2007.06.012)
- 9 Simpson JM & Miller O 2011 Three-dimensional echocardiography in congenital heart disease. *Archives of Cardiovascular Diseases* **104** 45–56. (doi:10.1016/j.avcd.2010.11.004)
- 10 Lang RM, Badano LP, Tsang W, Adams DH, Agricola E, Buck T, Faletra FF, Franke A, Hung J, de Isla LP *et al* 2012 EAE/ASE recommendations for image acquisition and display using three-dimensional echocardiography. *European Heart Journal Cardiovascular Imaging* **13** 1–46. (doi:10.1093/ehjci/er316)
- 11 Vettukattil JJ 2011 Three dimensional echocardiography in congenital heart disease. *Heart* **98** 79–88. (doi:10.1136/heartjnl-2011-300488)
- 12 Simpson JM 2011 Congenital heart disease in children. In *Three-Dimensional Echocardiography*, 1st edn, pp 201–221. Eds T Buck, A Franke & MJ Monaghan. Berlin, Heidelberg: Springer.
- 13 Simpson J, Miller O, Bell A, Bellsham-Revell H, McGhie J & Meijboom F 2011 Image orientation for three-dimensional echocardiography of congenital heart disease. *International Journal of Cardiovascular Imaging* **28** 743–753. (doi:10.1007/s10554-011-9893-3)
- 14 Anderson RH, Razavi R & Taylor AM 2004 Cardiac anatomy revisited. *Journal of Anatomy* **205** 159–177. (doi:10.1111/j.0021-8782.2004.00330.x)
- 15 Anderson RH & Kanani M 2007 Mitral valve repair: critical analysis of the anatomy discussed. *Multimedia Manual of Cardiothoracic Surgery* **2007** mmcts.2006.002147.
- 16 Ho SY 2002 Anatomy of the mitral valve. *Heart* **88** (Suppl 4) iv5–iv10. (doi:10.1136/heart.88.suppl_4.iv5)
- 17 Obadia JF, Casali C, Chassignolle JF & Janier M 1997 Mitral subvalvular apparatus: different functions of primary and secondary chordae. *Circulation* **96** 3124–3128. (doi:10.1161/01.CIR.96.9.3124)
- 18 Shone JD, Sellers RD, Anderson RC, Adams P, Jr, Lillehei CW & Edwards JE 1963 The developmental complex of “parachute mitral valve,” supravalvular ring of left atrium, subaortic stenosis, and coarctation of aorta. *American Journal of Cardiology* **11** 714–725. (doi:10.1016/0002-9149(63)90098-5)
- 19 Davachi F, Moller JH & Edwards JE 1971 Diseases of the mitral valve in infancy an anatomic analysis of 55 cases. *Circulation* **43** 565–579. (doi:10.1161/01.CIR.43.4.565)
- 20 Schaverien MV 2004 Independent factors associated with outcomes of parachute mitral valve in 84 patients. *Circulation* **109** 2309–2313. (doi:10.1161/01.CIR.0000129269.98460.C8)
- 21 Marino BS, Kruge LE, Cho CJ, Tomlinson RS, Shera D, Weinberg PM, Gaynor JW & Rychik J 2008 Parachute mitral valve: morphologic descriptors, associated lesions, and outcomes after biventricular repair. *Journal of Thoracic and Cardiovascular Surgery* **137** 385–3e4. (doi:10.1016/j.jtcvs.2008.09.016)
- 22 Layman TE & Edwards JE 1967 Anomalous mitral arcade. A type of congenital mitral insufficiency. *Circulation* **35** 389–395. (doi:10.1161/01.CIR.35.2.389)
- 23 Hakim FA, Krishnaswamy C & Mookadam F 2013 Mitral arcade in adults – a systematic overview. *Echocardiography* **30** 354–359. (doi:10.1111/echo.12126)
- 24 Smallhorn JF, de Leval M, Stark J, Somerville J, Taylor JF, Anderson RH & Macartney FJ 1982 Isolated anterior mitral cleft. Two dimensional echocardiographic assessment and differentiation from “clefts” associated with atrioventricular septal defect. *British Heart Journal* **48** 109–116. (doi:10.1136/hrt.48.2.109)
- 25 Sigfússon G, Etedgui JA, Silverman NH & Anderson RH 1995 Is a cleft in the anterior leaflet of an otherwise normal mitral valve an atrioventricular canal malformation? *Journal of the American College of Cardiology* **26** 508–515. (doi:10.1016/0735-0197(95)80030-K)
- 26 Kohli T & Silverman NH 1996 Comparison of cleft and papillary muscle position in cleft mitral valve and atrioventricular septal defect. *American Journal of Cardiology* **77** 164–169. (doi:10.1016/S0002-9149(96)90589-5)
- 27 Fraisse A, Massih TA, Kreitmann B, Metras D, Vouhé P, Sidi D & Bonnet D 2003 Characteristics and management of cleft mitral valve. *Journal of the American College of Cardiology* **42** 1988–1993. (doi:10.1016/j.jacc.2003.07.019)
- 28 Abadir S, Fouilloux V, Metras D, Ghez O, Kreitmann B & Fraisse A 2009 Isolated cleft of the mitral valve: distinctive features and surgical management. *Annals of Thoracic Surgery* **88** 839–843. (doi:10.1016/j.athoracsur.2009.06.004)
- 29 Wyss CA, Enseleit F, van der Loo B, Grünenfelder JR, Oechslin EN & Jenni R 2009 Isolated cleft in the posterior mitral valve leaflet: a congenital form of mitral regurgitation. *Clinical Cardiology* **32** 553–560. (doi:10.1002/clc.20608)
- 30 Kent SM, Markwood TT, Vernalis MN & Tighe JF, Jr 2001 Cleft posterior mitral valve leaflet associated with counterclockwise papillary muscle malrotation. *Journal of the American Society of Echocardiography* **14** 303–304. (doi:10.1067/mje.2001.110140)
- 31 Zalstein E, Hamilton R, Zucker N, Levitas A & Gross GJ 2004 Presentation, natural history, and outcome in children and adolescents with double orifice mitral valve. *American Journal of Cardiology* **93** 1067–1069. (doi:10.1016/j.amjcard.2004.01.015)
- 32 Sugiyama H, Hoshiai M, Toda T & Nakazawa S 2006 Double-orifice mitral valve associated with noncompaction of left ventricular myocardium. *Pediatric Cardiology* **27** 746–749. (doi:10.1007/s00246-006-1014-z)
- 33 Trowitzsch E, Baño-Rodrigo A, Burger BM, Colan SD & Sanders SP 1985 Two-dimensional echocardiographic findings in double orifice mitral valve. *Journal of the American College of Cardiology* **6** 383–387. (doi:10.1016/S0735-1097(85)80176-5)
- 34 Anwar AM, McGhie JS, Meijboom FJ & Ten Cate FJ 2008 Double orifice mitral valve by real-time three-dimensional echocardiography. *European Journal of Echocardiography* **9** 731–732. (doi:10.1093/ejechocard/jen149)
- 35 Baño-Rodrigo A, Van Praagh S, Trowitzsch E & Van Praagh R 1988 Double-orifice mitral valve: a study of 27 postmortem cases with developmental, diagnostic and surgical considerations. *American Journal of Cardiology* **61** 152–160. (doi:10.1016/0002-9149(88)91322-7)
- 36 Calabrò R & Limongelli G 2006 Complete atrioventricular canal. *Orphanet Journal of Rare Diseases* **1** 8. (doi:10.1186/1750-1172-1-8)
- 37 Anderson RH, Wessels A & Vettukattil JJ 2010 Morphology and morphogenesis of atrioventricular septal defect with common atrioventricular junction. *World Journal for Pediatric & Congenital Heart Surgery* **1** 59–67. (doi:10.1177/2150135109360813)
- 38 Adachi I, Uemura H, McCarthy KP & Ho SY 2008 Surgical anatomy of atrioventricular septal defect. *Asian Cardiovascular & Thoracic Annals* **16** 497–502. (doi:10.1177/021849230801600616)
- 39 Takahashi K, Mackie AS, Rebeyka IM, Ross DB, Robertson M, Dyck JD, Inage A & Smallhorn JF 2010 Two-dimensional versus transthoracic real-time three-dimensional echocardiography in the evaluation of the mechanisms and sites of atrioventricular valve regurgitation in a congenital heart disease population. *Journal of the American Society of Echocardiography* **23** 726–734. (doi:10.1016/j.echo.2010.04.017)
- 40 Hoohenkerk GJ, Wenink AC, Schoof PH, Koolbergen DR, Bruggemans EF, Rijlaarsdam M & Hazekamp MG 2009 Results of surgical repair of atrioventricular septal defect with double-orifice left atrioventricular valve. *Journal of Thoracic and Cardiovascular Surgery* **138** 1167–1171. (doi:10.1016/j.jtcvs.2009.05.012)

- 41 Stulak JM, Burkhart HM, Dearani JA, Cetta F, Barnes RD, Connolly HM & Schaff HV 2010 Reoperations after repair of partial atrioventricular septal defect: a 45-year single-center experience. *Annals of Thoracic Surgery* **89** 1352–1359. (doi:10.1016/j.athoracsur.2010.01.018)
- 42 Kaza AK, Colan SD, Jagers J, Lu M, Atz AM, Sleeper LA, McCrindle BW, Lambert LM, Margossian R, Lacro RV *et al* 2011 Surgical interventions for atrioventricular septal defect subtypes: the pediatric heart network experience. *Annals of Thoracic Surgery* **92** 1468–1475 (discussion 1475). (doi:10.1016/j.athoracsur.2011.04.109)
- 43 Macé L, Dervanian P, Houyel L, Chaillon-Fracchia E, Piot D, Lambert V, Losay J & Neveux J-Y 2001 Surgically created double-orifice left atrioventricular valve: a valve-sparing repair in selected atrioventricular septal defects. *Journal of Thoracic and Cardiovascular Surgery* **121** 352–365. (doi:10.1067/mtc.2001.111969)
- 44 Uemura H, Adachi I, McCarthy KP, Koh M & Ho SY 2008 Is the morphologic mitral valve in discordant atrioventricular connections always normal? *Annals of Thoracic Surgery* **86** 1328–1333. (doi:10.1016/j.athoracsur.2008.06.041)

Received in final form 14 January 2015

Accepted 16 January 2015