The right ventricle in congenital heart disease

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Heart 2006;**92**(Suppl I):i27–i38. doi: 10.1136/hrt.2005.077438

In patients with congenital heart disease the right ventricle (RV) may support the pulmonary (subpulmonary RV) or the systemic circulation (systemic RV). During the last 50 years evidence is accumulating that RV dysfunction develops in many of these patients and leads to considerable morbidity and mortality. Therefore RV function in certain groups of congenital heart disease patients needs close surveillance and timely and appropriate intervention to optimise outcomes. Despite major progress being made, assessing the RV either in the subpulmonary or the systemic circulation remains challenging, often requiring a multi-imaging approach and expertise (echocardiography, magnetic resonance imaging, nuclear and occasionally invasive assessment with angiography). This review discusses the implications of volume and pressure loading of the RV in the context of congenital heart disease and describes the most relevant imaging modalities for monitoring RV function.

The exploration and understanding of the heart's morphology, physiology, and function both in health and disease remains a challenging and still evolving field. Modern imaging modalities, mainly echocardiography, but also radionuclide imaging and lately computed tomography (CT) and cardiac magnetic resonance (CMR), have revolutionised clinical research on biventricular anatomy and function.[1–6] However, there are still numerous questions to be answered regarding left and right ventricular function and their contributions to cardiovascular disease prognosis.

Although LV function and dysfunction and its relationship to prognosis have been studied extensively, the role of RV morphology, function, and dysfunction in cardiovascular disease has not attracted the interest of scientists until recently. This was largely due to the fact that most acquired cardiovascular diseases affect primarily the left ventricle. Additionally, the RV has a very complex shape, which makes precise in vivo imaging and assessment challenging for most imaging modalities.

RV morphology and function is, however, of paramount importance in the rapidly growing field of congenital heart disease (CHD). Many CHD patients have become adolescents and adults thanks to major advances of paediatric cardiology and cardiac surgery in the latter half of the last century. This has created a patient population (adult CHD patients) in which the RV is often the centre of attention.[7–11] Such patients are unique models for the study of RV physiology and function. Accurate assessment of RV anatomy, volume, and ejection fraction in CHD, both before and after reparative surgery, requires one or more of the following imaging modalities: echocardiography, contrast angiography, radionuclide studies, CT, and or CMR.[12]

RIGHT VENTRICULAR MORPHOLOGY AND IMAGING

Echocardiography is the imaging modality of choice for the assessment of left ventricular function. However, most quantitative two dimensional echocardiographic measurements of ventricular performance are based on geometric assumptions that do not apply to the RV. The left ventricle is more conical in shape and has a wall thickness 3–4 times greater than the RV free wall.[13] RV trabeculations are coarse compared to the finely trabeculated left ventricle and the RV outflow tract is muscular and elongated, ending up in the pulmonary valve which does not have a real valvar annulus.[14] These differences in ventricular morphology reflect the genetically determined different role the two ventricles are called to play in the circulation.[15] Sir Magdi Yacoub described the left ventricle as a ''flask'' shape with the inlet and outlet sharing one orifice, enabling it to deliver a bolus of blood against high resistance, and the RV as a flattened tube wrapped around the left ventricle with separate inlet and outlet orifices and a presumed contraction pattern simulating peristalsis, an arrangement suited for pumping blood against low resistance.[16]

Having these important differences in mind we will briefly refer to imaging methods for the evaluation of RV anatomy and function and then examine the RV and the applicability of these methods in two broad contexts: the volume loaded RV, and the pressure loaded RV.

Angiographic assessment of the RV is invasive, involves ionising radiation and use of contrast agents, and is not as accurate as CMR[12] (fig 1). It used to be the gold standard for RV evaluation in the early era of imaging, but has largely been replaced by newer ''non-geometric'' techniques like three dimensional (3D) echocardiography, CMR, and multislice CT (MSCT), which permit accurate assessment of RV volume, mass and function.[2–4 17–20]

Indirect insights into RV systolic and diastolic function are given by conventional Doppler indices such as the duration of systolic time intervals derived by interrogation of the RV outflow[1] and Doppler recordings of the tricuspid inflow and hepatic venous flow.[1 21] M mode and tissue Doppler imaging examine myocardial velocities and time intervals, detectable at the level of the tricuspid annulus, as markers of RV systolic and diastolic longitudinal motion[1] (fig 2). Diastolic tricuspid annular velocities, in contrast to inflow velocities, correlate with invasively determined RV pressures.[22 23] Nongeometric and load independent, Doppler derived quantitative indices of global ventricular function, like the myocardial performance index (Tei index),[24 25] or tricuspid annular isovolumic acceleration,[26–28] may prove useful for evaluation of RV function. However, they correlate weakly with echocardiographic RV ejection fraction[29] and they have not been validated against quantitative methods of evaluation of RV function like CMR.[22 25 30] Transoesophageal echocardiography (TOE) has better sensitivity and specificity for evaluation of CHD compared to transthoracic echocardiography, but it is

Abbreviations: ASD, atrial septal defect; ccTGA, congenitally corrected transposition of the great arteries; CHD, congenital heart disease; CMR, cardiac magnetic resonance; CT, computed tomography; MRI, magnetic resonance imaging; MSCT, multislice computed tomography; PR, pulmonary valve regurgitation; RNA, radionuclide angiography; RV, right ventricle; TOE, transoesophageal echocardiography; TR, tricuspid regurgitation

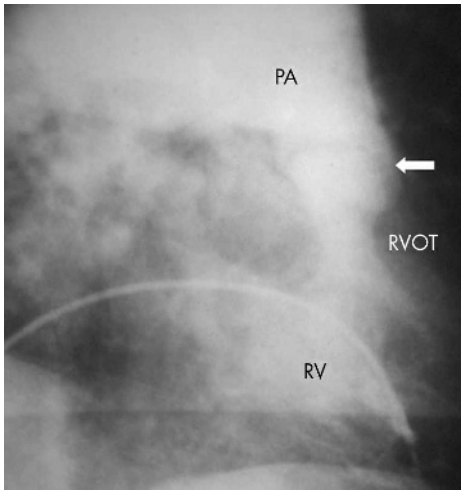

Figure 1 Right ventricular angiography, right anterior oblique (RAO) projection, end systole. Severe pulmonary valve stenosis. There is thickening and doming of the pulmonary valve (white arrow), secondary narrowing of the right ventricular outflow tract due to muscle hypertrophy, and poststenotic dilatation of the pulmonary artery. PA, pulmonary artery; RV, right ventricle; RVOT, right ventricular outflow tract.

semi-invasive, not well suited for evaluation of an anteriorly positioned RV, and requires special skills.[31]

Radionuclide angiography provides a reliable quantitative measurement of ventricular function that is not based upon assumptions of ventricular geometry, and its value in the routine clinical measurement of ventricular function is well established.[12 32] However, it requires the acquisition of views of the ventricles that exclude counts from other chambers, which can usually be achieved for the left ventricle, but often not satisfactorily for the RV.[33] Radionuclide imaging uses ionising radiation, although the radiation dose is low compared to cineangiography.[33] Additionally this modality requires an adequate bolus injection for first pass studies, and a regular rhythm with minimal R-R variability.[12] Its resolution is poor compared to more modern imaging methods. Finally, investigation of CHD has focused more on structural rather than functional abnormalities, although this is changing. Radionuclide imaging has, thus, been of limited use to date.[33]

Rapid advancements in the field of CMR have established this technique as the gold standard for quantitative assessment of RV volume, mass, and function regardless of its position in the thorax (subpulmonary *v* systemic RV).[4 18 20 34] Spin echo (black blood) sequences are used for exploration of anatomy and gradient echo (white blood) sequences for assessment of RV function.[35] Flow velocity mapping allows for accurate assessment of valvar regurgitation (regurgitant fraction) and magnetic angiography for assessment of great vessel anatomy.[36] CMR with late gadolinium enhancement can detect myocardial fibrosis in both ischaemic and non-ischaemic cardiomyopathies.[37 38] This technique has now been applied to CHD and is likely to make an important contribution to our understanding of the pathophysiology of RV dysfunction.[39] However, CMR has also limitations. It usually requires breath-holding, a regular heart rhythm, exclusion of patients with implantable metallic devices, and it has high cost with low availability at present.[39] MSCT is emerging as an alternative modality, especially for patients with implantable devices (contraindication for CMR); however, MSCT uses ionising radiation and requires a low heart rate for image acquisition.[2 3]

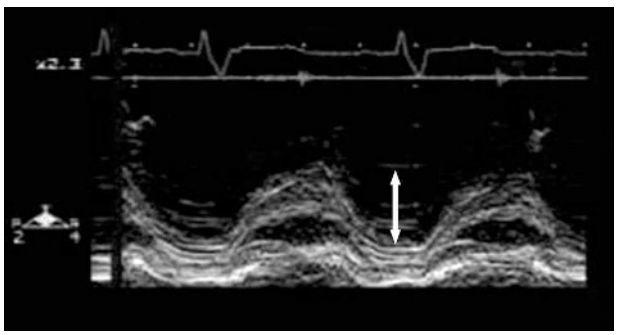

Figure 2 M mode echocardiogram of the lateral (free wall) tricuspid valve annulus. The height of the annular movement (white double arrow) is a surrogate marker of RV systolic function.

THE VOLUME LOADED RV

Three of the most common lesions associated with RV volume loading will be examined: atrial septal defect, significant pulmonary valve regurgitation, and significant tricuspid regurgitation.

Atrial septal defect (ASD)

There are three major types of ASDs: ostium secundum, ostium primum, and superior sinus venosus defect.[40] An isolated ASD results in left-to-right shunting, which when significant, leads to right atrial/ventricular and pulmonary arterial dilatation. These features are often evident on chest *x* ray (postero-anterior and lateral). Transthoracic echocardiography is invaluable for diagnosing an ASD and assessing its impact on RV size and function[40 41] (figs 3–5). TOE may be needed to diagnose a sinus venosus defect and assist in assessment of pulmonary venous drainage.[40–42] Paradoxical septal motion as a result of RV volume loading is evident both in M mode and 2D echocardiograms in most patients[41 43 44] (fig 6).

The RV tolerates volume loading well for a long time.[40] Although delayed RV contraction has been detected with radionuclide studies (in the absence of conduction defects),[45] echocardiographic assessment has shown RV systolic and diastolic function to be normal or exaggerated.[46 47] In older

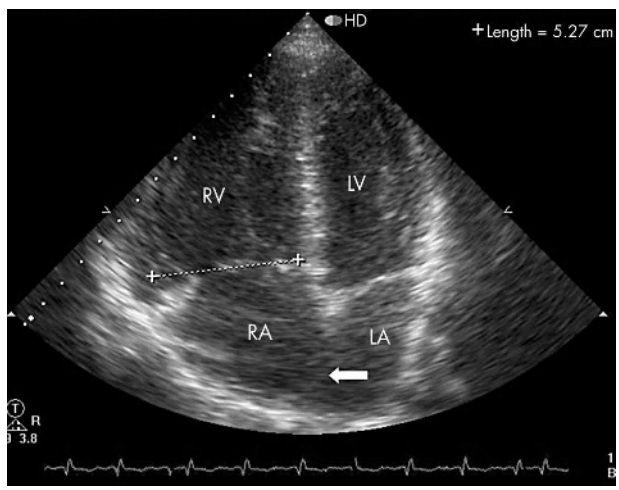

Figure 3 Two dimensional echocardiogram, apical four chamber view. There is dilatation and hypertrophy of the right ventricle, which is larger than the left ventricle. The right ventricular inflow diameter (dotted line), measures 5.27 cm (normal adult diameter < 4 cm). A large atrial septal defect (ASD) is present (white arrow). LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

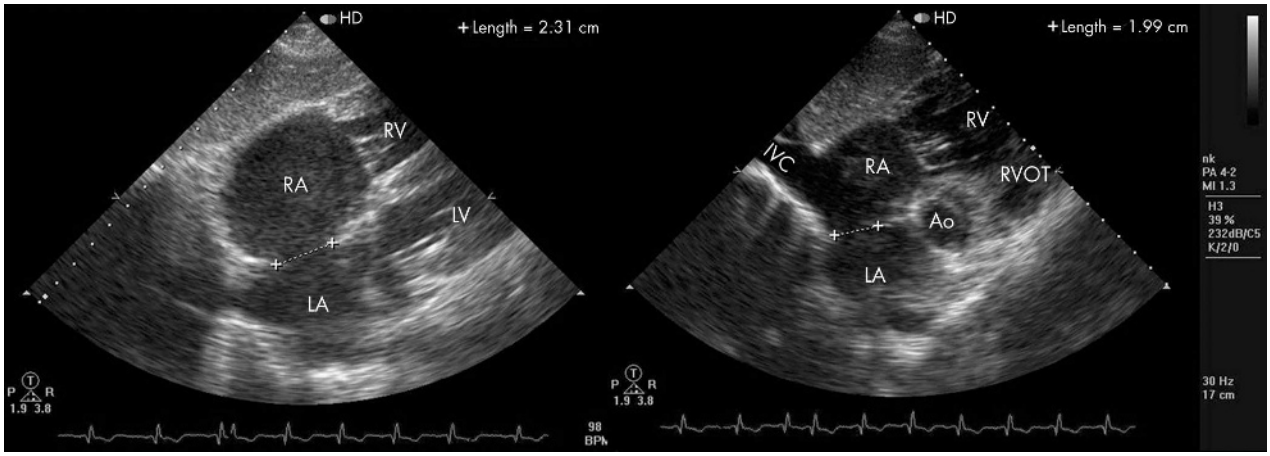

Figure 4 Two dimensional echocardiogram. Left panel: Subcostal four chamber view. There is a large (23 mm) ostium secundum ASD (dotted line). Right panel: Same patient in the left panel. Subcostal short axis view at the level of the great vessels. The relation of the ASD with the aortic root can be assessed. Ao, aortic root; IVC, inferior vena cava; LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle, RVOT, RV outflow tract.

patients with long standing volume overload, regional RV tissue Doppler imaging may disclose early relaxation abnormalities even with normal traditional tricuspid inflow velocities.[47]

It is now accepted that long standing right heart, pulmonary arterial and venous volume overload and dilatation in the setting of an ASD is detrimental and leads to morbidity (heart failure, arrhythmia, and thromboembolic events) and increased mortality.[42 48] These can all be reversed to variable degrees with catheter or surgical closure of the defect.[48 49] However, atrial arrhythmias may persist or develop in adults repaired after the age of 40 years.[48 50–53] Indeed, RV/RA remodelling is incomplete in the older patient undergoing transcatheter closure.[54] Therefore early defect closure is warranted if a significant shunt (with right heart dilatation) is present.[10 40 48]

Transcatheter ASD device occlusion has become the treatment of choice for most secundum ASDs. While many devices are being used for this purpose, the Amplatzer ASD occluder (AGA Corporation, Golden Valley, Minnesota, USA) is most widely used at present[55 56] (fig 7, 8). With appropriate patient selection, device closure in adults leads to symptomatic improvement and increased exercise capacity even in asymptomatic patients and is associated with fewer complications and shorter hospitalisation times compared to surgery.[57 58] However, very long term results are lacking at present.

Dilatation of the RV may not subside after ASD closure,[59] in some patients up to five years after repair.[60] Others have reported progressive normalisation of RV size during 1–24 months after surgical or device closure.[54 61 62] Atrial ''shrinkage'' is inversely proportional to age at repair and is related to the potential for atrial arrhythmias after late defect closure.[53 61] In patients with normal diastolic function, increased RV myocardial diastolic and systolic velocities (tissue Doppler) return to normal within one month after device closure.[47] In older patients, however, with abnormal relaxation myocardial velocities seem to be volume independent and do not change after device closure, suggesting altered myocardial structure and function.[47] CMR and MSCT are seldom needed in post-repair follow up (fig 8).

Pulmonary regurgitation (PR)

Isolated clinical PR is a rare problem, but not an innocent one.[63] Severe PR is very common after tetralogy of Fallot repair and is associated with RV dysfunction, diminished

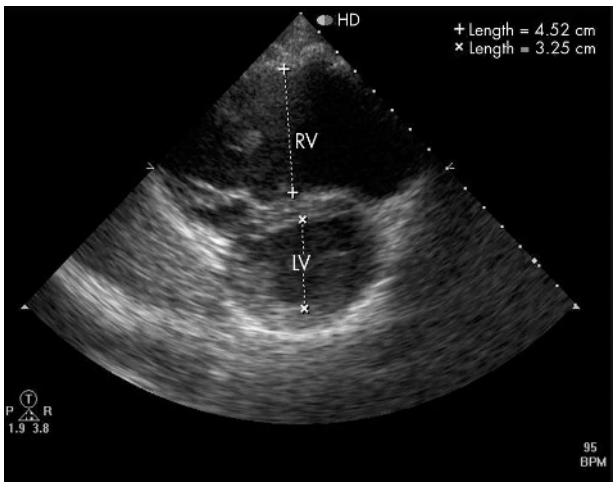

Figure 5 Two dimensional transthoracic echocardiogram, left parasternal short axis view. There is RV dilatation and hypertrophy in a young adult patient with an ostium secundum ASD and a Qp/Qs of 3.2. The left ventricle is ''squashed'' by the dilated RV. LV, left ventricle; RV, right ventricle.

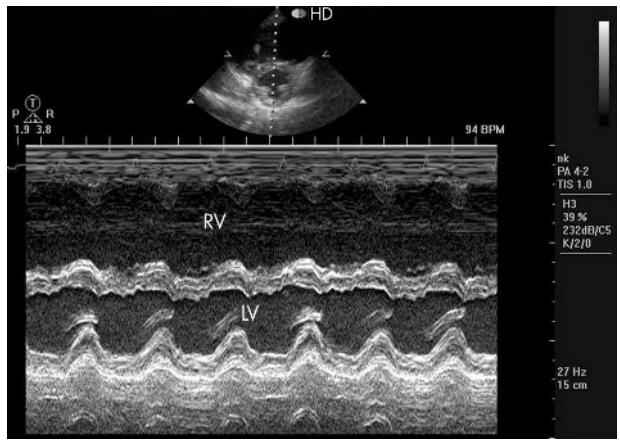

Figure 6 M mode echocardiogram of the same patient shown in fig 5. There is paradoxical septal motion, a sign of RV volume overload. RV enlargement relative to the left ventricle is evident. LV, left ventricle; RV, right ventricle.

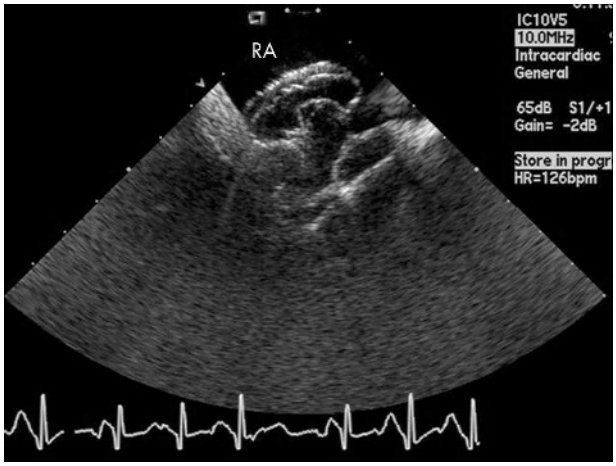

Figure 7 Intracardiac echocardiogram of a patient with an ostium secundum ASD just after apposition of an Amplatzer septal occluder umbrella device. The intravascular echocardiographic probe is located in the right atrium. Both sides of the umbrella and its main axis traversing the ASD are clearly visualised. RA, right atrium.

exercise capacity, atrial and ventricular arrhythmias, and sudden death. Timely pulmonary valve replacement may protect patients from PR related complications.[34 64–69] Therefore, serial quantitative assessment of PR and RV function are key to management. Echocardiography remains the most widely employed imaging modality (fig 9). However, CMR is considered the gold standard for both PR quantification (flow velocity mapping) and RV volumetric analysis (gradient echo)[18 34 70] (figs 9 and 10). Doppler echocardiography is a useful alternative for semiquantitive PR assessment as a new Doppler index (PR index: ratio of PR duration to diastolic duration) correlates well with the MRI derived pulmonary regurgitant fraction.[71] A PRi less than 0.77 yields 100% sensitivity and 85% specificity for identifying patients with a PR fraction $> 24.5\%$—that is, patients with significant PR.[71] A PR pressure half time < 100 ms has also been found to be a reliable indicator of haemodynamically significant regurgitation.[72]

Doppler detection of forward and laminar late diastolic pulmonary blood flow, coinciding with atrial systole, present throughout respiration, and associated with a prominent retrograde superior vena caval flow, defines the so called ''restrictive RV physiology''.[73] A non-compliant, usually hypertrophied RV, along with low pulmonary arterial diastolic pressures, results in partial presystolic opening of the pulmonary valve during right atrial contraction, which contributes to forward flow (fig 11). This physiology is commonly present early after tetralogy of Fallot repair, where it is associated with a low cardiac output (despite normal biventricular systolic function), leading to longer intensive care stay.[74 75] In contrast, restrictive RV physiology late after repair of tetralogy counteracts the effects of chronic pulmonary regurgitation and is associated with smaller RV size, shorter QRS duration, and better exercise capacity.[73 76–80]

Pronounced RV dilatation, especially if serial imaging demonstrates progression, may prompt referral for pulmonary valve replacement before RV dysfunction ensues. Thus, serial follow up of RV volumes—ideally with CMR—is recommended.[67–69 81 82] Following pulmonary valve replacement, RV volume usually decreases as evidenced by echocardiography,[83] radionuclide angiography (RNA),[84] or CMR.[68 85] However, there are contradictory reports on RV function[68 81 85] after pulmonary valve replacement, largely due to different timing of reoperation,[81 86] different imaging modalities employed and different parameters being measured,[68 81 86] presence of RV outflow aneurysms or akinesia[34 86] (fig 12), and variable re-evaluation intervals post-pulmonary valve replacement.[85 87]

Tricuspid regurgitation (TR)

Congenital TR may be primary, due to a malformed tricuspid valve, as exemplified by isolated tricuspid valve dysplasia or prolapse and Ebstein's anomaly or Ebstein's-like anomaly in patients with congenitally corrected transposition (in which case the tricuspid valve represents the systemic atrioventricular valve).[88] However, it is more frequently secondary, due to severe RV enlargement with resultant tricuspid annular dilatation as happens in patients with RV dysplasia, or free pulmonary regurgitation usually in the context of repaired tetralogy of Fallot.[88]

Ebstein's anomaly is a complex congenital heart malformation, characterised by an apical displacement of both the septal and the posterior tricuspid leaflets, exceeding 20 mm or 8 mm/m^2 in adults.[89] As a consequence, the right heart is divided in three components: the true right atrium, the functional RV, and an intervening zone that is anatomically ventricular but functionally right atrial (atria-

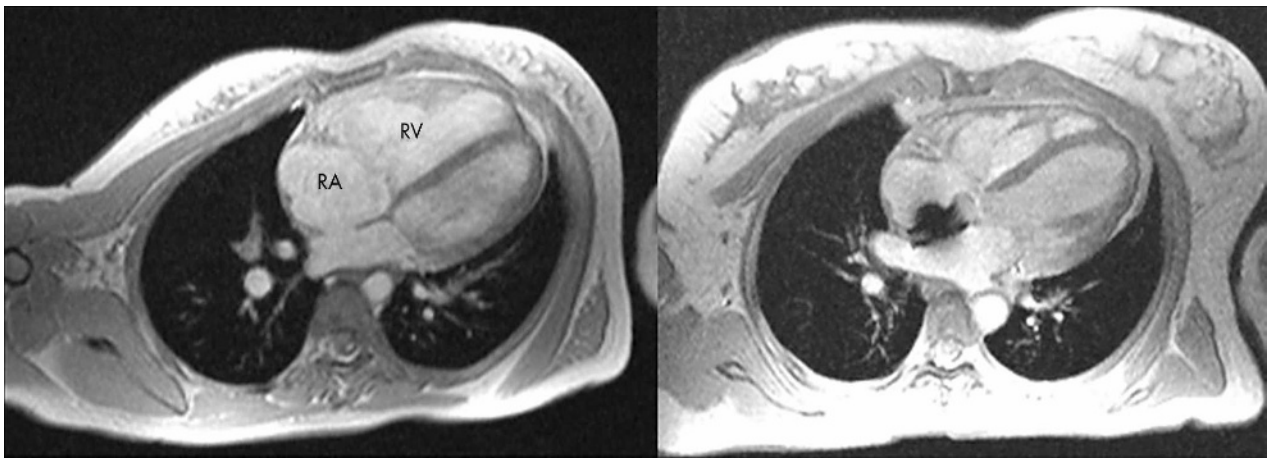

Figure 8 Gradient echo (white blood), magnetic resonance sequence. Four chamber transaxial (short axis of the thorax) view of a patient with an ostium secundum ASD. The defect is clearly seen on the left panel, along with right atrial/ventricular dilatation. On the right panel an Amplatzer septal occluder sealing the defect is seen, along with significant reduction of RV and atrial size six months after defect closure. RA, right atrium; RV, right ventricle.

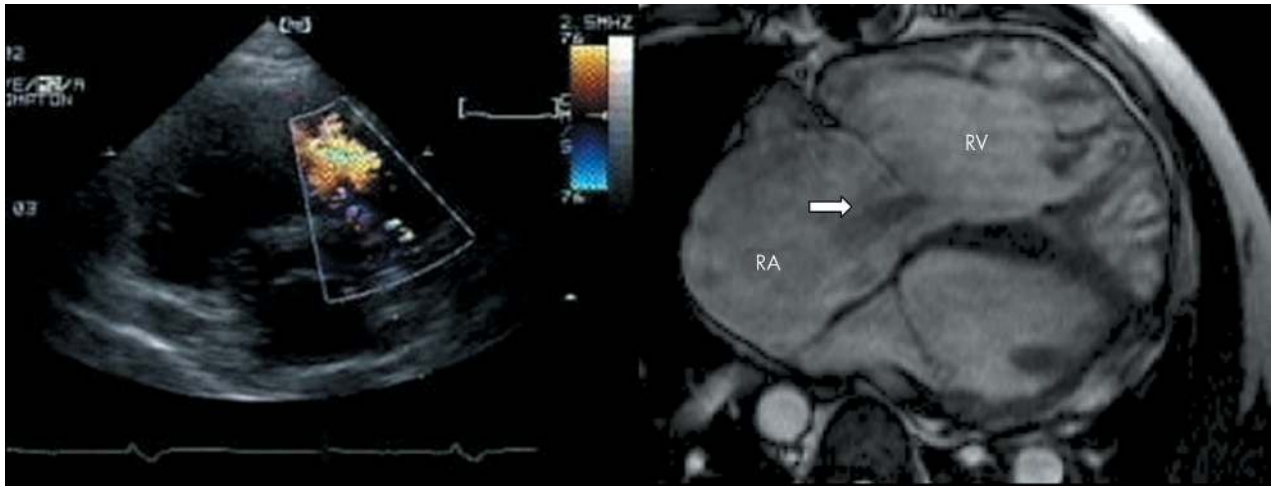

Figure 9 Left panel: Echocardiographic colour flow mapping (short axis left parasternal view), showing a pulmonary valve regurgitation jet. Right panel: gradient echo (white blood) cardiac magnetic resonance (CMR) sequence of the same patient, four chamber transaxial (short axis of the thorax) view. There is severe right atrioventricular dilatation and RV hypertrophy. Tricuspid regurgitation is depicted as signal void (black coloured jet of blood) at the level of the tricuspid valve towards the right atrium (white arrow). RA, right atrium; RV, right ventricle.

lised RV). The malformation results in moderate to severe TR and may be accompanied by pulmonary stenosis and an ASD with bidirectional shunting, which have a great impact on RV haemodynamics. All these features are adequately assessed with 2D and Doppler echocardiography. When other cardiac lesions are absent (for example, severe pulmonary stenosis), Ebstein's anomaly may be diagnosed in adolescence or adulthood, due to innocent murmurs or arrhythmias, with good long term outcome.[90] When severe, TR leads to RV volume loading and in the long term RV or biventricular dysfunction. Echocardiography may provide some information regarding the size, shape, and function of the functional RV, however CMR is best suited for a detailed study of the above features.[91 92]

Surgery should be performed for symptomatic adults.[93–95] Classical repair of Ebstein's anomaly is usually performed with transverse plication of the atrialised chamber and tricuspid valvoplasty if feasible, or tricuspid valve replacement.[93] However, with severely compromised RV or biventricular function, or in the presence of a relatively hypoplastic and/or malfunctioning RV chamber inadequate to sustain the entire systemic venous return but capable of managing part of the systemic venous return, a one and a half ventricular repair (superior cavopulmonary anastomosis) may provide good functional results.[96] Therefore a detailed preoperative assessment of RV size and function, but also of the valve leaflet attachments, commissures, and surface is mandatory. The latter cannot be achieved easily with 2D echocardiography; however, newer 3D echocardiographic techniques provide excellent intracardiac views of the valve commissures and leaflets' surface.[97] We would submit that for a complete evaluation of the right heart anatomy and function, combining echocardiography (2D, 3D, and transoesophageal) with CMR would be the best option in such cases.

In patients with repaired tetralogy of Fallot, TR is related to RV dilatation due to severe PR and possibly valvar trauma

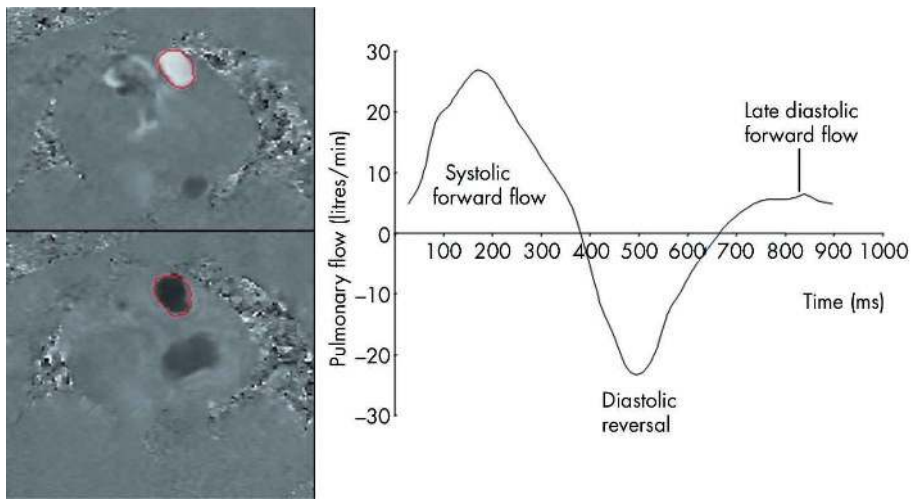

Figure 10 CMR flow velocity mapping of the pulmonary valve. Upper left panel: Forward flow through the valve (traced area) is encoded in white colour. Lower left panel: Backward flow through the pulmonary valve (pulmonary regurgitation) is encoded in black colour. Right panel: Pulmonary flow curve. The area under the curve represents flow. Forward flow is represented by the curve above the reference (zero) line. Backward flow is represented by the curve below the reference line. The last part of the curve above the reference line represents diastolic forward flow through the pulmonary valve, which suggests a restrictive RV. The pulmonary regurgitant fraction (PRF) is calculated as systolic forward flow–diastolic reversal/total flow. In this case the PRF is approximately 50%.

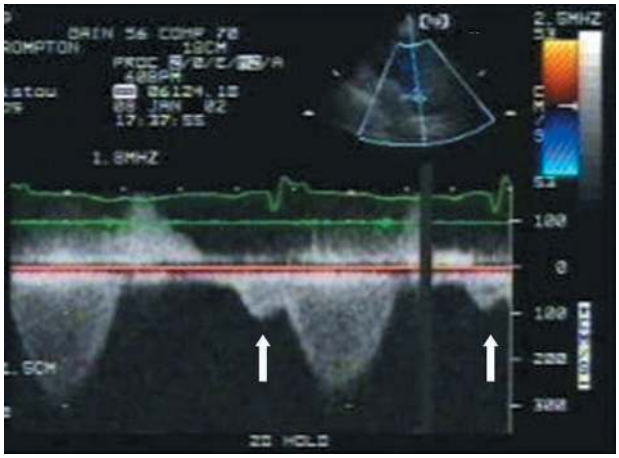

Figure 11 Continuous wave Doppler tracing of the RVOT of a patient with repaired tetralogy of Fallot and severe pulmonary regurgitation. There is residual pulmonary valve stenosis. The pulmonary regurgitation signal ends well before the next systolic signal, which suggests severe pulmonary regurgitation or the existence of a restrictive RV with high RV diastolic pressures, or both. There is a late diastolic signal of forward flow through the pulmonary valve (white arrows), suggesting the existence of a restrictive RV.

during reparative surgery.[98] When severe it contributes to further RV dilatation.[98] The existence of significant TR in such patients is considered an indication for pulmonary valve replacement.[99 100] However, when TR is severe, reoperation is associated with high surgical mortality and poor long term results due to postoperative RV dysfunction.[100 101] This supports the view that timely pulmonary valve replacement is mandatory before severe TR and RV dysfunction ensue.[102]

THE PRESSURE LOADED RV

Two major models exemplifying pressure loading of the RV will be discussed: RV outflow tract (RVOT) obstruction and the RV supporting the systemic circulation (systemic RV).

Right ventricular outflow tract obstruction–pulmonary stenosis

Isolated stenosis at the valvar level represents 80–90% of pulmonary stenosis cases.[14 103] However, obstruction may also occur at the subvalvar or supravalvar level. Regardless of the

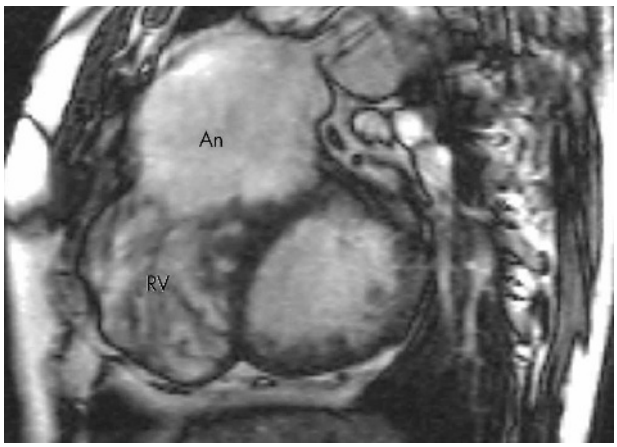

Figure 12 Gradient echo (white blood) CMR sequence, sagittal plane (RVOT view), end systole. The RV is severely dilated and hypertrophied. The upper part of the RV chamber, almost of same size with the main RV chamber below, represents a huge RVOT aneurysm. An, aneurysm; RV, right ventricle.

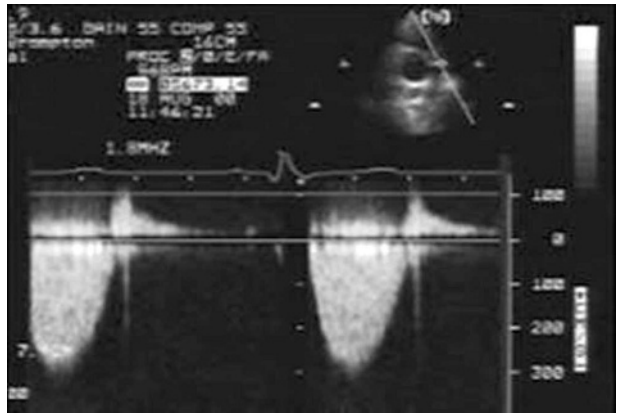

Figure 13 Continuous wave Doppler tracing of the RVOT in a patient with combined pulmonary valve stenosis and regurgitation. Note that the signal of pulmonary regurgitation (above the zero line) ends well before the next systolic Doppler wave (below the line). This may be either due to severe pulmonary regurgitation, or to elevated RV diastolic pressure.

level of obstruction, the RV exerts a hypertrophic response the degree of which varies with the magnitude of obstruction.[104] Echocardiography is the diagnostic method of choice.[103] Continuous wave Doppler is used for estimation of the pressure gradient across the RVOT[14] (fig 13). In contrast to left sided stenoses, the RVOT instantaneous gradient correlates well with catheter based peak-to-peak gradient, obviating the need for cardiac catheterisation.[105] The latter is saved for patients with long RVOT stenoses at the infundibular level where Doppler may be inaccurate. More sophisticated methods, mainly CMR, may be needed for detailed imaging of the RVOT and for assessment of RV size and function[103] (fig 14). This need is exemplified by more complex anomalies like the ''double chambered RV''.[106] The latter is a term used for anomalous muscle bundles that divide the RV into a high pressure apical chamber and a low pressure outlet/infundibular chamber[14 106] (fig 15).

The systemic right ventricle

In terms of physiology the RV is teleologically well suited for the changes in preload that normally occur with changes in intrathoracic pressure and systemic venous return and poorly tolerant of acute changes in afterload.[13] Fundamental anatomic and physiologic principles pose obvious disadvantages to the RV supporting the systemic circulation.

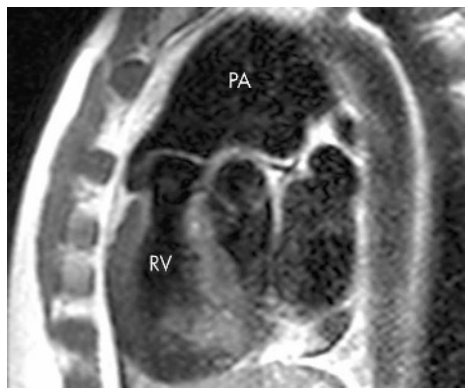

Figure 14 Spin echo (black blood) CMR sequence, sagittal plane (RVOT view), end systole. There is severe RV hypertrophy. The pulmonary valve is thickened and domed. The main pulmonary artery is severely dilated. PA, pulmonary artery; RV, right ventricle.

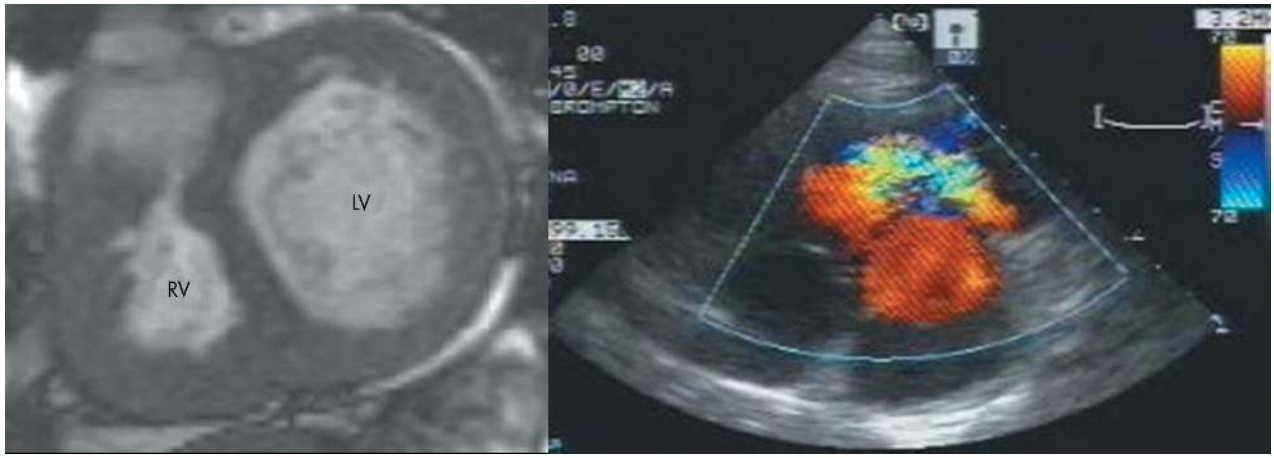

Figure 15 Left panel: Gradient echo (white blood) CMR sequence, oblique sagittal plane (short axis of the ventricles), end systole. The RV is divided into two chambers by a thick ring of ventricular myocardium. This anomaly is called ''double chambered RV''. Right panel: Echocardiographic colour flow mapping (short axis left parasternal view) of the same patient shown on the left. There is blood turbulence (aliasing) within the RV at the level of the hypertrophic muscle bands. LV, left ventricle; RV, right ventricle.

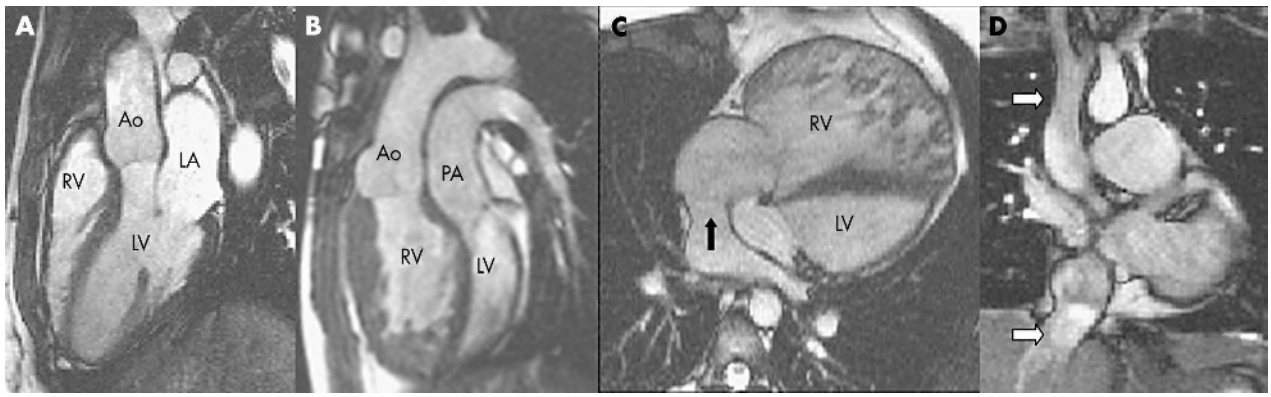

Figure 16 Gradient echo (white blood) CMR sequences. (A) Sagittal plane, normal cardiac anatomy. The left atrium is draining into the left ventricle, which is connected to the aorta. (B–D) Various CMR views of a patient with complete transposition of the great arteries and Mustard repair (atrial switch). (B) Sagittal plane. The left ventricle is connected to the pulmonary artery (subpulmonary left ventricle). The RV is connected to the aorta (systemic RV). The systemic RV is dilated and severely hypertrophied. (C) Four chamber view. The pulmonary venous pathway of the atrial switch is shown. The pulmonary veins (left lower pulmonary vein is clearly seen in this level) are draining into the right atrium through the baffle (arrow). The systemic RV is dilated and severely hypertrophied. (D) Coronal view. The systemic venous pathway of the atrial switch is shown. The inferior and superior vena cavae (arrows) are directed underneath the baffle to the left atrium. Ao, aorta; LA, left atrium; LV, left ventricle; PA, pulmonary artery; RV, right ventricle.

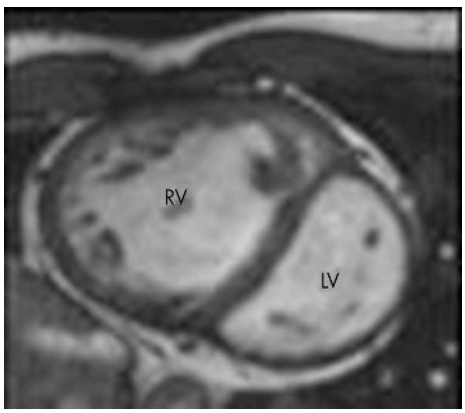

Figure 17 Gradient echo (white blood) CMR sequence, oblique sagittal (short axis of the ventricles view) of a patient with complete transposition of the great arteries and Senning (atrial switch) repair. The RV is severely hypertrophied and dilated. The interventricular septum is bowing towards the left ventricle. LV, left ventricle; RV, right ventricle.

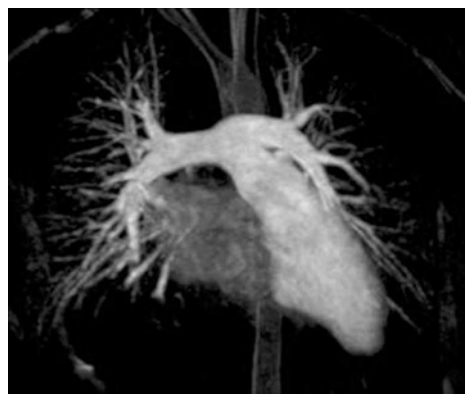

Figure 18 Magnetic resonance angiography (MRA) with gadolinium injection through a peripheral vein. Ventriculography, frontal projection. The chamber opacified demonstrates left ventricular characteristics (conical shape, smooth walls); however, it is connected to the pulmonary arteries, which are also opacified by gadolinium. This represents a subpulmonary left ventricle in a patient with complete transposition of the great arteries and Mustard repair (atrial switch). MRA allows for a detailed non-invasive assessment of the ventriculo-arterial connections and of the anatomy of the great vessels.

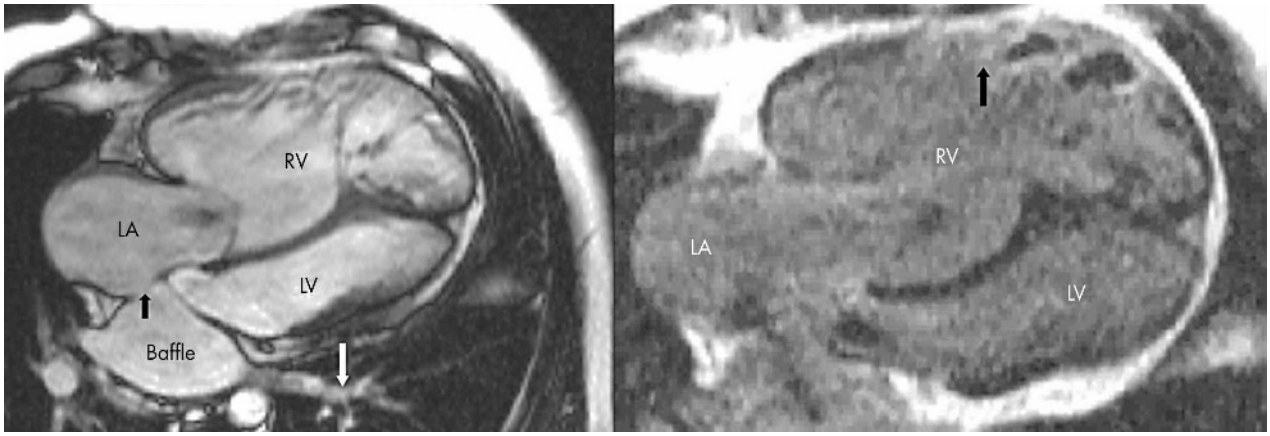

Figure 19 Left panel: Gradient echo sequence (white blood), short axis view of the RV and the pulmonary venous pathway in a patient with complete transposition of the great arteries and Mustard repair (atrial switch). The RV is dilated and hypertrophied. There is a signal void (black coloured jet) at the level of the tricuspid valve caused by tricuspid regurgitation. The pulmonary venous return finds its way back to the RV through a baffle (black arrow), which redirects blood from the pulmonary veins (white arrow) to the left atrium (LA). The RV is connected to the aorta (not shown) and supports the systemic circulation. The left ventricle is elongated and thin walled, and supports the pulmonary circulation. Right panel: The same patient imaged with a late gadolinium enhancement CMR sequence. Gadolinium is washed out from normal myocardium, which appears grey-black. Areas of necrosis and scarring demonstrate late gadolinium enhancement (white colour). There is gadolinium enhancement of an area of the RV free wall (black arrow), suggesting myocardial scarring. LA, left atrium; LV, left ventricle; RV, right ventricle.

Atrial switch operations (Mustard and Senning)

Complete transposition of the great arteries is incompatible with life without a surgical switch of the circulation either at atrial or great arterial level (physiologic or anatomic repair respectively). The former procedures are the Mustard and Senning operations, which have been performed for over 40 years now and have transformed the outlook for these patients[107] (figs 16–19). However, the atrial switch procedures result in the RV supporting the systemic circulation. Long term concerns remain: the prospect of RV failure, arrhythmias (atrial flutter variants and sick sinus syndrome), and accordingly compromised long term quality of life and survival for many of these patients. Assessment of RV function is paramount but also challenging, because of the inherent problems in assessing RV function (discussed above) and the absence of criteria for ''normal'' values.[108] Volumetric methods (echocardiography, RNA, CMR) have been the mainstay of RV assessment.[108] Cumulative survival 25–30 years after the Mustard repair is as high as 80%. However, it seems that there is progressive deterioration of RV function with time in most patients after the Mustard repair and this is often accompanied by significant systemic atrioventricular valve (tricuspid) regurgitation[109] (fig 19). This decline in RV function along with residual lesions (baffle obstruction or leakage, residual ventricular septal defect, and pulmonary valve stenosis), contribute to late morbidity and mortality manifested as reduced exercise capacity, heart failure, endocarditis, supraventricular arrhythmia, reoperation, and cardiac death.[109]

The cause of RV dysfunction is unclear, however. Myocardial perfusion defects[110] and impaired myocardial flow reserve in the systemic RV have been demonstrated in survivors of the Mustard operation, suggesting inadequate coronary blood supply.[111] The cause of these perfusion defects, however, is not classic coronary artery disease. It is more likely that they represent a supply/demand ischaemia in the context of severe RV hypertrophic response to systemic pressure loading.[112] Post-ejection RV longitudinal shortening (longitudinal excursion following the ejection phase), during stress echocardiography was shown in a significant number of patients,[113] suggesting incoordinated myocardial contraction, highly sensitive to myocardial ischemia.[113] Regions of abnormal RV myocardium can be visualised late after atrial switch with the use of CMR with late gadolinium enhancement and are likely to represent focal fibrosis[39] (fig 19). The presence and extent of such regions correlate with RV mass, RV dilatation, and impaired systolic function, suggesting that hypertrophy is associated with fibrosis in some patients, and correlates inversely with RV systolic performance.[39] Furthermore, gadolinium enhancement was associated with markers of adverse outcome like QRS duration and arrhythmia itself, underlining the prognostic significance of these findings.[39]

Although accurate assessment of RV ejection fraction is important, the definition of ''normal'' systemic RV ejection fraction remains problematic and depends on the method of determination. However, most authorities agree that a systemic RV ejection fraction $> 50\%$ can be considered normal (in the absence of significant valve regurgitation).[114 115] For the reasons discussed, CMR is considered the gold standard for the study of RV size and function in these patients.[115] Radionuclide angiography is a useful alternative for serial follow up when CMR or CT are not available.[114 116] Although transthoracic echo assessment of adult patients is limited in quantitative volumetric data, it provides invaluable information on baffle patency, leaks (with contrast studies), valvar regurgitation or stenosis and, in experienced hands, semiquantitative information on RV function.[117] MRI derived RV volumes correlate positively with echo derived RV inlet dimensions and negatively with the dP/dT of the tricuspid regurgitant jet (indirect measure of RV contractile function).[117] Furthermore, RV longitudinal function (M mode: wall excursion measured from the apex) correlates with CMR derived RV ejection fraction.[117]

Echocardiographic indices of systemic RV function during dobutamine stress (such as RV long axis excursion) predict exercise capacity, establishing stress echocardiography as an important semi-invasive, physiologic imaging modality.[113] Sinus node dysfunction is a relatively frequent finding in these patients, necessitating permanent pacing.[118] Pacing, in turn, constitutes a contraindication for CMR at present. Other emerging imaging modalities, namely MSCT, may prove to be useful alternatives for volumetric analysis of the RV.[119]

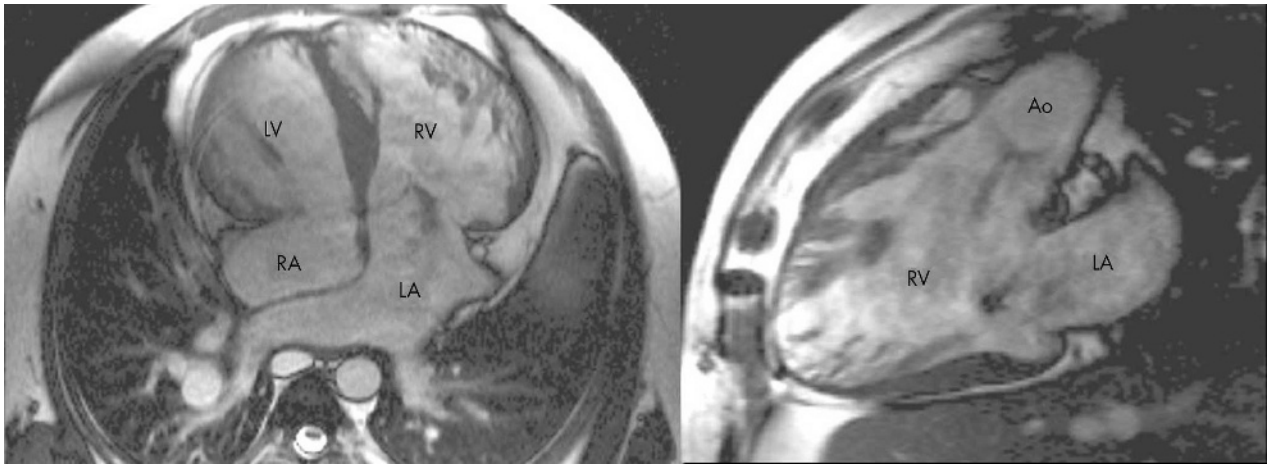

Figure 20 Gradient echo (white blood) CMR sequences of a patient with corrected transposition of the great arteries (double discordance). Left panel: Transaxial four chamber view. The pulmonary veins (left and right lower veins in this plane) are draining to the left atrium, which is however connected to the RV (systemic RV). The right atrium, in turn, is connected to the left ventricle (subpulmonary LV). Right panel: Same patient shown on the left. The relationships of the dilated and hypertrophied RV with the aorta and the left atrium are depicted. There is a signal void of tricuspid valve (systemic atrioventricular valve) regurgitation and a signal void of aortic regurgitation. Ao, aorta; LA, left atrium; LV, left ventricle, RA, right atrium; RV, right ventricle.

Congenitally corrected transposition of the great arteries (ccTGA)

The RV supports the systemic circulation in patients with ccTGA. Associated lesions (ventricular septal defect, pulmonary stenosis, and Ebstein's anomaly of the systemic tricuspid valve) are common.[120] Diagnosis may be made in adult life in non-cyanotic patients, usually by identifying a systemic ventricle with RV morphologic characteristics[121] (coarse trabeculations, moderator band, and the insertion of the septal leaflet of a morphologically tricuspid valve to the ventricular septum in conjunction with a morphologically mitral valve on the right side).[121] The relation of the two ventricles (and the two great arteries) is more side by side than the usual anteroposterior, rendering the apical and subxiphoid four chamber the most useful echocardiographic views for diagnosis. Transoesophageal echocardiography may be required and, as with complete TGA, additional imaging such as CMR is preferable for assessing systemic RV function[122] (fig 20).

Long term outcome is not normal even in patients without associated lesions due to a propensity to complete heart block,[123] tricuspid valve regurgitation (TR),[124] and the development of RV systolic dysfunction.[125–130] Not surprisingly, more than moderate TR and RV dysfunction are significantly related to increased mortality,[129] with TR being the most significant independent predictor of outcome.[130] However, TR strongly relates to RV dysfunction, raising the question whether TR leads to RV dysfunction or vice versa.[128 130] Despite difficulties in assessing the systemic RV in ccTGA, usually a semiquantitive evaluation of ventricular function and TR by echo is feasible. In contrast to patients with atrioventricular concordance, who often tolerate a significant degree of mitral insufficiency for decades before left ventricular failure ensues, RV dysfunction usually starts within five years from onset of TR in ccTGA patients (without associated lesions or surgery).[130–132] RV failure with ventricular enlargement results in worsening of TR due to annular dilatation. The factors responsible for accelerated failure of the systemic RV are not quite clear. It seems that ventricular geometry and the design of the respective atrioventricular valve is important.[133] Also perfusion defects at rest have been reported in patients with ccTGA without associated lesions.[134 135] Coronary flow reserve assessed with positron emission tomography is decreased, indicating altered vasoreactivity and quantitative changes in microcirculation.[136]

The adverse interplay between TR and RV dysfunction in these patients calls for timely tricuspid valve replacement (repair does not work), otherwise patients should be considered for transplantation.[130 137] In this regard, serial assessment of TR and RV function is mandatory, underscoring the benefits of a combined imaging approach with echo and CMR.[4 20 138 139] Gated equilibrium radionuclide angiocardiography may also be used for assessment of RV function at rest and during exercise.[126] Coronary artery origin and distribution are reversed and frequently anomalous in these patients and non-invasive coronary angiography (with CT or CMR) can delineate it.[2 3 140]

CONCLUSION

The RV, with its complex geometry and unique adaptive mechanisms in CHD, remains a challenge to the cardiologist. The RV is a pivotal chamber and its dysfunction—both systolic and diastolic—has clear implications to short and long term outcome. Recent advances in imaging, particularly in CMR, have revolutionised the exploration of RV anatomy and function and have shed light on late pathophysiology of many CHD defects. Transthoracic echocardiography remains the workhorse of non-invasive assessment of the RV in patients with CHD, however. Combined with other imaging, appropriately selected and timed for the individual patient with CHD, echocardiography remains key to assessing disease progression and timing of late re-intervention.

ACKNOWLEDGEMENTS

We thank Drs Wei Li, Philip Kilner, and Michael Mullen for providing us with figures. MAG and the Royal Brompton Adult Congenital Heart Programme have received support from the Clinical Research Committee and the Waring Trust both from the Royal Brompton & Harefield NHS Trust and from the British Heart Foundation.

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