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REVIEW ARTICLE

CT and MR imaging of patients with a dilated right ventricle due to congenital causes and their treatment

[1,2]RODRIGO SALGADO, MD, PhD, [3]RICARDO P.J. BUDDE, MD, PhD and [4]LUCA SABA, MD, PhD

[1]Department of Radiology, Antwerp University Hospital, Antwerp (Edegem), Belgium
[2]Department of Radiology, Holy Heart Hospital, Lier, Belgium
[3]Department of Radiology & Nuclear Medicine, Erasmus MC, Rotterdam, The Netherlands
[4]Department of Radiology, University of Cagliari, Cagliari, Italy

Address correspondence to: Ricardo P.J. Budde
E-mail: *r.budde@erasmusmc.nl*

ABSTRACT

A variety of both acquired and congenital conditions can significantly affect the right ventricle, with a variety of potential origins that can have substantial clinical ramifications. These conditions can range from the impact of diseases like pulmonary arterial hypertension and ischaemic heart disease to valvular deficiencies resulting in heart failure. Moreover, the right ventricle response to factors like abnormal loading conditions, and its subsequent clinical effects, are influenced by factors such as age, disease progression, potential interventions, and their immediate and long-term clinical outcomes. Therefore, a readily available and reproducible non-invasive imaging assessment can aid in diagnosing the underlying condition of a dilated right ventricle, track its evolution, and help devising the most appropriate treatment strategy and optimal timing for its implementation throughout the patient's life.
In this review, our primary focus will be on the non-invasive imaging with CT and MR of an enlarged right ventricle resulting from congenital causes and their treatment.

INTRODUCTION

For a long time, the right ventricle (RV) was considered a "forgotten" chamber, its contribution to cardiac function overshadowed by its left counterpart, and often thought of inferior clinical relevance. However, many acquired and congenital conditions can affect the RV spanning a wide range of possible aetiologies with significant clinical repercussions, ranging from the effects of pulmonary arterial hypertension and ischaemic heart disease to valvular deficiencies leading to heart failure.[1] Also, RV adaptation to, *e.g.* abnormal loading conditions and hence its clinical effects can vary according to natural ageing, disease progression, possible interventions and their short- and long-term clinical consequences. As such, an accessible and reproducible non-invasive imaging assessment will help in the establishing the disease, monitor its progression, and help devising the best possible treatment plan and timing of its execution during the patient's lifetime.

In this review, we will focus on the non-invasive imaging of a dilated RV arising from a congenital origin or its treatment using CT and MR, highlighting relevant information and the necessary topics which must be included in the final report.

AETIOLOGY OF A DILATED RV—VOLUME- *VS* PRESSURE-OVERLOAD

Causes of RV dilatation can be divided into three groups, secondary to disease causing (1) RV volume overload, (2) pressure overload or the consequence of (3) intrinsic or acquired myocardial disease. Of these possible aetiologies, the first two groups will cover most encountered cases in clinical practice.

As such, when initially approaching a patient with a dilated RV, an initial distinction between volume- *vs* pressure-overload is very useful, as this allows narrowing an initial broad differential diagnosis to a more specific targeted approach.

The initial response of the RV to increased pressure overload is myocardial hypertrophy, as can be seen in untreated tetralogy of Fallot or patients with chronic obstructive pulmonary disease. Conversely, a volume-overloaded RV will dilate initially without wall hypertrophy and occasionally conversely presenting with a rather thin ventricle wall, as can be witnessed in patients with pulmonary regurgitation and dilated cardiomyopathy. As the RV will more

easily adapt to volume- than to pressure-overload, RV systolic function can remain unaffected over longer periods of time, explaining the sometimes discrepant relation between clinical symptoms and disease severity. This is illustrated by the fact that many patients with repaired Tetralogy of Fallot generally tolerate varying degrees of RV dilatation well until adulthood.

Altered trans-septal loading conditions will also influence septal motion. In patients with RV volume-overload, the interventricular septum (IVS) will progressively flatten during the mid- to end-diastolic phase, even moving paradoxically towards the left ventricle, with loss of its normal curvature. During the systolic phase both motion and morphology of the IVS are subsequently restored.

The interatrial septum may also flatten during diastole to accommodate for the increased volume.

Conversely, in patients with RV pressure overload, the flattening of the IVS during diastole will become more pronounced in the systolic phase. A transient leftward motion of this IVS can also occur at the end of the systolic phase secondary to a preceding drop in LV pressure before the RV pressure decrease. Increased atrial pressure may also cause the interatrial septum to bow towards the left atrium, especially during atrial systole.

In patients with both RV-pressure and -volume overload, IVS motion can be complex, including both flattening and bowing of the IVS in different phases of the cardiac cycle.

Nevertheless, it is important to realise that, as disease progresses and cardiac adaption evolves to the limits of compensatory remodelling with progressive dysfunction, end-stage disease eventually develops where both RV hypertrophy and RV dilatation can co-exist.

Finally, knowledge of previous medical history (*e.g.* previous interventions for congenital heart disease or known chronic pulmonary hypertension) is also very helpful. Table 1 provides a broad overview that can serve as initial guidance.

IMAGING THE RIGHT VENTRICLE WITH CMR AND CT

Echocardiography remains the first-line imaging modality, delivering both morphological and functional information in a non-invasive manner.[2] However, it has known limitations due to both the nature of the imaging modality and patient-related factors. Its utility is further compromised by the complex anatomy and mechanics of the right ventricle, features that also require precautions in its evaluation with CT and MR.

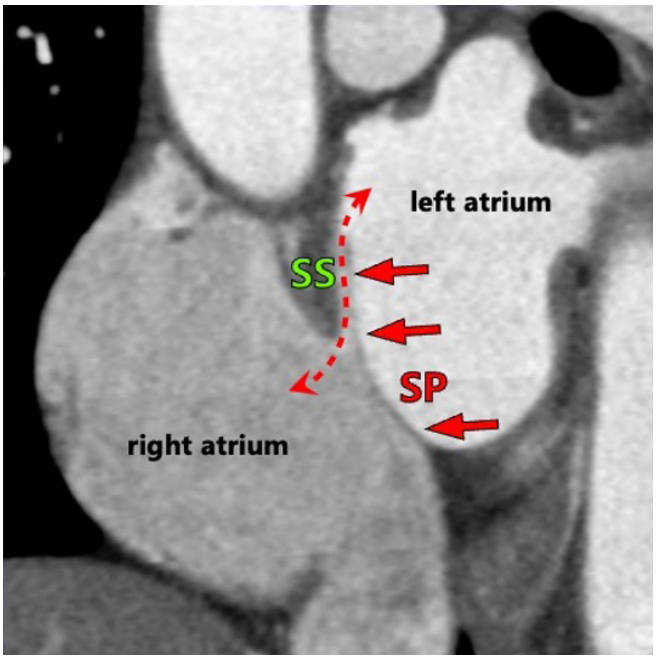
Despite the unmistakeable role of echocardiography, cardiac MR (CMR) has emerged as an essential tool in the further evaluation of the right ventricle, delivering a reproducible quantitative assessment of cardiac function, global and regional wall motion, myocardial tissue characterisation, as such overcoming many drawbacks of echocardiography.[3,4] It is therefore the *de-facto* reference standard for cardiac volumetry, mass, and systolic function, and is very well-suited for both the initial assessment as well the longitudinal follow-up of patients. Furthermore, the use of phase-contrast techniques, and more recently 4D flow techniques, allow the calculation of pulmonary and tricuspid transvalvular flow and also pulmonary to systemic (Qp:Qs) fractional shunt calculation. These parameters, especially the Qp:Qs ratio, provide important insights into the haemodynamic repercussions of disease and are important elements for consideration

Table 1. Different origins of a dilated RV

Driving mechanism	Aetiology group	Pathology
Volume-overload	Abnormal vascular connection	
	Atrial septal defect	ASD I
		ASD II
		Sinus venosus defect
	Other abnormal vascular connections	PAPVR (may co-exist with ASD)
		Extracardiac shunts
		Coronary fistulae to right heart
		Unroofed coronary sinus
	Regurgitant RV valve	Tricuspid valve dysfunction
		Pulmonary valve dysfunction
		Ebstein's anomaly
Pressure-overload	Increased afterload	Pulmonary hypertension
		RVOT obstruction
Cardiomyopathy	Congenitally absent or reduced RV myocardium	Uhl's anomaly
Other		ventricular aneurysms/diverticulum

ASD: atrial septal defect. PAPVR: partial anomalous pulmonary venous return;RV: right ventricle.

Figure 1. Illustration of the anatomy of a patent foramen ovale. In this cross-sectional CT image at the level of the fossa ovalis, the failed fusion between the SP (arrows) and SS can be appreciated. This leads to an interatrial communication oblique to the atrial septum (dashed line). This is a common finding during routine cardiac-CT examinations with no haemodynamic clinical consequences. SP, septum primum; SS, septum secundum.



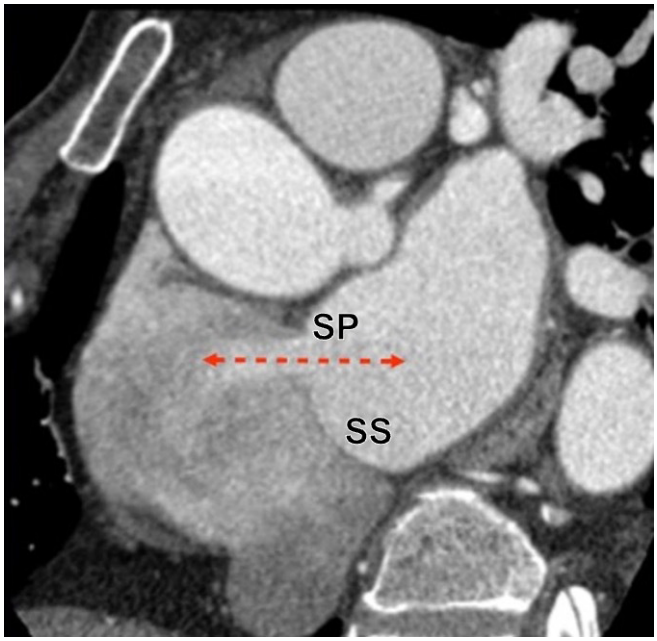
regarding indication and timing of additional interventional procedures.[3,5]

A further detailed overview of the various imaging techniques and normal RV anatomy can be found in many excellent reviews but is outside the scope of this paper.[3,6]

As CMR and echocardiography have some fundamental differences, measurements are not easily comparable. While authors have reported modality-specific normal values ranges for different cardiac measurements, a recent publication reported normal CMR ranges in adults and children based on data from several studies.[7–9] It is also important to have a standardised approach for both image acquisition and post-processing (*e.g.* epi- and endocardial contours for mass and volume quantification) in order to have reliable and reproducible longitudinal data over the lifetime of a patient.[10] Although some authors had initially reported differences in calculated RV volumes when using a RV axial- *vs* short-axis image stack approach, as well as interobserver variability, recent publications argue to that both methods are highly accurate compared with echocardiography-derived measurements.[10–13] As technology progresses and new artificial intelligence-driven analysis methods become more mainstream, it is safe to assume that correct quantification of RV volumes will only improve over time.[14,15]

Due to its different nature, CT is in practice reserved for a more detailed anatomical evaluation of the heart and great vessels, as

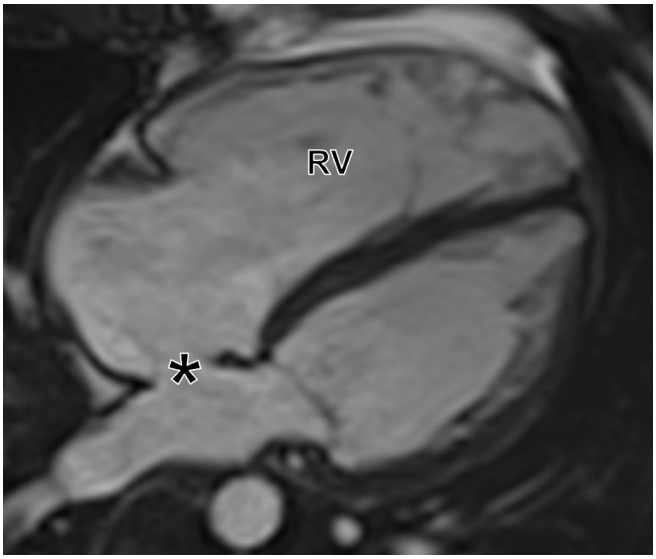
Figure 2. Ostium secundum ASD at the same level and orientation as Figure 1. In contrast to a PFO, an ASD is a permanent communication between left and right atrium which does not vary with the cardiac cycle. A further distinguishing feature with a PFO is the perpendicular transeptal flow of blood, as opposed to the more oblique orientation of flow through a PFO as illustrated in Figure 1. While ASD's are far less prevalent than PFO's, they have significantly more clinical impact as the left-right shunt will often lead to volume overload of the right heart with subsequent right ventricular dilatation. ASD, atrial septum defect; PFO, patent foramen ovale; SP, septum primum; SS, septum secundum.



it can cover the whole chest in a fraction of the time compared with CMR. It is as such an excellent imaging modality to detect and evaluate abnormal vascular connections like a partial anomalous pulmonary venous return (PAPVR) and has been shown to reliably evaluate intracardiac shunts.[16] While investigators have shown that quantification of right ventricular function and volumes is feasible, this remains a niche application reserved for cases in which other imaging modalities deliver equivocal results or CMR is not possible due to the presence of non-MR compatible devices *in-situ*.[17] Also, RV functional analysis requires retrospective gating to cover the whole range of the cardiac cycle, which is associated with an increased radiation exposure over prospective scans which usually register only portions of the cardiac cycle.

When considering congenital reasons for a dilated RV, pathology leading to an isolated volume overload of the RV can be divided between a RV valvular insufficiency (*e.g.* tricuspid or pulmonary insufficiency) and the presence of pre-tricuspid shunts like atrial septal defects (ASD) and PAPVRs. This latter cause constitutes a far more frequent reason for RV dilatation encountered in clinical practice, also outside specialised centres.

Figure 3. Ostium secundum ASD in a 57-year-old man. A dilated RV can be appreciated due to chronic volume overload through an ASD II defect (asterisk). Cardiac MR allows the quantification of the Qp:Qs shunt fraction, which is an important parameter when considering ASD closure. In this case, the Qp:Qs fraction was 2.5, making this an important factor in the final decision to proceed to deployment of a septal closure device in this particular case. ASD, atrial septum defect; RV, right ventricle.



ABNORMAL VASCULAR CONNECTIONS

In the spectrum of congenital conditions leading to a dilated RV, abnormal vascular connections have a prominent position. These include intracardiac shunts like ASD and PAPVR.

Atrial septal defects

Many cardiac shunts like a patent ductus arteriosus and ventricular septal defects are detected and treated during childhood. While ASD's are also commonly diagnosed during early life, they can also go undetected until adulthood as they form an anatomically heterogenous group with a wide range of clinical presentations depending on many factors.

In short, an ASD is a persistent interatrial communication after birth, allowing oxygenated blood to flow between the atria. The embryology of the atrial septum and the embryological events leading to these defects have been detailed elsewhere[16] Briefly summarising, during embryonic life the septum primum and secundum, the two components of the atrial septum, exist side-by-side with incomplete closure allowing passage of blood through the tunnel-like opening (fossa ovalis) between both atria. This is necessary for oxygenated blood from the umbilical vein to reach the foetal left atrium. As left atrial pressure increases after birth, fusion between the septum primum and secundum closes this defect in most of the population. In a varying percentage of the population, an incomplete atrial septal closure exists (Figure 1).

The most common defect is a patent foramen ovale (PFO), which occurs in up to 25–30% of the population and is mostly of no clinical haemodynamic significance.[18] However, it must be distinguished from an ASD which is rarer but has more clinical impact. ASD's account for up to 13% of all CHD, and up to 25–30% of CHD cases diagnosed in adulthood.[19] They form at predictable locations and allow as such targeted imaging.

The presence of an ASD will, in an initial stage, present as a left-to-right shunt, leading to a volume overload of the right heart with secondary dilatation of the right ventricle but without wall hypertrophy. While the volume overload through this interatrial shunt depends on the size of the defect, it is remarkable that even large ASD's can remain undetected until adult life, producing little or non-specific symptoms. As time progresses, progressive shunting leads to various haemodynamic-related symptoms including limited exercise intolerance, increased risk for atrial fibrillation (due to increased left atrial wall pressure), and the effects of eventually right-to-left shunting with decompensated heart failure and pulmonary hypertension.

While the initial diagnosis of a septal defect is usually made with echocardiography, both PFO and ASD can be diagnosed using CT or MR. Due to the different embryological origin, several imaging features allow a clear distinction in a vast majority of cases (Figure 2).[20,21] While a PFO is often incidentally noted during cardiac-CT examination performed for unrelated reasons, its detection can be increased when reviewing images across the full cardiac cycle.[22]

In practice, CT and CMR are performed for different indications in patients with an ASD. These imaging modalities are very rarely used for the diagnosis of a septal defect, as most patients will already have a diagnosis made on echocardiography.

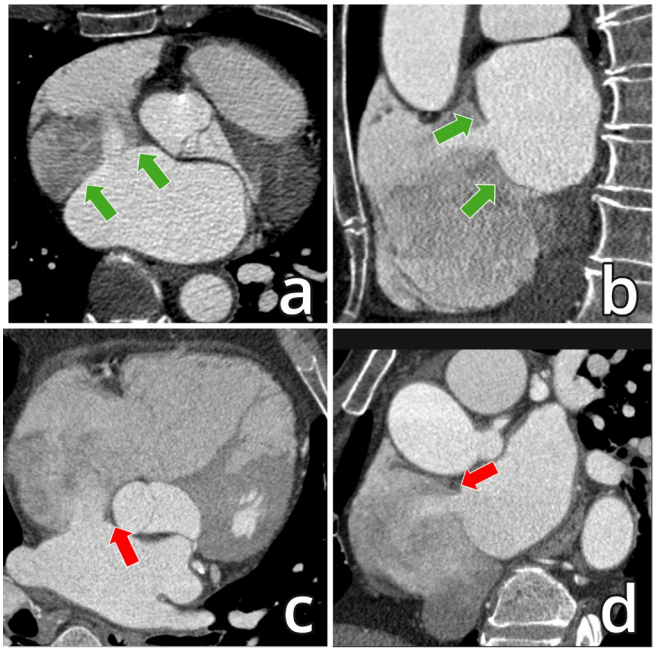
However, CMR will be the imaging modality of choice to evaluate the haemodynamic repercussions and calculate the Qp:Qs shunt fraction (Figure 3) . In the absence of an intracardiac shunt, the effective volume of blood reaching the systemic vascular bed (Qs) equals the effective volume reaching the pulmonary vasculature (Qp). However, the presence of a haemodynamically significant ASD will increase Qp and hence the shunt fraction Qp:Qs. A shunt fraction Qp:Qs > 1.5 is one of the most important parameters in selecting patients for a surgical or endovascular closure, as patients with a lower shunt fraction are less likely to benefit from an intervention.[2,23]

The role of CT lies mostly in the pre-procedural evaluation of the anatomical location of the ASD and its relationship with the atrial septum borders (Figure 4). Several devices are now available that allow closure of an ASD.[24] The excellent isotropic resolution of CT and its ability to obtain motion-free images through an ECG-gated acquisition makes its especially suited to help select patients and minimise procedural complications.[20] Knowledge of the specific intended devices is mandatory as the pre-requisites are device-specific.

Partial anomalous pulmonary venous return

A PAPVR is a vascular congenital condition in which at least one pulmonary vein drains in a systemic vein or the right atrium, leading to a left-to-right shunt and volume-overload of the right

Figure 4. Two patients with an ostium secundum ASD but different configuration of adjacent septal rims. In the first patient (a, b), there is sufficient length of the remaining atrial septum (arrows) adjacent to the ASD to attach a septal closure device. Conversely, there is almost no septal rim available (arrow) on the septum primum side of the ADS in the second patient (c, d), making this patient unsuited for endovascular treatment. Note the dilated right ventricle due to volume overload in the second patient. ASD, atrial septum defect.

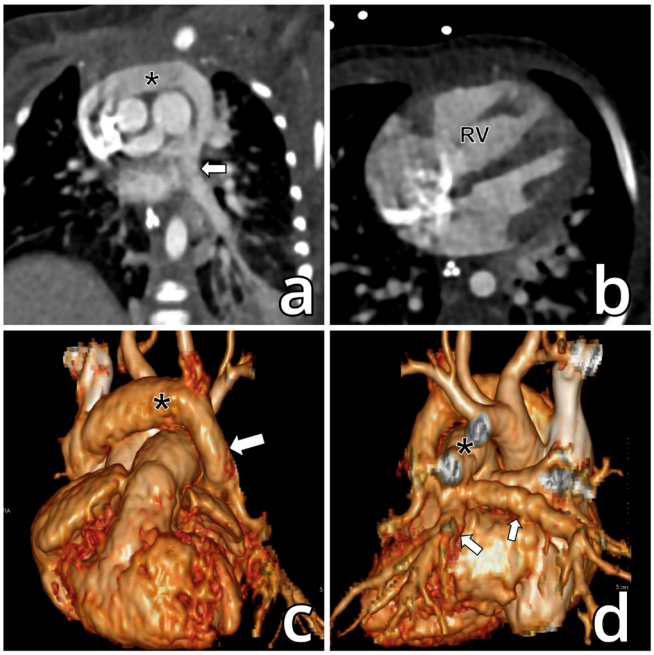


heart. A total anomalous pulmonary venous return (TAPVR) is an even rarer congenital disease with embryonical failure of connecting the common pulmonary vein to the left atrium, with all pulmonary veins connecting to the systemic veins, right atrium, or coronary sinus (Figure 5). A TAPVR has an incidence of about 7.1 per 1000,000 live births, while a PAPVR has been found in up to 0.7% of autopsies.[25,26]

Although a PAPVR may be an isolated finding, both conditions may appear in conjunction with an ASD, especially a sinus venosus defect in PAPVR and an ASD with right-to-left shunt in TAPVR. Both isolated PAPVR or in combination with an ASD can contribute to a dilated right ventricle (Figure 6).[27,28]

While a PAPVR can on occasion be appreciated on conventional chest radiographs, they are mostly identified on CT and MR imaging, sometimes on examinations performed for unrelated reasons (Figure 6). Both imaging modalities are equally suited for initial detection, CT having the additional advantage of providing an isotropic data set in a single short acquisition.[29–32] Echocardiography will often be the first-line imaging modality in the investigation of a patient with potentially cardiac-related symptoms, revealing the presence of a dilated right ventricle. It is however less-suited to always detect the abnormal connecting vein(s) directly, especially when located high in the chest. This, together with historical reluctance from surgeons to not routine extend dissection around the superior vena cava to avoid phrenic

Figure 5. Contrast-enhanced CT of a 14-day-old patient with a supracardial total anomalous abnormal venous return (arrows), draining to the left brachiocephalic vein (asterisk). RV, right ventricle.



nerve damage, may be one of the reasons why one may still encounter patients with a dilated RV secondary to a PAPVR that was initially missed during the workup of corrective congenital heart surgery during the neonatal period or early infancy.[33]

While both CT and MR allows excellent anatomical visualisation and classification of this vascular abnormality, CMR is the modality of choice to evaluate the haemodynamic repercussions by calculation of the Qp:Qs shunt fraction. This is important, as in the absence of a significant shunt or RV dilatation, PAVR can be treated conservatively. In practice, this will mostly apply when there is only a single, mostly left-sided abnormal draining vein (Figure 7). However, this is not an absolute guideline as recent publications point to the likely presence of a significant left-to-right shunt (Qp:Qs > 1.5) when PAPVR and ASD co-exist, and in patients with a single right-sided PAPVR (Figure 8).[34,35] In these cases, surgical correction can be indicated, even in patients with a single PAPVR.[23,34]

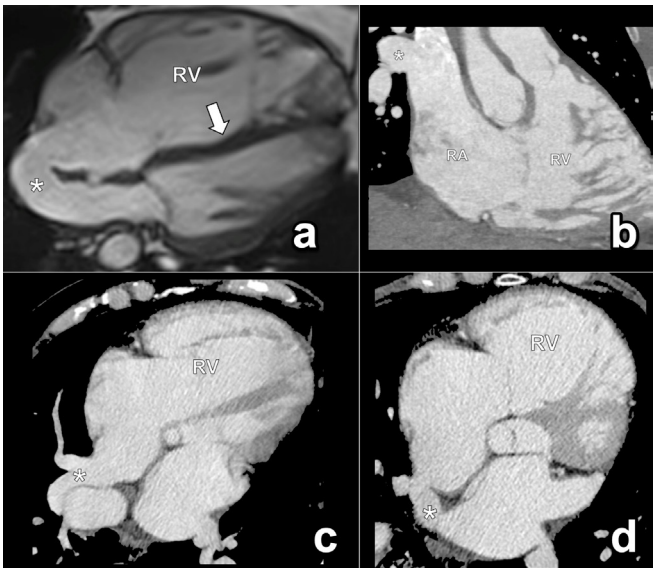
VALVULAR AETIOLOGY

Both pulmonary and tricuspid valve deficiencies can lead to RV dilatation in congenital heart disease.

Pulmonary valve dysfunction

While pulmonary regurgitation (PR) is usually an acquired condition, it plays an important role in RV dilatation in the setting of repaired tetralogy of Fallot. Advances in the conservative and surgical treatment of congenital heart disorders have achieved that specifically for this condition, large proportions of patients now survive into adulthood. According to recent data, about 90% of patients with surgically repaired tetralogy

Figure 6. 52-year-old patient with a superior sinus venosus defect in combination with a partial anomalous venous return of the right upper pulmonary vein. The cardiac MR 4-chamber view clearly illustrated the sinus venosus defect between RA and left atrium (asterisk), with a dilated right ventricle (RV). Note also the inward septal deviation (arrow). A contrast-enhanced CT-examination further illustrates the abnormal venous return (asterisk in panel b, c), together with the abnormal interatrial communication (asterisk in d). RA, right atrium; RV, right ventricle.



of Fallot (rTOF) survive at least the first two decades.[36] Despite these advances, survival eventually drops in the third decade due to long-term complications, of which PR is one of the most important driving factors. The underlying mechanism for this can be traced back to the original intervention, as surgical right

Figure 7. 37-year-old woman with an incidentally detected partial anomalous venous return (arrow) of the left pulmonary veins to the left brachiocephalic vein during a MR examination performed for unrelated reasons. There was no dilatation of the right ventricle, and no treatment was deemed necessary.

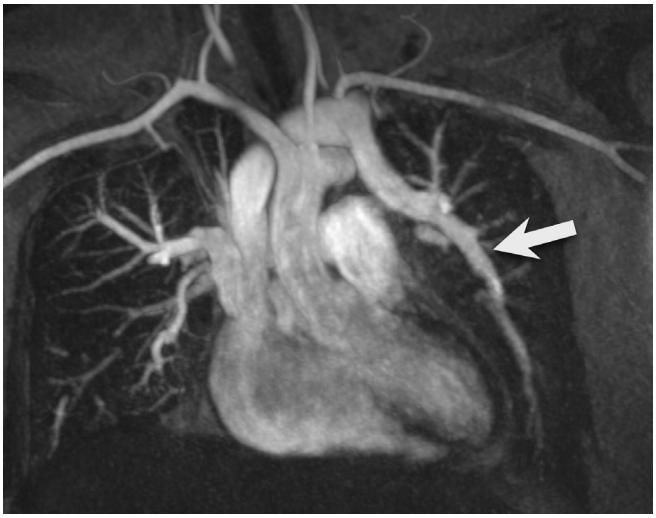
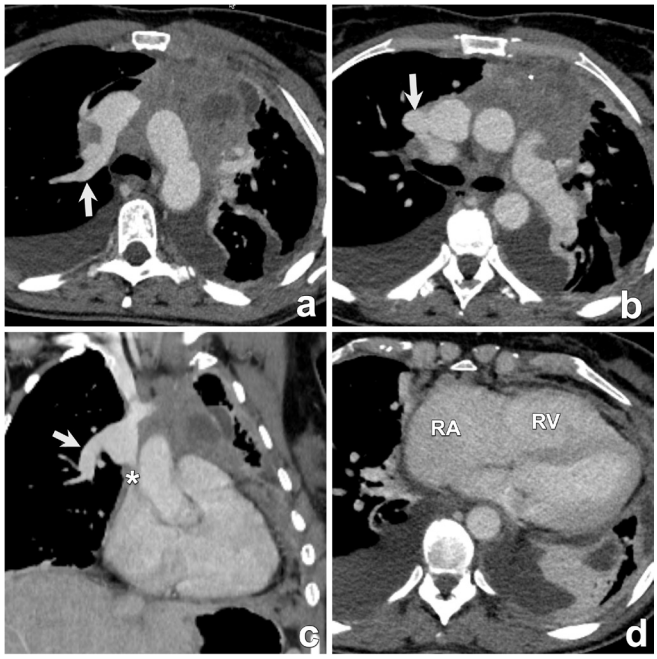


Figure 8. 52-year-old woman with thymic carcinoma and anomalous venous return from the right upper pulmonary veins (arrows in a–c) in the superior vena cava. Dilatation of the RA and RV secondary to the chronic volume overload (d). Note also the focal stricture of the superior vena cava secondary to tumoral infiltration (asteriks in c). RA, right atrium; RV, right ventricle.



ventricular outflow tract resection with often transannular patch repair usually involves severe disruption of the native pulmonary valve. Therefore, a compromised pulmonary valve function leads to PR with over time progressive RV volume-overload. Concomitant tricuspid regurgitation can also occur.

While symptoms can be initially limited and often well-tolerated during childhood, this progressive and long-standing volume-overload will eventually deteriorate RV function and progress to heart failure. Also, severe RV dilatation and chronic PR are also recognised haemodynamic substrates for the development of arrhythmias and sudden cardiac death.

CMR is the modality of choice of longitudinal follow-up of rTOF patients, delivering quantitative functional and morphological assessment of both left and right ventricles, together with non-invasive determination of the pulmonary regurgitant fraction. This long-term follow-up is important to monitor serial changes in RV size and function.

As pulmonary valve function progressively deteriorates and compromises RV morphology and function, definitive pulmonary valve replacement (PVR) will eventually become necessary. As this is a major intervention, guidelines support the use of CMR to determine the optimal time point for PVR, currently based on CMR-derived indexed RV-volumes (Figure 9**,** including supplementary movie files).[2,37] Close monitoring is important, as PVR must be performed before irreversible RV dysfunction occurs to

Figure 9. Repaired tetratology of Fallot with severe pulmonary insufficiency. Marked turbulent regurgitant flow can be appreciated in the right ventricle (asteriks in a), with severe turbulant flow at the level of the pulmonary valve (asteriks in c). Note also the dilated right ventricle with septal flattening (arrow in b). Regurgitant fraction can be calculated with phase-contrast sequences,indicating in a different patient a regurgitation fraction of 62%, which is a further important argument for pulmonary valve replacement. See also supplementary movie files for a–c.

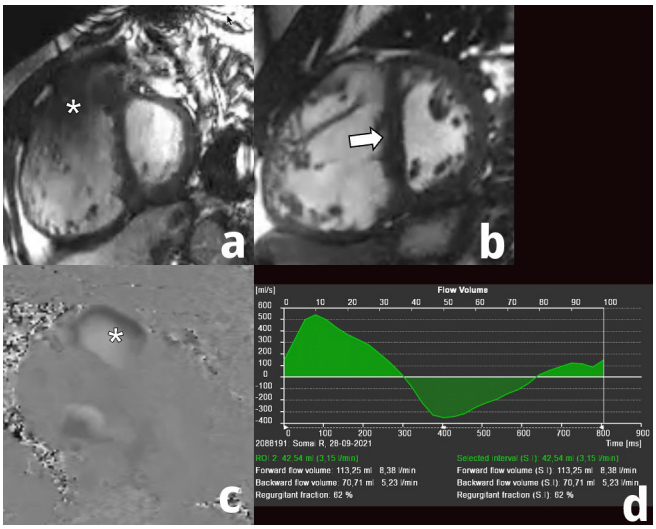
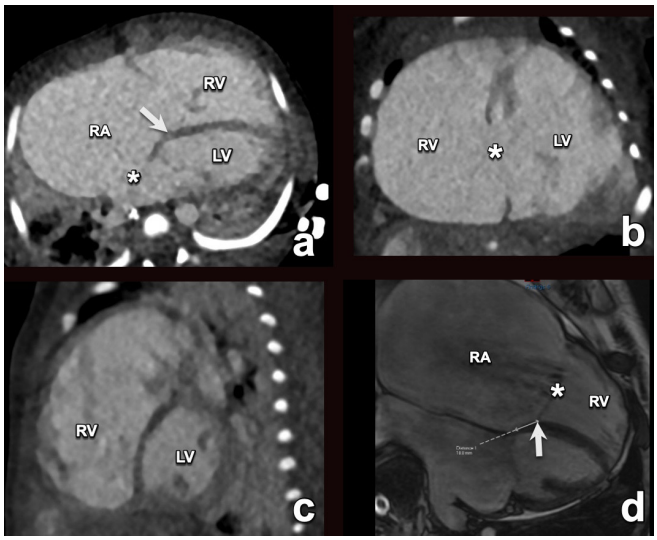


Figure 10. A 10-day-old child with Ebstein anomaly. The abnormal apical displacement of the septal tricuspid valve leaflet is clearly illustrated (arrows in a, d) on both the CT images (a–c) and 4-chamber CMR image (d). The secondary larger appearance of the LA is evident, as are the signs of volume overload with a dilated RV compared with the left counterpart. Note also a large ASD (asteriks in a, b). CMR (d) can additionally deliver quantification of ventricular volumes and regurgitation fraction. ASD, atrial septum defect; CMR, cardiac MR; LA, right atrium; LV, left ventricle; RA, right atrium, RV, right ventricle.



preserve long-term RV function. As such, many centres will start considering PVR when RV end-diastolic volumes are between 150 and 170 ml/m^2. A recent meta-analysis showed that larger indexed RV ESV and EDV are associated with a lower change of RV volume normalisations after PVR.[38] Nevertheless, while PVR can provide clinical relief with reduced or normalised RV volume, a clear survival benefit remains unclear.[39]

CMR can also adequately quantify PVR, but in practice, this will often add little to the already established echocardiographic assessment of PV function in most patients, its use reserved for specific cases.

When assessing potential PVR candidates, longitudinal data are more important than single measurements.[40] It is therefore important that all quantitative data are obtained using a standardised approach, both in the acquisition of different sequences and the tracing of endocardial RV contours for quantification.

Tricuspid valve dysfunction

Ebstein anomaly is a congenital valvular condition characterised by an apical displacement of the septal and posterior leaflets of the tricuspid valve. The ensuing valvular dysfunction leads to severe tricuspid valve regurgitation and volume overload of the right heart. Concomitant right-to-left shunting through a patent foramen ovale can lead to cyanosis, although there is a wide spectrum of ages and clinical symptoms at first presentation, some being even asymptomatic. Besides cyanosis, volume overload may lead to left ventricular diastolic dysfunction secondary to massive right-sided cardiac enlargement. Finally, patients may experience arrhythmias due to conduction abnormalities, which may lead to sudden death.

Both CT and CMR can be used to visualise the abnormal position of the tricuspid valve, with an enlarged right atrium and ventricle due to volume overload.[41,42] CMR can additionally provide quantification of ventricular volume, right- and left ventricular function and regurgitation fraction (Figure 10).

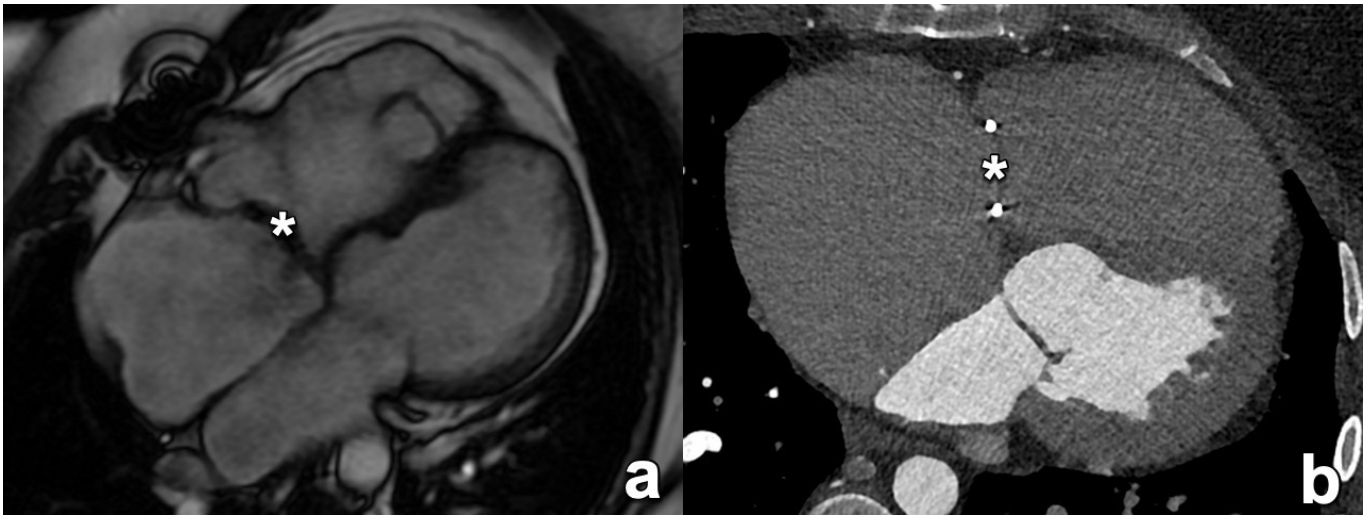
While treatment can initially be supportive in cyanotic neonates, definite repair is currently achieved by valvuloplasty or tricuspid valve repair (Figure 11).

Isolated tricuspid regurgitation is a morphological subtype that is being increasingly recognised as a distinct entity. In the absence of, *e.g.* concomitant pulmonary hypertension or co-existing left-sided heart disease, isolated tricuspid regurgitation will mostly be found present in elderly patients with a high prevalence of atrial fibrillation causing right atrial dilation, tricuspid valve annulus dilation, and tricuspid valve leaflets malcoaptation.

MYOCARDIAL CAUSES

Congenital myocardial causes of right ventricular dilatation are rare. In Uhl's anomaly, there is almost complete absence of right ventricular myocardium, with however a normal tricuspid valve.[43] The septal myocardium is preserved, as is the left ventricular myocardium. Both right atrial and ventricular dilatation can be seen, and it also often associated with other congenital disorders including Ebstein anomaly.

Figure 11. CMR (a) and CT (b) images after tricuspid valve repair, showing the bioprosthetic tricuspid valve prosthesis (asteriks) in normal position. Note the persisting dilatation of the right heart. CMR, cardiac MR.



Finally, arrhythmogenic right ventricular cardiomyopathy should always be considered in a young patient with a dilated and impaired RV. Task force criteria exist encompassing a combination of clinical, pathologic, electrophysiological, and imaging information. An in-depth discussion is however beyond the scope of this paper.

CONCLUSION

Congenital causes of right ventricular dilatation are more common than usually perceived, as abnormal vascular connections like PAVR and intracardiac shunts are often incidentally seen during CT- and MR-examinations performed for other reasons. Also, CMR has evolved into a crucial imaging tool to follow disease progression and determine optimal time point for (re)intervention, due to its capabilities to quantify both ventricular volumes and function as well as the regurgitation fraction in valvular causes of RV dilatation.

A good knowledge of the different aetiologies as explained in this paper will help the radiologist in planning and executing the examination, as well as integrating all necessary information in the final report.

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