Journal of the American College of Cardiology
© 2009 by the American College of Cardiology Foundation
Published by Elsevier Inc.

Vol. 54, No. 21, 2009
ISSN 0735-1097/09/$36.00
doi:10.1016/j.jacc.2009.06.048

STATE-OF-THE-ART PAPERS

Adult Congenital Heart Disease

Importance of the Right Ventricle

Carole A. Warnes, MD

Rochester, Minnesota

The right ventricle (RV) is of lesser importance in acquired heart disease, but its role is of increasing importance in congenital heart disease. It may function as a subpulmonary ventricle or as a subaortic (systemic) ventricle in transposition complexes. The RV has a remarkable ability to adapt to pressure and volume load, but its size and function are often overlooked. Patients usually develop symptoms only after RV dysfunction has occurred, and in many diverse clinical scenarios, late referral is common. Advanced RV enlargement and dysfunction are responsible for not only impaired functional capacity but also lethal ventricular arrhythmias and sudden death. Appropriate imaging of the size and function of the RV are important because timely surgery on the pulmonary and tricuspid valves may preserve RV size and function. Adults with congenital heart disease should be followed at centers where there is an understanding of these problems so that valvular surgery can be considered when appropriate. (J Am Coll Cardiol 2009;54:1903–10) © 2009 by the American College of Cardiology Foundation

In the last 2 decades, many investigations surrounding acquired cardiac disease have focused on the left ventricle (LV) and the importance of preserving its systolic function. Examples include coronary revascularization and appropriate timing for mitral and aortic valve disease to avoid LV dysfunction, heart failure, and risk of sudden cardiac death, which ensues if surgery is performed too late. In stark contrast, the right ventricle (RV) is largely overlooked, and surgery for tricuspid regurgitation (TR), for example, is usually reserved for patients who are refractory to medical therapy, even if the RV dysfunction is severe. The surgical mortality in these circumstances may approach 30% (1). Whether earlier surgery would improve the surgical mortality or whether residual RV dysfunction portends a poor prognosis is unclear. As such, the RV is of secondary importance in acquired heart disease, and there is little or no impetus to preserve its function.

In adult congenital heart disease, the RV has also been "forgotten," but in the last decade, increasing attention has been focused on the RV in an effort to avoid the adverse outcomes that may be associated with RV dysfunction and fibrosis. These outcomes include not only exercise limitation and RV failure, but importantly, lethal ventricular arrhythmias. In a variety of clinical scenarios, pulmonary and tricuspid valve surgery is being performed earlier in an attempt to "save" the RV, and the surgical mortality in experienced centers is acceptably low. Although the timing of these surgeries remains controversial, it is clear that earlier interventions can have excellent long-term outcomes.

It is recognized that the RV has a completely different shape, structure, and contraction pattern than the LV, and measurement of its size and function to facilitate timing of surgery is more challenging. Most often it exists as a subpulmonary ventricle but may be the subaortic (systemic) ventricle in transposition complexes. This article reviews some of the current issues related to the RV and the important role it plays in congenital heart disease.

Anatomy

The RV is composed of 3 anatomic and functional subunits, which include the inlet portion extending from the tricuspid valve to the insertions of the papillary muscles onto the ventricular wall, the trabecular portion involving the RV body and apex (the fundamental component of the pump mechanism), and the outflow or infundibular portion extending to the pulmonary valve that is generally free of trabeculations (Fig. 1A). This complex, more triangular shape of the RV contrasts with the more conical shape of the LV; when viewed in short axis, the RV has a crescent shape and "wraps around" the LV (Fig. 1B). The muscular wall of the normal RV is usually 3 to 5 mm in thickness, but in conditions of pressure overload, the RV wall thickness may even exceed that of the LV. The myocardial fiber arrangement of the RV is different than that of the LV, and its contraction relies more heavily on longitudinal shortening than the circumferential fiber arrangement of the LV (2,3). As a subpulmonary ventricle pumping to a low-resistance circuit, the RV functions at a lower ejection fraction (EF) than its counterpart. The coronary arteries are

From the Divisions of Cardiovascular Diseases and Pediatric Cardiology and Department of Internal Medicine, Mayo Clinic, Rochester, Minnesota.
Manuscript received April 6, 2009; accepted June 3, 2009.

JACC Vol. 54, No. 21, 2009
November 17, 2009:1903–10

Abbreviations and Acronyms

AV = atrioventricular
EF = ejection fraction
LV = left ventricle/ventricular
PS = pulmonary stenosis
PVR = pulmonary valve replacement
RV = right ventricle/ventricular
RVOT = right ventricular outflow tract
TR = tricuspid regurgitation
VSD = ventricular septal defect
VT = ventricular tachycardia

concordant; therefore, regardless of the anatomic arrangement, a single right coronary artery supplies the RV myocardium, with the inherent vulnerability to oxygen supply/demand mismatch. As such, a morphologic RV seems inherently incapable of functioning as a subaortic systemic ventricular pump, although as this review will discuss, it has the remarkable capability to adapt and can function at systemic pressures for decades.

Imaging

The chest radiograph may be misleading in the assessment of RV size because the RV may be significantly enlarged and may compress the LV, resulting in a normal cardiothoracic ratio. Two-dimensional echocardiography facilitates assessment of LV size and function, and reliable mathematical models can be applied because it has a prolate ellipsoid shape. In contrast, precise measurements of the RV are challenging because of its complex shape (4). It must be imaged in multiple planes, although the short-axis view is usually the most helpful. A qualitative visual assessment is usually applied, with the RV size being described as either normal or mildly, moderately, or severely enlarged. If the RV is the same size as the LV, it is usually characterized as moderately enlarged, and if larger than the LV, it is severely enlarged. Function of the RV is usually characterized as normal or mildly, moderately, or severely dysfunctional. With serial follow-up, there may be little interobserver variability; however, among different interpreters and institutions, there may be considerable differences in the assessment of RV size and function. Because of these limitations, 3-dimensional echocardiography is being increasingly utilized although not yet well validated.

Magnetic resonance imaging is also increasingly being used to assess the RV; although it is often referred to as the "gold standard," there are difficulties with its use—including considerable interobserver/intraobserver variability (5). Endocardial border definition is essential to aid RV volume calculation, and both trabeculations and papillary muscles in the RV make this more difficult (6,7). Although many centers compute RV volumes from short-axis images, there is not universal consensus (5). The most basal RV slice near the tricuspid valve is particularly difficult to define; because this is a large area, the potential for error is significant. The RV outflow may be excluded from the volume measurements if there is an aneurysm of the right ventricular outflow tract (RVOT), which may occur after patching. Increasingly, as RV end-diastolic volumes are being used to facilitate decisions about timing of valve surgery in congenital heart disease, these caveats must be remembered. A consistent and reproducible approach should be adopted by all.

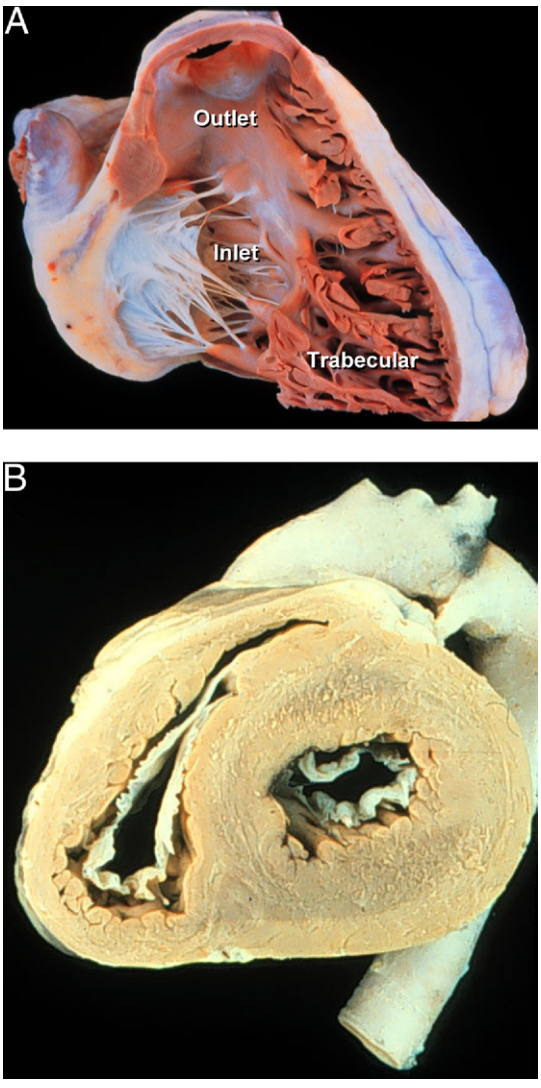

Figure 1 Pathologic Specimens

(A) Pathologic specimen of the right ventricle with the free wall removed to demonstrate the 3 anatomic regions. **(B)** Pathologic specimen of the heart cut transversely demonstrating the crescent shape of the right ventricle. Photos courtesy of Dr. W. D. Edwards, consultant in pathology, Mayo Clinic.

Volume-based indexes of RV function have limitations because they are load dependent, and other Doppler measurements may add insights into RV function. These include the dP/dt of the TR velocity and the index of myocardial performance of the RV (8). Other measurements include tissue Doppler imaging of the tricuspid annulus (9) and myocardial acceleration during isovolumic contraction (10), which measure intrinsic contractility but are not used routinely. RV diastolic function may also be assessed by Doppler interrogation of hepatic and superior vena cava inflow patterns as well as the RVOT. The

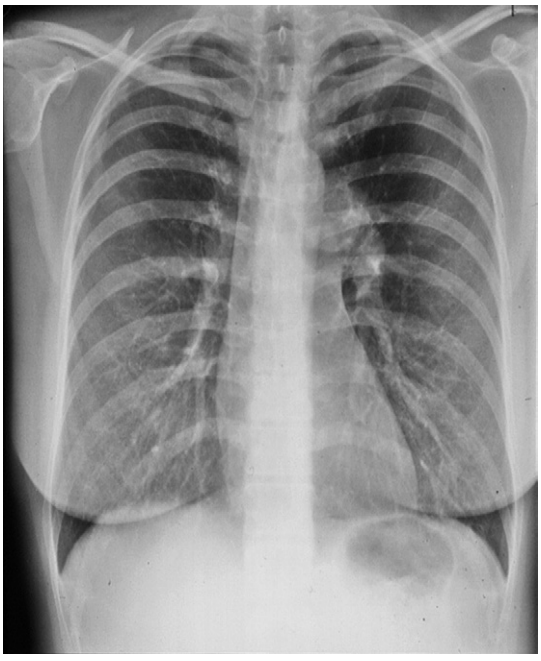

Figure 2 **Chest Radiograph of a Patient With Severe Valvular Pulmonary Stenosis**

The heart size is normal even though the right ventricular pressure in this patient is 150 mm Hg. Note the characteristic post-stenotic dilation of the pulmonary artery.

demonstration of atrial contribution to forward flow with retrograde flow in the superior vena cava suggests restrictive physiology in the RV (11).

Pulmonary Valve Stenosis (PS)

PS exemplifies the ability of the RV to adapt to pressure overload and is the most common form of RV outflow obstruction. Obstruction may also occur at the supravalvular level; below the valve, obstruction is commonly at the infundibular level and is usually secondary to PS. The chest radiograph often has characteristic features, with a normal heart size and post-stenotic dilation of the pulmonary artery (Fig. 2). The diagnosis and severity of PS is usually made by clinical examination and confirmed by transthoracic echocardiography as long as multiple windows are explored and the RV pressure confirmed by the TR velocity.

The RV responds to PS by hypertrophy and maintains its function for years even when the pressure is systemic. It is a misconception that the RV dilates and fails when exposed to high pressure; indeed, as long as sinus rhythm is preserved, and there is no additional volume lesion, the RV can maintain good function until the patient is in the fifth decade. When the RV pressure exceeds 50% of the systemic pressure, however, many patients develop symptoms, typically dyspnea, dizziness, or chest pain.

The first operation for PS was performed in 1948, and subsequent procedures in the early 1950s involved a closed valvotomy. Later, open pulmonary valvotomy yielded superior results. If the pulmonary annulus is small, a transannular patch may be needed; if the valve is dysplastic, a valvectomy may be necessary. These operations always result in pulmonary regurgitation, which is tolerated well for years. Initially the RV compensates by dilation but maintains contractility and stroke volume. Often decades later, however, RV systolic function deteriorates, and patients develop symptoms, usually dyspnea, fatigue, and atrial arrhythmias.

PS is generally considered a benign lesion, and operations for PS were considered "curative." In the Second Natural History Study, only 14 of 277 (5%) patients treated surgically for isolated PS required reintervention at 25-year follow-up (12). In the longer term, however, particularly after a closed procedure when significant pulmonary regurgitation is a common sequela, the outcome is not so benign, and further surgery may be necessary. In the series by Earing et al. (13), 21 of 53 (40%) adult patients underwent pulmonary valve replacement (PVR), with a mean interval after initial surgery to the time of PVR of 33 years. Most patients had moderate or severe RV dilation at the time of PVR, two-thirds were New York Heart Association functional class II or III, and one-half had at least moderate TR requiring concomitant tricuspid valve annuloplasty. Two patients in the follow-up group who had not undergone PVR experienced sudden cardiac death, and another 3 developed a ventricular arrhythmia. These data are similar to those in a study of congenital pulmonary valve regurgitation in which 21 of 72 (29%) patients developed symptoms after 40 years and the rate of development (hazard function) of symptoms was markedly increased after 40 years (14).

Thus, in unoperated PS, the RV remains well compensated; however, after valvotomy, the resultant pulmonary regurgitation may cause RV dysfunction. Pulmonary valve replacement should be performed before severe RV dysfunction occurs; otherwise, RV function never recovers. This emphasizes the importance of lifelong follow-up for these patients, many of whom consider themselves "cured." Serial exercise testing facilitates the detection of subtle decline in exercise capacity, and echocardiography permits assessment of RV size and function. Development or progression of secondary TR should prompt consideration of PVR, and the onset of atrial arrhythmias should prompt an assessment of the underlying hemodynamics. The exact timing for PVR remains controversial because all available bioprosthetic valves have a finite life expectancy.

The current treatment for severe valvular PS is balloon valvotomy, which has results comparable to surgical valvotomy. Current American College of Cardiology/American Heart Association guidelines recommend balloon valvotomy when the Doppler peak instantaneous gradient across the pulmonary valve is >60 mm Hg, even if the patient is asymptomatic (15). Whether the resultant pulmonary regurgitation will ultimately cause the same problems as surgical valvotomy remains to be determined.

JACC Vol. 54, No. 21, 2009
November 17, 2009:1903–10

Tetralogy of Fallot

This anomaly exemplifies the problems that beset the RV after surgical repair. Tetralogy is one of the most common cyanotic congenital heart defects and consists of sub-PS (often with coexistent valve and supravalvular stenosis), RV hypertrophy, a subaortic ventricular septal defect (VSD), and an aorta that overrides the VSD. Repair of tetralogy was first accomplished by Lillehei et al. (16) in 1954 with closure of the VSD and resection of the RVOT stenosis. Relief of the RVOT stenosis may necessitate an RVOT patch when the pulmonary annulus is small or a more extensive transannular patch; both procedures distort the pulmonary valve. If the pulmonary valve itself is abnormal, a pulmonary valvotomy or even a valvectomy may be necessary.

Late survival after tetralogy repair is excellent, but like all patients with repaired congenital heart disease, there are residua and sequelae and patients are not completely cured. An RVOT patch always results in pulmonary regurgitation, which is usually well tolerated for many years. The RV maintains systolic function for a long time unless there is an added hemodynamic burden such as peripheral PS that impedes forward flow or another volume lesion such as a residual VSD. Atrial tachycardia or fibrillation may also accelerate RV dysfunction. Often, after 2 decades or so, RV contractile function deteriorates, resulting in decreased stroke volume and increased RV end-systolic volumes. Importantly, severe RV enlargement and severe RV dysfunction can antedate the onset of symptoms. Most patients after tetralogy repair have right bundle branch block on electrocardiography, but the wider the QRS, the worse the RV function—the so-called "mechano-electrical interaction" (17). When the QRS duration exceeds 180 ms, patients are more vulnerable to ventricular tachycardia (VT) and sudden death; however, this correlation is inexact in individuals, and precise risk stratification remains challenging.

As such, patients should be considered for PVR before there is irreversible RV dysfunction. Certainly PVR should be performed when patients develop dyspnea, but many patients with congenital heart disease have no perception of "normal" and may therefore have advanced RV dysfunction by the time they complain of symptoms. Serial exercise testing may help to delineate subtle changes in exercise capacity before the patient becomes symptomatic.

In addition to exercise capacity, the size and function of the RV help to determine appropriate timing of PVR. Indications for PVR include moderate to severe RV enlargement, moderate to severe RV dysfunction, moderate to severe TR that develops as the RV dilates, and symptomatic atrial and ventricular arrhythmias that usually manifest as part of the underlying hemodynamic problem.

Usually no single parameter, but several variables, are considered to facilitate correct timing of PVR (15). Coexistent PS, although uncommon, may necessitate earlier operation because the RV then has to withstand both pressure and volume load, and patients are often symptomatic before the RV is severely dilated. When there is significant TR, concomitant repair or replacement of the tricuspid valve may also be necessary. Cryoablation of the RVOT may be performed to treat ventricular tachyarrhythmias, and a concomitant Maze procedure if there are atrial arrhythmias can also be accomplished with little prolongation of the operative time.

Magnetic resonance imaging is increasingly being used to measure RV volumes and function to facilitate timing of PVR. In 2002, Vliegen et al. (18) demonstrated that after PVR, patients improved their New York Heart Association functional classification and RV volumes diminished, but RVEF did not improve. The authors concluded that PVR should be performed earlier. In 2004, Geva et al. (19) reported that all patients with repaired tetralogy who had an RV end-systolic volume index >95 ml/m^2 had RV dysfunction and that patients who had an RVEF $<35\%$ tended to have worse clinical status. Importantly, they also reported the unfavorable ventricular-ventricular interaction that occurs with severe RV enlargement; if the RVEF was impaired, the LVEF was also worse.

More recently, "cut-off" values for RV volumes have been described beyond which the RV size will never normalize. In a small series, Therrien et al. (20) reported that only those with RV end-diastolic volumes <170 ml/m^2 normalized their end-diastolic volumes after PVR and only those with end-systolic volumes <85 ml/m^2 could normalize their end-systolic volumes. Oosterhof et al. (21) reported that even very large RVs get smaller after PVR, but normalization of RV size only occurred when the pre-operative RV end-diastolic volume was <160 ml/m^2. The RVEF did not improve, however, and only 17% of patients normalized RV end-systolic volumes. More recently the proposed "cut-off" value has moved even lower to 150 ml/m^2 (22). In none of these studies was consistent improvement in RVEF observed. Because of the challenges of RV measurements outlined previously, RV volume alone should not be the only variable used to determine timing of PVR.

Whether achieving *normalization* of RV volumes improves long-term outcomes remains uncertain, and there are no data that preserving RV *function* by earlier operation portends a better prognosis. Although the bad RV is associated with an increased risk for VT and sudden death, there is no evidence that PVR alone protects against subsequent VT. Because QRS duration may be a "proxy" for RV function, PVR alone does not usually result in shortening of the QRS duration. A recent nonrandomized study (23) reported that PVR conferred no improvement in survival or the incidence of VT compared with controls, although both groups were fairly small and the follow-up was relatively short. It is possible that in both groups, the RV size and dysfunction were already advanced and surgery was too late to confer a survival advantage.

Thus, although the "window of opportunity" for PVR is shifting earlier, we await evidence that earlier surgery to "save" the RV results in improved survival and decreased

incidence of VT. This may be because surgery is still being performed too late, and the focus should be on the preservation of RVEF rather than RV volume. Certainly this is in contrast to the LV, for which maintenance of a normal EF is the "holy grail" of valve intervention. Although normalization of RV volume and RVEF could be achieved with earlier operation, it remains unproven whether this would result in better long-term outcomes, especially because pulmonary bioprostheses have a limited life span, necessitating reoperation.

Ebstein Anomaly

Ebstein anomaly is an uncommon congenital abnormality that involves failure of delamination of the tricuspid valve and variable degrees of TR. The tricuspid valve is displaced inferiorly with an "atrialized" RV above and a true RV below. In extreme cases, the atrialized portion of the RV can occupy more than one-half of the RV volume, and the RV enlargement may be so pronounced that the ventricular septum shifts leftward, compressing the LV. The LV itself may also be abnormal (24). The anomaly encompasses a variable spectrum, and patients may present in infancy with cyanosis and heart failure, or in adulthood with dyspnea, fatigue, palpitations, or cyanosis when there is an associated atrial septal communication.

Not well recognized as part of the anomaly is the accompanying RV myopathy that adds to the propensity for RV dysfunction, in addition to that posed by the TR. The RV is usually very thin walled (Fig. 3) and vulnerable to progressive dilation and dysfunction as TR increases. Like other congenital heart diseases, subtle decline in exercise capacity may go unnoticed until severe RV dysfunction and severe TR supervenes. In addition, the clinical signs of TR are easily missed because the right atrium is so large and encompasses all the volume of the regurgitant jet without elevation of the jugular venous pressure. The TR may also be underestimated on echocardiography because color flow may be laminar with low velocity. For these reasons, adults with Ebstein anomaly are often referred late with severe RV enlargement and dysfunction (Fig. 4). Historically, the results of tricuspid valve operation were also poor, in part because of the advanced RV dysfunction, more severe forms of the anomaly, and worse functional status.

In the current era, however, tricuspid valve repair or replacement can be accomplished with a low mortality and morbidity in experienced centers, particularly when patients do not have severe pre-operative RV dysfunction. A recent study of 539 patients (25) reported an early mortality of 6.3%. Importantly, pre-operative variables of higher mortality included RV and LV dysfunction, emphasizing the need for earlier referral and consideration of operation. In the earlier era, indications for surgery included a cardiothoracic ratio on chest radiograph of >65%. In the last decade, however, we recognized that when the cardiac silhouette is so large, the RV is severely enlarged because the LV is usually relatively small; therefore, when possible, surgery was performed earlier. Improvement in functional class and exercise capacity resulted in the majority of patients, and 10-year survival free of any reoperation was 74% (25).

Figure 3 **Pathologic Specimen Cut in the 4-Chamber Plane From a Patient With Ebstein Anomaly**

The tricuspid valve is displaced markedly inferiorly, and the right ventricular wall is extremely thin. Photo courtesy of Dr. W. D. Edwards, consultant in pathology, Mayo Clinic.

Congenitally Corrected Transposition

This anomaly demonstrates the remarkable ability of the RV to adapt to systemic pressure. It comprises both atrioventricular (AV) and ventriculoarterial discordance so that the right atrium enters the morphologic LV, which gives rise to the pulmonary artery, and the left atrium enters the morphologic RV, which gives rise to the aorta. Thus, blood flows in the normal direction but through the "wrong" ventricle (Fig. 5). Because the tricuspid valve always enters the morphologic RV, it too is on the left side in the systemic circulation and is more correctly termed the systemic AV valve (26). The single right coronary artery supplies the morphologic RV, making it vulnerable to perfusion mismatch and ischemia (27). Interestingly, the contraction pattern of a systemic (subaortic) RV may resemble that of a normal LV, with predominant circumferential shortening over longitudinal free wall shortening, opposite from the normal RV. This may represent an adaptive response to the systemic load (28).

JACC Vol. 54, No. 21, 2009
November 17, 2009:1903–10

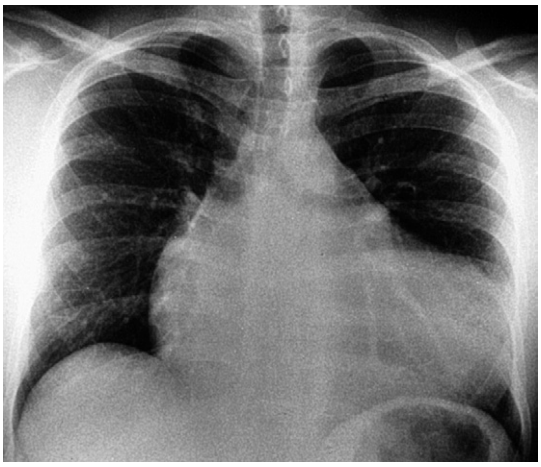

Figure 4 **Chest Radiograph of a 32-Year-Old Man With Severe Ebstein Anomaly**

The patient was referred late with severe right ventricular (RV) enlargement, severe RV dysfunction, and severe tricuspid regurgitation. He was cyanotic secondary to a small atrial septal defect and could walk only a few yards.

It is a misconception that when the morphologic RV is the systemic ventricle, it inevitably fails early, and remarkably, survival to the seventh and eighth decade has been reported. An added volume lesion, however, will precipitate ventricular dysfunction, and most commonly, failure of the morphologic RV occurs as a sequel to systemic AV valve regurgitation (29). The systemic (tricuspid) valve may have marked inferior displacement, the so-called "Ebstein-like" deformity of the systemic AV valve, although the inferior displacement is the only morphologic feature it shares with Ebstein anomaly of the right AV valve. In this setting, AV valve regurgitation is even more likely, and such patients should be evaluated regularly. At the earliest sign of deterioration in systemic ventricular function, systemic AV valve regurgitation should be suspected (30,31).

Much has been written about the importance of early surgery for mitral regurgitation and the imperative of maintaining LV function. In stark contrast, however, it is disappointing that patients with congenitally corrected transposition are often referred for surgery late with severe systemic AV valve regurgitation and advanced ventricular dysfunction. In a Mayo Clinic series, 59% of patients were referred with significant (grade >3/4) AV valve regurgitation, and more than one-half of those who needed systemic AV valve replacement were referred late with clinical ventricular dysfunction >6 months (32). Systemic AV valve replacement can be accomplished with acceptable mortality in experienced centers, but ventricular function remains depressed (33).

An important marker for poor survival was a poor pre-operative systemic EF, and patients surviving operation are at risk of needing cardiac transplantation in the subsequent decade. If systemic AV valve replacement is performed before the EF is depressed, however, systemic ventricular function may be maintained even in older patients (Fig. 6).

Conclusions

Adults with congenital heart disease often have right-sided cardiac disease, which has a negative impact on long-term outcomes. This may occur as part of their intrinsic abnor-

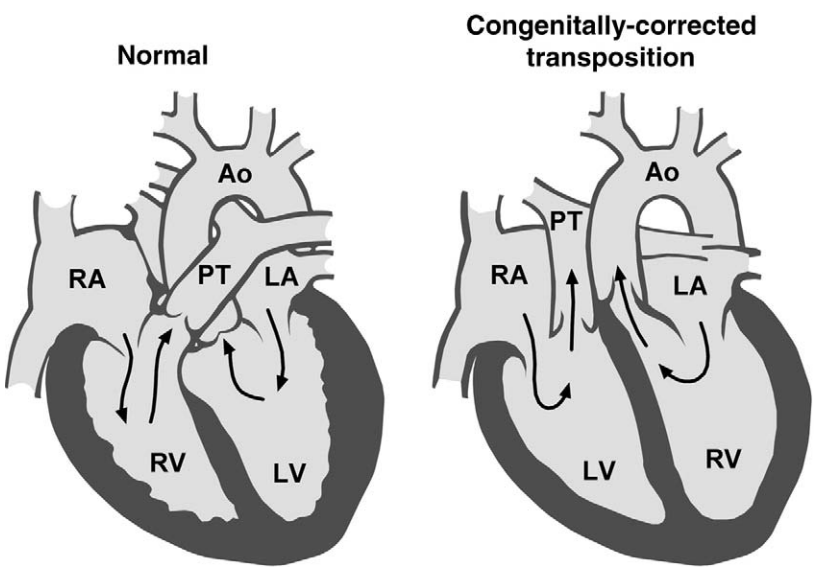

Figure 5 **Congenitally Corrected Transposition**

Schematic diagram of a normal heart **(left)** and congenitally corrected transposition **(right)** in which there is both atrioventricular and ventriculoarterial discordance. Reproduced with permission from Warnes (31). Ao = aorta; LA = left atrium; LV = morphologic left ventricle; PT = pulmonary trunk; RA = right atrium; RV = morphologic right ventricle.

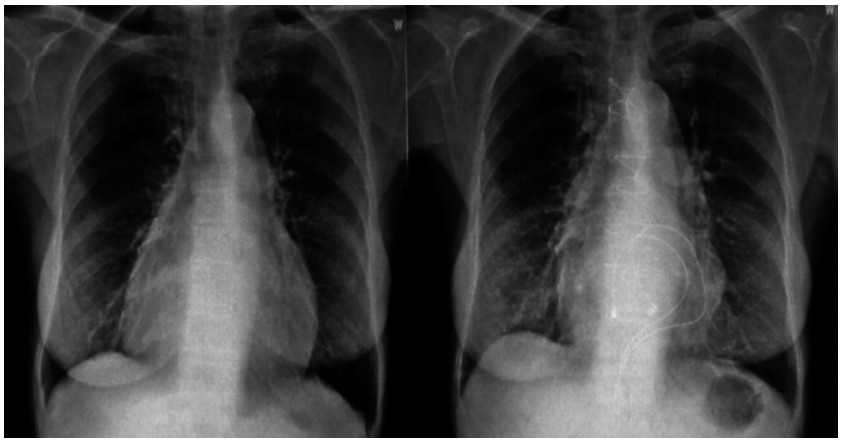

Figure 6 **Chest Radiographs of a Patient With Congenitally Corrected Transposition Before and After a Mechanical Systemic AV Valve Replacement**

At age 71 years **(left)**, the patient had severe atrioventricular (AV) valve regurgitation, but the function of her systemic ventricle was preserved with an ejection fraction (EF) of 50%. Now at age 85 years **(right)**, the EF of her systemic ventricle is still 50%, and her exercise capacity exceeds 100% of predicted.

mality or as a result of prior surgery. When the RV is significantly enlarged and dysfunctional, there is an increased likelihood of exercise limitation, VT, and sudden death. Enlargement and dysfunction of the RV may be easily overlooked, particularly when patients do not complain of symptoms. Patients should be followed at congenital heart disease centers where there is an understanding of these problems so that earlier valvular surgery may be considered when appropriate. The precise timing of these operations remains a "moving target"; although the trend is to operate earlier to improve RV size and symptoms, we await data indicating that preservation of RV function to improve outcome and survival should be the goal.

Reprint requests and correspondence: Dr. Carole A. Warnes, Mayo Clinic, Gonda 5-368, 200 First Street SW, Rochester, Minnesota 55905. E-mail: warnes.carole@mayo.edu.

REFERENCES

1. Staab ME, Nishimura RA, Dearani JA. Isolated tricuspid valve surgery for severe tricuspid regurgitation following prior left heart valve surgery: analysis of outcome in 34 patients. J Heart Valve Dis 1999;8:567–74.
2. Naito H, Arisawa J, Harada K, Yamagami H, Kozuka T, Tamura S. Assessment of right ventricular regional contraction and comparison with the left ventricle in normal humans: a cine magnetic resonance study with presaturation myocardial tagging. Br Heart J 1995;74:186–91.
3. Ho SY, Nihoyannopoulos P. Anatomy, echocardiography, and normal right ventricular dimensions. Heart 2006;92 Suppl 1:i2–13.
4. Lang RM, Bierig M, Devereux RB, et al. Recommendations for chamber quantification. Eur J Echocardiogr 2006;7:79–108.
5. Sheehan FH, Ge S, Vick GW 3rd, et al. Three-dimensional shape analysis of right ventricular remodeling in repaired tetralogy of Fallot. Am J Cardiol 2008;101:107–13.
6. Winter MM, Bernink FJ, Groenink M, et al. Evaluating the systemic right ventricle by CMR: the importance of consistent and reproducible delineation of the cavity. J Cardiovasc Magn Reson 2008;10:40.
7. Niemann PS, Pinho L, Balbach T, et al. Anatomically oriented right ventricular volume measurements with dynamic three-dimensional echocardiography validated by 3-Tesla magnetic resonance imaging. J Am Coll Cardiol 2007;50:1668–76.
8. Eidem BW, O'Leary PW, Tei C, Seward JB. Usefulness of the myocardial performance index for assessing right ventricular function in congenital heart disease. Am J Cardiol 2000;86:654–8.
9. Harada K, Tamura M, Toyono M, Yasuoka K. Comparison of the right ventricular Tei index by tissue Doppler imaging to that obtained by pulsed Doppler in children without heart disease. Am J Cardiol 2002;90:566–9.
10. Vogel M, Derrick G, White PA, et al. Systemic ventricular function in patients with transposition of the great arteries after atrial repair: a tissue Doppler and conductance catheter study. J Am Coll Cardiol 2004;43:100–6.
11. Gatzoulis MA, Clark AL, Cullen S, Newman CG, Redington AN. Right ventricular diastolic function 15 to 35 years after repair of tetralogy of Fallot. Restrictive physiology predicts superior exercise performance. Circulation 1995;91:1775–81.
12. Hayes CJ, Gersony WM, Driscoll DJ, et al. Second natural history study of congenital heart defects. Results of treatment of patients with pulmonary valvar stenosis. Circulation 1993;87:I28–37.
13. Earing MG, Connolly HM, Dearani JA, Ammash NM, Grogan M, Warnes CA. Long-term follow-up of patients after surgical treatment for isolated pulmonary valve stenosis. Mayo Clin Proc 2005;80:871–6.
14. Shimazaki Y, Blackstone EH, Kirklin JW. The natural history of isolated congenital pulmonary valve incompetence: surgical implications. Thorac Cardiovasc Surg 1984;32:257–9.
15. Warnes CA, Williams RG, Bashore TM, et al. ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease). J Am Coll Cardiol 2008;52:e1–121.
16. Lillehei CW, Cohen M, Warden HE, et al. Direct vision intracardiac surgical correction of the tetralogy of Fallot, pentalogy of Fallot, and pulmonary atresia defects; report of first ten cases. Ann Surg 1955; 142:418–42.
17. Gatzoulis MA, Till JA, Somerville J, Redington AN. Mechanoelectrical interaction in tetralogy of Fallot. QRS prolongation relates to right ventricular size and predicts malignant ventricular arrhythmias and sudden death. Circulation 1995;92:231–7.
18. Vliegen HW, van Straten A, de Roos A, et al. Magnetic resonance imaging to assess the hemodynamic effects of pulmonary valve replacement in adults late after repair of tetralogy of Fallot. Circulation 2002;106:1703–7.
19. Geva T, Sandweiss BM, Gauvreau K, Lock JE, Powell AJ. Factors associated with impaired clinical status in long-term survivors of

tetralogy of Fallot repair evaluated by magnetic resonance imaging. J Am Coll Cardiol 2004;43:1068–74.
20. Therrien J, Provost Y, Merchant N, Williams W, Colman J, Webb G. Optimal timing for pulmonary valve replacement in adults after tetralogy of Fallot repair. Am J Cardiol 2005;95:779–82.
21. Oosterhof T, van Straten A, Vliegen HW, et al. Preoperative thresholds for pulmonary valve replacement in patients with corrected tetralogy of Fallot using cardiovascular magnetic resonance. Circulation 2007;116:545–51.
22. Dave HH, Buechel ERV, Dodge-Khatami A, et al. Early insertion of a pulmonary valve for chronic regurgitation helps restoration of ventricular dimensions. Ann Thorac Surg 2005;80:1615–20.
23. Harrild DM, Berul CI, Cecchin F, et al. Pulmonary valve replacement in tetralogy of Fallot: impact on survival and ventricular tachycardia. Circulation 2009;119:445–51.
24. Attenhofer Jost CH, Connolly HM, Dearani JA, Edwards WD, Danielson GK. Ebstein's anomaly. Circulation 2007;115:277–85.
25. Brown ML, Dearani JA, Danielson GK, et al. Functional status after operation for Ebstein anomaly: the Mayo Clinic experience. J Am Coll Cardiol 2008;52:460–6.
26. Warnes CA. Congenitally corrected transposition: the uncorrected misnomer. J Am Coll Cardiol 1996;27:1244–5.
27. Hauser M, Bengel FM, Hager A, et al. Impaired myocardial blood flow and coronary flow reserve of the anatomical right systemic ventricle in patients with congenitally corrected transposition of the great arteries. Heart 2003;89:1231–5.
28. Pettersen E, Helle-Valle T, Edvardsen T, et al. Contraction pattern of the systemic right ventricle shift from longitudinal to circumferential shortening and absent global ventricular torsion. J Am Coll Cardiol 2007;49:2450–6.
29. Prieto LR, Hordof AJ, Secic M, Rosenbaum MS, Gersony WM. Progressive tricuspid valve disease in patients with congenitally corrected transposition of the great arteries. Circulation 1998;98:997–1005.
30. Warnes CA. The adult with congenital heart disease: born to be bad? J Am Coll Cardiol 2005;46:1–8.
31. Warnes CA. Transposition of the great arteries. Circulation 2006;114:2699–709.
32. Beauchesne LM, Warnes CA, Connolly HM, Ammash NM, Tajik AJ, Danielson GK. Outcome of the unoperated adult who presents with congenitally corrected transposition of the great arteries. J Am Coll Cardiol 2002;40:285–90.
33. van Son JA, Danielson GK, Huhta JC, et al. Late results of systemic atrioventricular valve replacement in corrected transposition. J Thorac Cardiovasc Surg 1995;109:642–52.

Key Words: adult congenital ■ pediatric ■ heart disease.