Int J Cardiovasc Imaging (2008) 24:867–870
DOI 10.1007/s10554-008-9344-y

EDITORIAL COMMENT

Diastolic dysfunction: a new additional criterion for optimal timing of pulmonary valve replacement in adult patient with tetralogy of Fallot?

Barbara J. M. Mulder · Hubert W. Vliegen · Ernst E. van der Wall

Received: 8 July 2008 / Accepted: 8 July 2008 / Published online: 24 July 2008

Over the past decades, life expectancy of patients with congenital heart disease has dramatically increased. The prognosis of patients with tetralogy of Fallot (TOF) was invariably fatal until the development of palliative and later corrective surgical procedures. Repaired TOF has an excellent long-term prognosis, but survival is still suboptimal. Serious complications may develop late after total repair during infancy. These complications are usually the result of long-standing pulmonary regurgitation, leading to dilatation of the right ventricle, decreased exercise tolerance, an increased risk for severe arrhythmias, and even sudden death. Pulmonary valve replacement has beneficial effects on right ventricular size and function, provided it is performed early, before irreversible right ventricular dysfunction ensues. Moreover, pulmonary valve replacement is associated with an improvement in patients' symptoms and exercise tolerance. Combined with arrhythmia surgery pulmonary valve replacement leads to a dramatic decrease in the incidence of fatal ventricular arrhythmias. Although late pulmonary valve replacement offers excellent mid-term results, homografts and xenografts, usually used for right ventricular outflow tract reconstruction, suffer late dysfunction and failure, committing patients and surgeons to further operations. Therefore, the timing of surgery must be carefully considered, weighing the up-front risks of surgery and possible repeat surgery against the risk of ongoing pulmonary regurgitation [1, 2]. Over the past years, many investigators have attempted to find the optimal moment of surgery [3–8], but presently there are no universally accepted criteria to determine optimal timing for valved conduit placement in patients with TOF.

A combination of clinical, electrocardiographic and imaging criteria is presently used to determine the timing of valve replacement. Exercise tolerance, signs of heart failure, prolonged QRS duration play an important role, but nowadays criteria based on cardiovascular imaging are gaining increased interest. In the past 20 years, cardiovascular magnetic resonance (CMR) imaging has become a predominant tool in the clinical management of patients with cardiovascular disease [9–15], in particular in patients with congenital heart disease [16–24]. CMR imaging has evolved into an imaging technique that provides adequate and unique information on residual problems in the follow-up of patients operated for TOF [8]. Spin-echo and gradient-echo CMR imaging allow detailed assessment of intracardiac and large vessel anatomy, which is particularly helpful in TOF patients with residual abnormalities

B. J. M. Mulder
Cardiology Department, B2-240 Academic Medical Center, Meibergdreef 9, 1105 AZ Amsterdam, The Netherlands

H. W. Vliegen · E. E. van der Wall (✉)
Department of Cardiology, Leiden University Medical Center, Leiden, The Netherlands
e-mail: e.e.van_der_wall@lumc.nl

of right ventricular outflow and/or pulmonary artery. Multisection gradient-echo cine CMR can be used to obtain accurate measurements of biventricular size, left and right ventricular ejection fraction, and wall mass. This allows serial follow-up of biventricular function. Knauth et al. showed that severe right ventricular dilatation and either left or right ventricular dysfunction assessed by CMR predicted major adverse clinical events [25].

At present, CMR velocity mapping is the only imaging technique available that provides practical quantification of pulmonary regurgitation volume. CMR velocity mapping can also be used to quantify right ventricular diastolic function in the presence of pulmonary regurgitation. Niezen et al. used CMR imaging to assess right ventricular diastolic function in young patients with corrected TOF and pulmonary regurgitation [26]. Nineteen children with repair of TOF and 12 healthy children were studied using CMR The authors showed impaired relaxation and restriction to filling affect right ventricular function in children with repair of TOF and pulmonary regurgitation. Based on the above-mentioned studies, CMR imaging may play a key role in indications, accurate diagnosis and prognosis, and timing of pulmonary valve replacement after TOF repair [27].

In a recent study, Oosterhof et al. analyzed preoperative thresholds of right ventricular volumes above which no decrease or normalization of right ventricular size takes place after surgery [28]. The authors studied 71 adult patients with corrected TOF, who underwent pulmonary valve replacement in a nationwide, prospective follow-up study. Patients were evaluated with CMR imaging both preoperatively and postoperatively. The authors could not find a threshold above which right ventricular volumes did not decrease after surgery. Preoperative right ventricular volumes were independently associated with right ventricular remodeling and also when corrected for a surgical reduction of the right ventricular outflow tract. However, normalization could be achieved when preoperative enddiastolic volume was <160 ml/m^2 or right ventricular endsystolic volume was <82 ml/m^2.

The purpose of the present study by Greenberg et al. reported in this issue, was to identify E- and A-wave flow patterns across the tricuspid valve in TOF patients [29]. Results from CMR phase contrast velocity-encoded flow quantification correlated well with measurements of right ventricular enlargement. The authors studied 33 children following TOF repair who had CMR examinations that included cine imaging to quantify ventricular size and function and flow analysis across the atria-ventricular valves to evaluate ventricle in-flow patterns. The E:A ratio was calculated for each patient and the population was separated into two different groups: alpha (E:A ratio ≥ 1.4) and beta (E:A ratio < 1.4) groups. A significant association was present between the group with E:A ratio < 1.4 and right ventricle enddiastolic volume index ≥ 140 ml/m^2 ($P = 0.046$), right ventricular endsystolic volume index ≥ 70 ml/m^2 ($P = 0.02$), and enddiastolic volume right ventricle to left ventricle ≥ 2.0 ($P = 0.003$). A reduction in the E:A wave ratio across the tricuspid valve was associated with right ventricular diastolic dysfunction and correlated well with right ventricular enlargement. The reduction in the E:A wave ratio across the tricuspid valve may be considered a new useful criterion for determining the timing of valved pulmonary conduit surgery in children following TOF repair.

The decision to operate adult patients with TOF for pulmonary regurgitation should be based on the balance between progressive right ventricular dilatation, exercise intolerance, symptoms, the occurrence of arrhythmias and the fact that further reoperations will be needed [30–34]. Research on the ideal valve for right ventricular outflow tract reconstruction is ongoing. Oosterhof et al. analyzed the long-term outcomes after pulmonary valve replacement in patients with a previous correction for TOF [35]. In a retrospective study, 158 adult patients who had undergone a pulmonary valve replacement after initial total correction for TOF in childhood, were identified from the Dutch CONCOR (CONgenital CORvitia) registry [36]. All patients underwent pulmonary valve replacement between June 1986 and June 2005. After 10 years of pulmonary valve replacement, 47% of the patients were free from homograft dysfunction and event-free survival after pulmonary valve replacement was 78%. Significant residual lesions directly after surgery influenced event-free survival. A smaller diameter of the pulmonary homograft and severe pre-surgical pulmonary regurgitation were related to early homograft dysfunction after surgery.

Prospective follow-up of patients with TOF with exercise testing and assessment of right ventricular

size and function will better define the natural history of the disease. At present, CMR imaging plays a key role in assessing ventricular function and volumes, thereby predicting clinical outcomes and providing firm guidelines for timing of pulmonary valve replacement, especially in asymptomatic patients. The current study of Greenberg et al. [29] provides a valid additional CMR criterion to determine proper timing of surgery in adult patients with TOF and serious pulmonary regurgitation.

References

1. Greenberg SB, Faerber EN, Balsara RK (1995) Tetralogy of Fallot: diagnostic imaging after palliative and corrective surgery. J Thorac Imaging 10:26–35
2. Gatzoulis MA, Balaji S, Webber SA et al (2000) Risk factors for arrhythmia and sudden cardiac death late after repair of tetralogy of Fallot: a multicentre study. Lancet 356:975–981
3. Therrien J, Marx GR, Gatzoulis MA (2002) Late problems in tetralogy of Fallot-recognition, management, and prevention. Cardiol Clin 20:395–404
4. van Dijkman PR, Voskuil K, Hazekamp MG, van der Wall EE (1996) Imaging of recurrent ventricular septal defect and supravalvular pulmonary stenosis eight years after assumed total surgical repair of tetralogy of Fallot. Int J Card Imaging 12:79–83
5. Henkens IR, van Straten A, Schalij MJ et al (2007) Predicting outcome of pulmonary valve replacement in adult tetralogy of Fallot patients. Ann Thorac Surg 83:907–911
6. Hooft van Huysduynen B, Henkens IR et al (2008) Pulmonary valve replacement in tetralogy of Fallot improves the repolarization. Int J Cardiol 124:301–306
7. Gregg D, Foster E (2007) Pulmonary insufficiency is the nexus of late complications in tetralogy of Fallot. Curr Cardiol Rep 9:315–322
8. Helbing WA, Roest AA, Niezen RA et al (2002) ECG predictors of ventricular arrhythmias and biventricular size and wall mass in tetralogy of Fallot with pulmonary regurgitation. Heart 88:515–519
9. Holman ER, Buller VG, de Roos A et al (1997) Detection and quantification of dysfunctional myocardium by magnetic resonance imaging. A new three-dimensional method for quantitative wall-thickening analysis. Circulation 95:924–931
10. van Rugge FP, Holman ER, van der Wall EE et al (1993) Quantitation of global and regional left ventricular function by cine magnetic resonance imaging during dobutamine stress in normal human subjects. Eur Heart J 14:456–463
11. van der Wall EE, van Dijkman PR, de Roos A et al (1990) Diagnostic significance of gadolinium-DTPA (diethylene-triamine penta-acetic acid) enhanced magnetic resonance imaging in thrombolytic treatment for acute myocardial infarction: its potential in assessing reperfusion. Br Heart J 63:12–17
12. van Dijkman PR, van der Wall EE, de Roos A et al (1991) Acute, subacute, and chronic myocardial infarction: quantitative analysis of gadolinium-enhanced MR images. Radiology 180:147–151
13. van Rugge FP, van der Wall EE, Spanjersberg SJ et al (1994) Magnetic resonance imaging during dobutamine stress for detection and localization of coronary artery disease. Quantitative wall motion analysis using a modification of the centerline method. Circulation 90:127–138
14. Bax JJ, Lamb H, Dibbets P, Pelikan H et al (2000) Comparison of gated single-photon emission computed tomography with magnetic resonance imaging for evaluation of left ventricular function in ischemic cardiomyopathy. Am J Cardiol 86:1299–1305
15. Langerak SE, Vliegen HW, de Roos A et al (2002) Detection of vein graft disease using high-resolution magnetic resonance angiography. Circulation 105:328–333
16. Vliegen HW, van Straten A, de Roos A et al (2002) Magnetic resonance imaging to assess the hemodynamic effects of pulmonary valve replacement in adults late after repair of tetralogy of fallot. Circulation 106:1703–1707
17. van Straten A, Vliegen HW, Lamb HJ et al (2005) Time course of diastolic and systolic function improvement after pulmonary valve replacement in adult patients with tetralogy of Fallot. J Am Coll Cardiol 46:1559–1564
18. Oosterhof T, Mulder BJ, Vliegen HW, de Roos A (2006) Cardiovascular magnetic resonance in the follow-up of patients with corrected tetralogy of Fallot: a review. Am Heart J 151:265–272
19. Tulevski II, Van der Wall EE, Groenink M et al (2002) Usefulness of MRI dobutamine stress in asymptomatic and minimally symptomatic patients with decreased cardiac reserve from congenital heart disease. Am J Cardiol 89:1077–1081
20. Tulevski II, Lee PL, Groenink M et al (2000) Dobutamine-induced increase of right ventricular contractility without increased stroke volume in adolescent patients with transposition of the great arteries: evaluation with magnetic resonance imaging. Int J Cardiovasc Imaging 16:471–478
21. Dodge-Khatami A, Tulevski II, Bennink GB et al (2002) Comparable systemic ventricular function in healthy adults and patients with unoperated congenitally corrected transposition using MRI dobutamine stress testing. Ann Thorac Surg 73:1759–1764
22. Tulevski II, Zijta FM, Smeijers AS et al (2004) Regional and global right ventricular dysfunction in asymptomatic or minimally symptomatic patients with congenitally corrected transposition of the great arteries. Cardiol Young 14:168–173
23. Tulevski II, Hirsch A, Dodge-Khatami A et al (2003) Effect of pulmonary valve regurgitation on right ventricular function in patients with chronic right ventricular pressure overload. Am J Cardiol 92:113–116
24. Greenberg SB, Eshaghpour E (2001) The importance of the maximum pulmonary artery regurgitant velocity following

repair of tetralogy of Fallot: a pilot study. Int J Cardiovasc Imaging 17:221–226
25. Knauth AL, Gauvreau K, Powell AJ et al (2008) Ventricular size and function assessed by cardiac MRI predict major adverse clinical outcomes late after tetralogy of Fallot repair. Heart 94:211–216
26. Niezen RA, Helbing WA, van der Wall EE, van der Geest RJ, Rebergen SA, de Roos A (1996) Biventricular systolic function and mass studied with MR imaging in children with pulmonary regurgitation after repair for tetralogy of Fallot. Radiology 201:135–140
27. Geva T (2006) Indications and timing of pulmonary valve replacement after tetralogy of Fallot repair. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2006:11–22
28. Oosterhof T, van Straten A, Vliegen HW et al (2007) Preoperative thresholds for pulmonary valve replacement in patients with corrected tetralogy of Fallot using cardiovascular magnetic resonance. Circulation 116:545–551
29. Greenberg SB, Shah CC, Bhutta ST (2008) Tricuspid valve magnetic resonance imaging phase contrast velocity-encoded flow quantification for follow up of tetralogy of Fallot. Int J Cardiovasc Imaging. doi:10.1007/s10554-008-9331-3
30. Oosterhof T, Jacobs M, Cramer MJ, Mulder BJ (2006) Survival into seventh decade after a potts palliation for tetralogy of Fallot. Congenit Heart Dis 2:55–57
31. Davlouros PA, Karatza AA, Gatzoulis MA, Shore DF (2004) Timing and type of surgery for severe pulmonary regurgitation after repair of tetralogy of Fallot. Int J Cardiol 97 (Suppl) 1:91–101
32. Mulder BJ, van der Wall EE (2005) Pulmonary valve replacement in patients with tetralogy of Fallot and pulmonary regurgitation: early surgery similar to optimal timing of surgery? Eur Heart J 26:2614–2615
33. Henkens IR, Van Straten A, Hazekamp MG et al (2007) Preoperative determinants of recovery time in adult Fallot patients after late pulmonary valve replacement. Int J Cardiol 121:123–124
34. Greenberg SB, Eshaghpour E (2001) The importance of the maximum pulmonary artery regurgitant velocity following repair of tetralogy of Fallot: a pilot study. Int J Cardiovasc Imaging 17:221–226
35. Oosterhof T, Meijboom FJ, Vliegen HW et al (2006) Long-term follow-up of homograft function after pulmonary valve replacement in patients with tetralogy of Fallot. Eur Heart J 27:1478–1484
36. van der Velde ET, Vriend JW, Mannens MM, Uiterwaal CS, Brand R, Mulder BJ (2005) CONCOR, an initiative towards a national registry and DNA-bank of patients with congenital heart disease in the Netherlands: rationale, design and first results. Eur J Epidemiol 20:549–557