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Echocardiographic evaluation of the systemic ventricle after atrial switch procedure. The usefulness of subcostal imaging

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Abstract

Background: *Subcostal planes allow demonstration of the entire right ventricular cavity and are frequently used in patients with congenital heart disease; however, their clinical utility in the evaluation of systemic right ventricular function after atrial switch procedure for complete transposition has never been verified in adolescent and adult patients.*

Methods: *In unselected patients with simple transposition who had had an atrial switch performed between 1982 and 1990, echocardiographic and myocardial perfusion imaging were performed. Systolic function of the right ventricle was assessed from the subcostal window, and the right ventricular area change was calculated. Right ventricular systolic function was defined as impaired when the right ventricular area change was equal to or less than 0.35.*

Results: *Sixty [43 male and 17 female, mean age (standard deviation) 14.9 (4.5) years] patients were included in the analysis. Echocardiographic right ventricular area change ranged from 0.14 to 0.66 [0.42 (0.12)]. Twenty-one patients (35%) had significant impairment of right ventricular systolic function [0.29 (0.06)]. Right ventricular area change equal to or less than 0.35 detected moderate-to-severe perfusion abnormalities with 78% sensitivity and 62% specificity.*

Conclusions: *Right ventricular area change evaluated from the subcostal plane provides significant clinical information in patients with complete transposition. A cutoff value of 0.35 can be used as an indication of right ventricular impairment associated with significant perfusion abnormalities.* (Cardiol J 2008; 15: 156–161)

Key words: transposition of great arteries, right ventricular function, perfusion abnormalities

Introduction

Atrial switch was the treatment of choice for patients with transposition of the great arteries long before the introduction of the arterial switch operation. The procedure has dramatically changed the prognosis of children with this disease, but late follow-up has identified several unfavourable squeals, including progressive right ventricular failure and sudden death.

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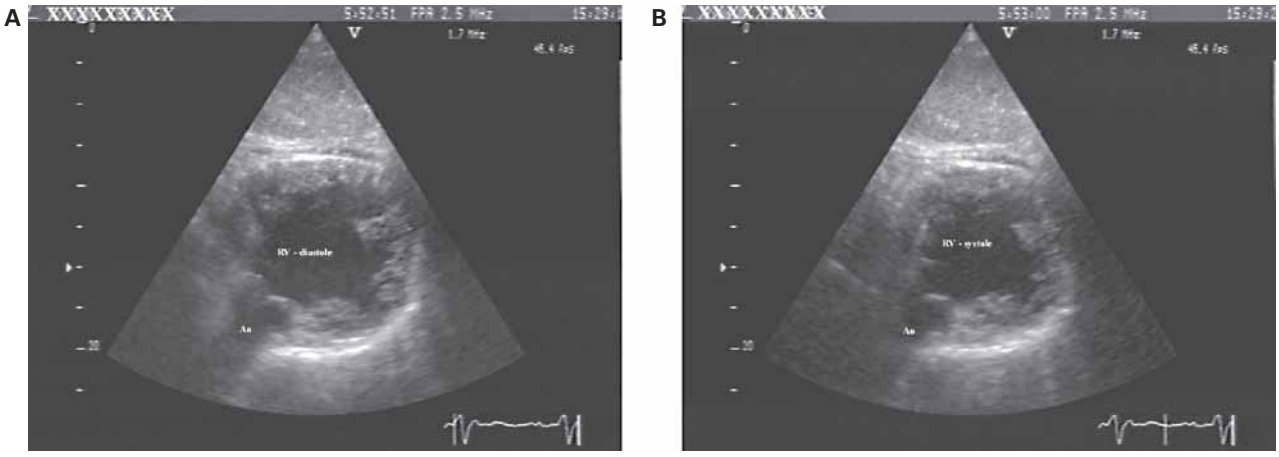

Figure 1. Echocardiographic assessment of the right ventricular area change from the subcostal window, enabling simultaneous demonstration of the entire right ventricular cavity (both inflow and outflow tracts). Right ventricle (RV) in the end-diastolic (**A**) and in the end-systolic (**B**) frames.

The use of echocardiography for evaluation of right ventricular function is limited because of the complex spatial morphology of the chamber, which prevents quantitative studies in standard echocardiographic planes. After the Mustard/Senning procedure, the right ventricle has to work as a systemic ventricle, which results in its substantial remodelling over time [1–4]. Long after atrial switch, the right ventricle resembles the left in shape, but the standard apical planes used to evaluate left ventricular systolic function do not allow its comprehensive functional assessment. Subcostal imaging planes, on the contrary, permit simultaneous demonstration of both the inflow and outflow of the right ventricle. We assessed the clinical value of the latter approach to evaluate the function of the systemic right ventricle in patients after atrial switch procedure for complete transposition.

Methods

Unselected patients with simple complete transposition who had had a Mustard or Senning operation performed between 1982 and 1990 were included. Patients with small (hemodynamically insignificant) ventricular septal defects, left ventricular outflow tract obstruction and/or patent arterial duct were included. Patients younger than eight years old and those with moderate-to-large ventricular septal defects were excluded. Each patient underwent a comprehensive clinical examination. Echocardiogram, chest X-ray, electrocardiogram, radionuclide angiography and myocardial perfusion imaging using technetium 99-m methoxyisobutyl isonitrile were performed. The right ventricular ejection fraction (right ventricular ejection fraction) was calculated from the background-corrected end-diastolic and end-systolic counts of the first-pass angiogram. Single photon emission computed tomographic images were recorded using a Siemens Orbiter Gamma Camera 750 ZLC. Images were evaluated off-line by two observers trained in nuclear cardiology. The right ventricular cross-sectional horizontal long-axis and vertical long-axis views were analyzed. Lesions, defined as areas of reduced radioactivity, were graded in a 5-point scale. Scores from three to five were defined as moderate-to-severe perfusion abnormalities. The reviewers were blinded to the results of the echocardiographic evaluation and other test results.

Echocardiographic examination

Comprehensive transthoracic two-dimensional and Doppler scans were performed with commercially available equipment. Systolic function of the right ventricle was assessed from the subcostal window, with the transducer rotated nearly into the frontal plane and tilted upwards, enabling simultaneous demonstration of the entire right ventricular cavity (i.e. both inflow and outflow tracts) (Fig. 1). The recordings were made at the end of the quiet expiration. The endocardial borders of the right ventricle in the end-diastolic and in the end-systolic frames were traced in three consecutive cardiac cycles. End-diastolic and end-systolic right ventricular endocardial borders were delineated and right ventricular area change was calculated from subcostal planes [5]. Right ventricular systolic

Table 1. Demographic data.

Variable	Mean (SD)
Age at the time of surgery (years)	3.3 (3.3)
Age at the time of the study (years)	14.9 (4.5)
Duration of postoperative follow up (years)	11.5 (2.7)
Weight [kg]	42.8 (16.9)
Height [cm]	149.9 (17.3)
Body surface area [m^2]	1.3 (0.3)

function was defined as impaired when the echocardiographic right ventricular area change was equal to or less than 0.35 (based on the receiver operating curves, drawn in order to determine the cut-off value of echocardiographic right ventricular area change with optimal sensitivity and specificity for the detection of moderate-to-severe perfusion abnormalities at rest). Tricuspid regurgitation was classified as absent, mild, moderate or severe using a standard semi-quantitative Doppler method [6]. All echocardiograms were recorded and evaluated by a single cardiologist (P.H.), experienced in adult congenital heart disease, who was blind to the results of other diagnostic investigations and clinical data.

The study protocol was approved by the Human Ethics Committee at our institution.

Statistical analysis

The data are presented as means (standard deviation). Student's t test (Mann-Whitney U test when data were not normally distributed) analysis of variance and univariate logistic regression were used for statistical analysis of the data. Variables that were statistically significant on univariate analysis were evaluated by multivariate analysis using multivariate logistic regression. Correlations between variables were assessed by Pearson or Spearman correlation coefficients. A p value of less than 0.05 was considered significant.

Results

Demographic and clinical data

Eighty patients met the eligibility criteria and 61 (76%) took part in the study. In one patient, the subcostal window was inadequate. Sixty (43 male and 17 female) patients were therefore included in the final analysis (Table 1). Nineteen patients (31.7%) had undergone the Mustard procedure and 41 patients (68.3%) the Senning procedure. Before this surgery, 52 patients (86.7%) had undergone Rashkind procedures and one had had a Blalock--Taussig shunt. Fifty-five patients were in New York Heart Association functional class I and five in class II.

Echocardiographic findings

Colour Doppler echocardiography disclosed mild tricuspid regurgitation in 28 (47%) patients, moderate regurgitation in 9 (15%) and severe regurgitation in 3 (5%). No significant obstruction of pulmonary and systemic inflow was found. Ventricular septal defect with hemodynamically insignificant left-to-right shunt was present in 6 (10%) patients. Left ventricular outflow tract obstruction due to systolic anterior movement of the mitral leaflet (with gradients from 20 mm Hg to 65 mm Hg), was detected in 9 (15%) patients. Echocardiographic right ventricular area change ranged from 0.14 to 0.66 [mean (standard deviation) 0.42 (0.12)]. Twenty-one patients (35%) had significant impairment of right ventricular systolic function 0.29 (0.06) and 39 (65%) had relatively preserved right ventricular systolic function 0.49 (0.07). Patients with impaired right ventricular area change were older at the time of operation and at the time of the study, had greater body surface area and perfusion abnormalities as well as more frequently significant tricuspid regurgitation (p equal to or less than 0.01 for all of them) (Fig. 2). Right ventricular area change correlated with: radionuclide angiographic right ventricular ejection fraction and inversely with the degree of perfusion abnormalities; severity of tricuspid regurgitation; height; weight; body surface area; age at the time of operation; age at the time of the study; the duration of follow-up; and QRS width on electrocardiogram (Table 2). A cut-off value of right ventricular area change set at 0.35 detected moderate-to-severe perfusion abnormalities with 78% sensitivity and 62% specificity. Multivariate logistic regression demonstrated that body surface area and perfusion abnormalities significantly influenced right ventricular area change (p equal to or less than 0.004 and 0.008, respectively). Age, duration of follow-up, operation type (Mustard *vs.* Senning), ventricular septal defect closure and degree of tricuspid insufficiency did not independently influence the echocardiographic right ventricular area change.

Discussion

Actuarial survival of early survivors at 10, 20, and 30 years after surgery is approximately 92%, 89% and 79%, respectively [7]. Since the vast majority of adolescent patients with complete

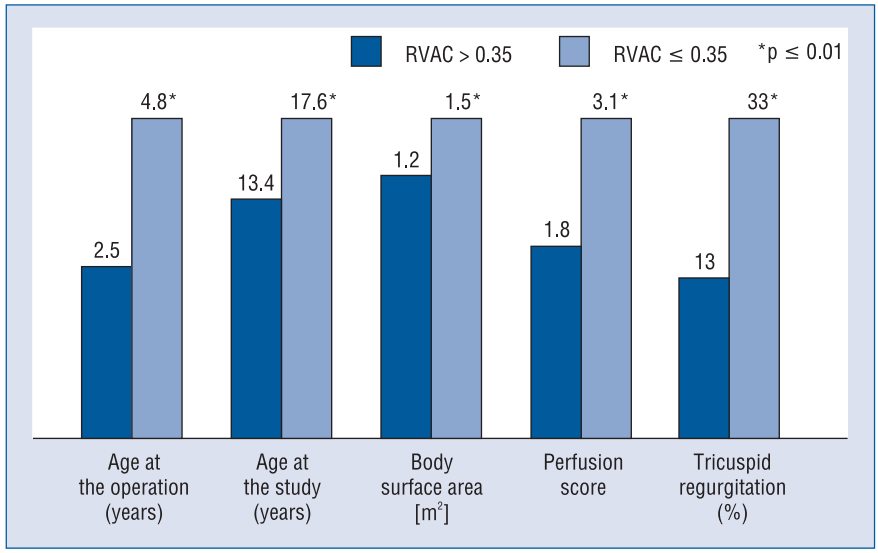

Figure 2. The comparison of selected demographic, echocardiographic and perfusion variables in patients with impaired versus preserved right ventricular function [right ventricular area change (RVAC) ≤ 0.35 *vs*. > 0.35].

Table 2. Correlation coefficients between the echocardiographically determined ejection fraction of the right ventricle and selected variables.

Variable	Correlation coefficient	p
Angiographic right ventricular ejection fraction	0.3704	0.006
Severity of tricuspid regurgitation	–0.4170	0.001
Perfusion abnormalities	–0.3756	0.004
Body surface area	–0.5603	0.0001
Age at surgery	–0.4449	0.0001
Age at the time of the study	–0.5503	0.0001
QRS width	–0.3584	0.005

transposition have undergone atrial switch procedure, cardiologists caring for adults need to be aware of its remote squeals.

The right ventricular ability to cope with systemic circulation over a normal lifetime after atrial switch has been extensively investigated. Roest et al. [8] reported an abnormal response to exercise of both systemic right and left ventricles examined with magnetic resonance imaging during exercise. Progressive right ventricular dilation and loss of contractility have been described [9]. Reich and coworkers, on the other hand, found no deterioration of systolic performance over a median interval of 8.8 years using radionuclide testing [10]. Nonetheless, diminished right ventricular performance is generally thought to be one of the key factors affecting the longer-term quality of life of these patients [11]. For this reason, a reliable and easily available method for assessment of right ventricular function is needed.

Echocardiographic evaluation of right ventricular function

In patients with complete transposition who underwent atrial correction, the right ventricle resembles the left in shape and function. Consequently, it is tempting to assess right ventricular performance using echocardiographic formule devised for the quantification of the left ventricle. Lidegran et al. [12] described the utility of transthoracic echocardiography for assessing right ventricular function in patients with complete transposition who had undergone atrial correction. These authors calculated ventricular volumes by means of a multiple-slice method applied to four- and two-chamber apical views, comparing them with magnetic resonance

imaging, and observed an overall tendency to underestimate ventricular volumes with echocardiography; this has also been reported in another study [13]. The explanation may be that apical scans leave right ventricular outflow tract out of the study planes since the complex spatial geometry of the right ventricle cannot be adequately demonstrated using the standard apical planes that are used routinely for calculating the left ventricular ejection fraction. Since, in patients after atrial correction, the right ventricle dilates considerably, eventually resembling left ventricle in shape, the entire right ventricular anatomy (both inflow and outflow) can be displayed in one subcostal plane. We took advantage of it when calculating the right ventricular area change in this study.

Subcostal view

In order to analyze the systolic performance of the entire right ventricle (inflow and outflow) we visualized the right ventricle from a subcostal view. By doing so, the ellipsoid right ventricular cavity could be demonstrated and this allowed accurate tracing of the endocardial border in diastole and systole. We are aware of only one study verifying subcostal projections in the assessment of right ventricular function, performed by Trowitzsch et al. [5] over 20 years ago in a pediatric population and verified with biplane cineangiography. No data in adults are available.

Right ventricular area change measured from subcostal view by echocardiography correlated inversely with the severity of perfusion abnormalities — impaired systolic function detected with echocardiography predicted moderate-to-severe perfusion defects with 78% sensitivity and 62% specificity. Similar information was recently provided by means of contrast-enhanced magnetic resonance imaging, in which the presence of abnormal myocardial regions was associated with right ventricular dysfunction [14]. The significant correlations between echocardiographic right ventricular area change and age at the time of operation, age at entry to the study, duration of follow-up and body surface area implied a progressive deterioration of ventricular systolic function with the growth of the patients. In other words, subcostal echocardiographic evaluations of the right ventricle provided data about its function and perfusion that were clinically important.

Limitations of the study

The right ventricular area change cutoff value of 0.35 was chosen rather arbitrarily, based on the receiver operating curves, but there is no data concerning "normal" limits of function for the systemic right ventricle area change following atrial correction of transposition of great arteries. All echocardiographic examinations were performed by one cardiologist experienced in using these techniques in adults with congenital heart disease; for this reason, we have no data about inter-observer variability. Although this is a potential source of error, the measurements we describe should be valid when undertaken by cardiologists able to recognize the late squeals of atrial switch surgery. A second limitation of the study is that radionuclide assessment of right ventricular ejection fraction serving as a reference method is not free of flaws associated, for example, with the presence of significant tricuspid regurgitation. On the other hand, it has been demonstrated that equilibrium radionuclide ventriculography shows good agreement with magnetic resonance imaging and provides a good alternative in cases in which magnetic resonance imaging is not available or appropriate [15].

Conclusions

Since the introduction of the arterial switch, Mustard/Senning procedure is no longer the surgical treatment of choice for complete transposition. At present, patients who have undergone these procedures are being cared for by cardiologists whose practice concerns adolescents and adults. This study provides a useful, widely-available means of monitoring these patients. Evaluating right ventricular function by means of subcostal planes with the application right ventricular ejection fraction area change provides significant clinical information that has not previously been verified in adults, in spite of routine use of echocardiography in these patients in their everyday assessment. The proven relationship between the right ventricular ejection fraction area change and perfusion abnormalities similar to that observed with contrast-enhanced magnetic resonance imaging suggests that the former can be used not only as a simple measure of systolic function but also as an index of right ventricular structural alterations. Hence, a right ventricular area change cutoff value of 0.35 can be used as a useful indication of right ventricular impairment associated with significant perfusion abnormalities.

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