Tricuspid valve repair improves early right ventricular and tricuspid valve remodeling in patients with hypoplastic left heart syndrome

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Objective: Tricuspid regurgitation is a significant risk factor for reoperation and mortality in patients with hypoplastic left heart syndrome. The effects of tricuspid valve repair on quantitative measures of right ventricle and tricuspid valve remodeling have not been well documented.

Methods: We reviewed retrospectively the 2-dimensional echocardiograms of 17 tricuspid valve repairs (male, n = 12; female, n = 5; median age, 30 months; age range, 1.5-53 months) performed 1 month before and after tricuspid valve repair between 2005 and 2011. From the apical 4-chamber view, we measured right ventricle end-diastolic area, right ventricle fractional area change, and tricuspid valve leaflet coaptation length. The severity of tricuspid regurgitation was graded qualitatively. A 2-sided paired *t* test was used to compare changes in tricuspid valve and right ventricle outcomes, and the Wilcoxon signed-rank test was used to compare changes in tricuspid regurgitation grades.

Results: Right ventricle end-diastolic area decreased significantly after tricuspid valve repair from 14.1 ± 5.2 to $11.8 \pm 3.9\ cm^2$ ($P = .001$), whereas right ventricle fractional area change declined from $44.4\% \pm 6.4\%$ to $39.7\% \pm 8.5\%$ ($P = .016$). The coaptation length of the lateral and septal leaflet improved significantly after tricuspid valve repair (0.4 ± 2.4 mm vs 3.1 ± 2.7 mm, $P = .002$; 2.0 ± 2.7 vs 3.4 ± 2.0 mm, $P = .036$; respectively). Furthermore, the tricuspid regurgitation grade improved after tricuspid valve repair (3.1 ± 0.6 to 1.7 ± 0.9, $P < .001$).

Conclusions: Tricuspid valve repair improved significantly the tricuspid valve coaptation length and reduced right ventricle volume in children with hypoplastic left heart syndrome. Further follow-up of decreased right ventricle function is required to determine whether this is a temporary phenomenon related to reduced right ventricle preload, permanent right ventricle dysfunction from late repair of the tricuspid valve, or unavoidable sequelae of a right ventricle exposed to systemic vascular resistance. (J Thorac Cardiovasc Surg 2013;145:446-50)

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The outcome of patients with hypoplastic left heart syndrome (HLHS) has improved significantly during the past 2 decades. However, there remain several factors that affect long-term survival negatively after the Norwood procedure. The development of tricuspid regurgitation (TR) is an ominous finding and is regarded as an important risk factor for increased mortality in children with HLHS.[1-4] Tricuspid regurgitation occurs in approximately 25% of survivors of the Norwood procedure within 10 years.[4] Tricuspid regurgitation may occur because of congenital tricuspid valve (TV) dysplasia, malformation of support structures or secondary to right ventricle (RV) dilatation. Several reports have suggested that TV repair decreases the severity of TR and may improve the functional outcome of patients with HLHS.[2-4] However, the effects of TV repair on RV and TV remodeling have not been documented well in patients with HLHS.

Recent advances in echocardiography and understanding of the mechanisms of atrioventricular valve regurgitation have made it possible to evaluate valve function, before and after intervention, both more accurately and quantitatively. Mitral valve repair in adult patients with mitral regurgitation has been reported to increase the coaptation length of mitral valve leaflets and lead to favorable mitral valve remodeling.[5,6] However, there is a paucity of data on the effect of TV repair on coaptation length, and TV and RV remodeling in congenital heart diseases with RVs exposed to the systemic circulation. Therefore, we sought to assess quantitatively the changes in TV and RV function and morphology after TV repair in patients with HLHS.

MATERIALS AND METHODS

Patients

We reviewed retrospectively all patients with classic HLHS who underwent TV repair after a stage 1 Norwood procedure at our institution between January 2005 and December 2011. The study was approved by the University of Alberta Institutional Review Board, and the need for parental consent was waived. We examined hospital records, surgical records, and echocardiographic data, and collected demographic information, diagnoses, surgical procedures, and data on the indications for TV repair, complications, and outcomes.

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Abbreviations and Acronyms

ECMO = extracorporeal membrane oxygenation
HLHS = hypoplastic left heart syndrome
RV = right ventricle
RVEDA = right ventricular end-diastolic area
RVESA = right ventricular end-systolic area
RVFAC = right ventricular fractional area change
TR = tricuspid regurgitation
TV = tricuspid valve

Surgical Procedures

Several procedures for TV repair were used, depending on the mechanism of the TR (Table 1). Posterior annuloplasty was applied in patients with central regurgitation or regurgitant flow through the posterior leaflet of the TV. We performed this technique also if there was residual regurgitant flow after cleft closure or commissure closure. Posterior annuloplasty was accomplished by approximating the anteroposterior commissure with the septal–posterior commissure using mattress pledgeted sutures. Cleft closure was performed in patients with clefts, causing significant TR. Commissure closure was used if prolapsed leaflets, near the commissure, produced TR. The repaired TV was confirmed to have adequate tricuspid annular size. Posterior annuloplasty was performed in 14 patients, cleft closure in 5 patients, commissure closure in 4 patients, and with anteroseptal annuloplasty in 2 patients. Fourteen children had concomitant procedures together with TV repair, including the modified Fontan operation in 10 patients, bidirectional Glenn in 3 patients, and aortic arch repair in 2 patients. Three children underwent TV repair after the bidirectional Glenn but before the Fontan completion. Tricuspid valve repair was performed a median of 4.0 months (range, 0.1-42.7 months) after the occurrence of significant TR.

In the latter part of the series, 3-dimensional echocardiography was helpful in preoperative evaluation and influenced intraoperative strategy.[7] However, during the study period, not all patients underwent 3-dimensional echocardiography.

Echocardiographic Examination

A full 2-dimensional Doppler and color Doppler echocardiographic assessment was conducted routinely within 1 month before and after TV repair. The apical 4-chamber view was used to evaluate TV and RV outcomes. Right ventricular end-diastolic area (RVEDA) and RV end-systolic area (RVESA) were measured by tracing the area of the RV at end diastole and end systole, respectively. Right ventricular fractional area change (RVFAC) was calculated using the following equation:

$$\text{RVFAC}\ (\%) = \frac{\text{RVEDA} - \text{RVESA}}{\text{RVEDA}} \times 100$$

Tricuspid valve end-diastolic annular diameter was the distance between the insertion of the septal and the lateral leaflets at end diastole. The septal and lateral leaflet length (Ld) was measured from the tricuspid annulus to the tip of each leaflet during the diastole (Figure 1). The length of uncoapted segments of each leaflet (Lc) was the distance from the annulus to the coaptation point at end systole. Therefore, the coaptation length for each leaflet was calculated using the following equation:

$$\text{Coaptation length} = \text{Ld} - \text{Lc}$$

The TV tenting height was the distance between the septal and lateral annuli to the coaptation point of the tricuspid leaflet at end systole. The TV tenting area was calculated by multiplying the annular diameter at end systole by 0.5 times the tenting height. The vena contracta width was measured at the narrowest region of the regurgitant flow just distal to the flow convergence area at mid systole.[8] The degree of TR was determined qualitatively. TR was graded on the ratio of the color Doppler jet to the area of the atrium. TR was ranked as none (0), trivial (1), mild (2), moderate (3), and severe (4). The RV sphericity index was the ratio between the short diameter and the long diameter of the RV at end systole.

Data Analysis

Categoric variables are described as frequencies and percentages, and continuous variables are described as mean and standard deviation or median and range. A 2-sided paired sample *t* test was used to compare changes in TV and RV outcomes, and the Wilcoxon signed-rank test was used to compare changes in TR grades before and after TV repair. SAS/Base software version 9.3 (SAS Institute Inc, Cary, NC) was used for analysis.

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RESULTS

Surgical Results

There were 19 consecutive patients who underwent TV repair for severe TR during the study period. Four of 19 children who received TV repair during the study period underwent a heart transplant a median of 6.5 months (range, 3-18 months) after TV repair. Heart transplantation was performed for 1 patient because of persistent TR and severe RV dysfunction, and was performed in 3 patients because of intractable protein-losing enteropathy despite mild TR and preserved RV function. Two patients were excluded. One patient died after TV repair. He could not be separated from cardiopulmonary bypass after the modified Norwood stage 1 operation because of severe RV dysfunction and TR, and he required extracorporeal membrane oxygenation (ECMO). This patient received 7 days of ECMO support with persistent, severe TR and RV dysfunction. Tricuspid valve repair was performed, but RV function remained severely depressed and the patient could not be weaned from ECMO. He developed mediastinitis, sepsis, and died. Thus, 1 of 19 patients died after TV repair, giving a hospital mortality of 5.3%. One patient had inadequate echocardiographic images. Therefore, 17 patients were included in the study. We studied 12 males and 5 females with a median age at tricuspid valve repair of 30 months (range, 1.5-53 months) and a median weight of 13 kg (range, 3.1-17.2 kg). Median myocardial ischemic time at TV repair was 22 minutes (range, 0-61 minutes). The median follow-up time was 31 months (range, 1-63 months).

Three patients underwent reoperation for persistent TV regurgitation. Two patients required a second posterior TV annuloplasty. In 1 of these patients, the cause of the TR was tethered TV chords. In the second patient, the TV leaked at the junction of the posterior and septal leaflet. In the third patient, a scallop of the posterior leaflet prolapsed and we closed with sutures the cleft between 2 scallops of the posterior leaflet. At reoperation we found an eccentric jet arising in the commissure between the posterior leaflet and the anterior leaflet adjacent to the previous repair. Subsequently, the cleft was closed between the anterior and