Tricuspid Valve Surgery in Adults With a Dysfunctional Systemic Right Ventricle
Repair or Replace?

Roderick W.C. Scherptong, MD; Hubert W. Vliegen, MD, PhD; Michiel M. Winter, MD; Eduard R. Holman, MD, PhD; Barbara J.M. Mulder, MD, PhD; Ernst E. van der Wall, MD, PhD; Mark G. Hazekamp, MD, PhD

Background—In patients with a right ventricle (RV) in the systemic position, tricuspid valve surgery for regurgitation beyond adolescence is a subject of debate. The aim of the present study was to evaluate the complications, survival, and benefit of tricuspid surgery in adult patients with an atrium-level correction for transposition of the great arteries or congenitally corrected transposition of the great arteries.

Methods and Results—All adult patients (n=16; 7 men, 9 women; age 35±11 years) who underwent tricuspid valvuloplasty (n=8) or replacement (n=8) in the period 1999 to 2008 were included. Complications and survival were analyzed, and postoperative changes in RV function and functional class were evaluated. Tricuspid regurgitation was graded 1 to 4 according to its severity, RV dysfunction was graded as 1 to 4 (1=no dysfunction to 4=severe dysfunction), and functional status was determined according to New York Heart Association class. Although complications occurred in 11 patients, all could be managed adequately. Three patients died 109, 180, and 659 days after surgery, respectively, the first patient after tricuspid valve replacement and the latter 2 after tricuspid valvuloplasty. Overall, tricuspid valve function improved (from grade 3.1±0.8 to 0.9±1.0; P=0.001) and functional class improved (from 2.7±0.6 to 2.1±0.8; P=0.007), whereas RV function remained unchanged. After tricuspid valvuloplasty, however, recurrent moderate tricuspid valve regurgitation was observed frequently (n=3; 37%).

Conclusions—Mortality is rather low after tricuspid surgery in adult patients with mild to moderate RV dysfunction. In general, tricuspid valve function and functional class improve significantly after surgery, and systemic RV function is preserved. Tricuspid valvuloplasty, however, is associated with a high rate of recurrence of regurgitation. (Circulation. 2009;119:1467-1472.)

Key Words: heart defects, congenital ■ transposition of great vessels ■ valves ■ surgery ■ valvuloplasty ■ survival

Patients with congenitally corrected transposition of the great arteries (ccTGA) and patients with an atrium-level correction for transposition of the great arteries (acTGA) have a morphological right ventricle (RV) that sustains the systemic circulation.1–3 In these patients, regurgitation of the systemic atrioventricular valve, the tricuspid valve (TV), is a common finding. When TV regurgitation becomes severe, it is associated with RV failure and decreased survival.4 In adolescent or younger patients, this can be treated surgically by retraining the left ventricle by means of pulmonary artery banding and subsequent relocation of the left ventricle into the systemic position. In adults beyond the second decade of life, however, results of such operations are still mainly negative.5–7 Another surgical option, replacement or repair of the TV, is controversial, because the amount of data that demonstrates favorable survival after these operations is currently insufficient. The only large study to date in which beneficial effects of TV replacement (TVR) have been demonstrated is the study by van Son et al8 published in 1995, which reports the follow-up of 40 patients with ccTGA who had surgery for TV regurgitation. In other reports, attempts to repair the TV have been described as largely unsuccessful in patients with ccTGA, and in these reports, consideration for the above-described double-switch procedure is advocated.9,10 Study results of TV operations in patients with acTGA are even scarcer than for ccTGA. The general opinion in literature is that the role of TV surgery, as a treatment option for patients with acTGA and impending RV failure, should be limited.11 However, studies that focus on systematic evaluation of TV surgery in adult patients with acTGA and RV dysfunction is rather low after tricuspid surgery in adult patients with mild to moderate RV dysfunction.

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Circulation is available at http://circ.ahajournals.org DOI: 10.1161/CIRCULATIONAHA.108.805135
dysfunction are lacking.12–15 Therefore, the aim of the present study was to evaluate the complications, survival, and benefit of tricuspid valvuloplasty (TVP) or TVR as the first option in patients with important tricuspid regurgitation and a dysfunctional RV in the systemic position.

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Methods

Patient Population
All patients referred either from other medical centers or from the outpatient clinics of our centers (Leiden, Amsterdam, Utrecht) with ccTGA or acTGA who underwent TVP or TVR for tricuspid regurgitation between 1998 and 2008 were included in the present study. Patients were considered for surgery when the combination of important tricuspid regurgitation (grade 3 or 4) and mild to moderate systemic RV dysfunction was present on echocardiographic evaluation. RV function had to be sufficiently preserved, because tricuspid surgery in the later stages of RV dysfunction had a poor outcome in previous studies.8,16 Furthermore, only those patients with functional impairment who were in New York Heart Association (NYHA) class 2 or 3 were regarded as suitable for elective TV surgery. Cardiac function and anatomy were evaluated preoperatively with cardiac MRI or multislice CT if a recent evaluation was not available. These evaluations were also used to assess whether concomitant procedures, including baffle and/or conduit revisions, were necessary.

Study Protocol
Complications related to the procedure, survival, and operation benefit in terms of TV competence, RV function, and NYHA class were evaluated. First, the primary diagnosis (ccTGA or acTGA), previous operations, and preoperative TV competence, RV function, and NYHA class were noted. Second, the type of procedure (TVP or TVR) and any concomitant procedures were documented. Third, we observed postoperative complications, and finally, to assess operation benefit, we analyzed postoperative changes in RV function, tricuspid valve competence, and functional class during follow-up.

Complications and Survival
Postoperative complications were categorized into categories as arrhythmia, low cardiac output (defined as persistently low arterial pressures in the postoperative period that did not respond to plasma volume expansion), renal insufficiency, infection, and rethoracotomy. Maximum survival was defined as the period between the first postoperative day until the last available follow-up date or the occurrence of the composite end point of death or recurrent severe tricuspid regurgitation. Differences in survival were compared between the TVP and TVR procedures.

Operation Benefit
Echocardiography was used for the assessment of RV and TV function. For the purpose of the present study, echocardiograms, made preoperatively and during periodic follow-up after surgery, were reviewed for RV and TV function. Echocardiographic evaluations were performed 3 weeks, 3 months, and 10 months after surgery. Tricuspid regurgitation was graded on a scale of 1 to 4 according to its severity. RV function was analyzed semiquantitatively and classified as either normal (1), mildly depressed (2), depressed (3), or severely depressed (4).17,18 Furthermore, patient functional status was assessed preoperatively and postoperatively according to NYHA class.

Operative Technique
All patients were operated on by the same surgeon through median (re)sternotomy with total cardiopulmonary bypass, mild to moderate hypothermia, and antegrade perfusion of a crystalloid cardioplegic solution. The TV was approached via either the right atrium (acTGA) or the atrial septum and left atrium (ccTGA). Initially, TVP was intended in all patients. For this type of surgery, a classic Carpentier-Edwards or the newer Edwards MC3 annuloplasty ring (Edwards Lifesciences Inc, Irvine, Calif) was used. However, preoperatively or intraoperatively, either when the quality of the TV leaflets was regarded as insufficient (eg, dysplastic tricuspid leaflets) or when significant leaflet tethering was present, it was decided to replace rather than repair the TV. Moreover, when the initial TVP did not result in reduction of regurgitation during the operation, the TV was also replaced within the same operation. For this procedure, a CarboMedics bileaflet mechanical prosthesis (Sulzer CarboMedics Inc, Austin, Tex), a St. Jude Medical mechanical prosthesis (St. Jude Medical Inc, St. Paul, Minn), or a Medtronic Mosaic porcine bioprosthesis (Medtronic Inc, Minneapolis, Minn) was used.

Statistical Analysis
SPSS (version 12.0.1; SPSS Inc, Chicago, Ill) was used for statistical analysis. Data are presented as mean±SD unless otherwise noted. Because of the small number of patients, the Friedman test and the Mann-Whitney U test were used to compare preoperative and postoperative TV function, RV function, and functional class. Kaplan-Meier curves were drawn, and the log-rank test was used to compare the difference between the TVP and TVR procedure. P values <0.05 were considered statistically significant. The authors had full access to and take full responsibility for the integrity of the data. All authors have read and agree to the manuscript as written.

Results
Sixteen patients underwent surgery, 9 with ccTGA and 7 with acTGA. Characteristics of these patients are summarized in Table 1. Preoperatively, grade 3.1±0.8 TV regurgitation was observed. Most patients exhibited mildly depressed RV function (n=12), and preoperative NYHA functional class was 2.7±0.6. Figure 1 provides an overview of patient surgery. In 14 patients, the mechanism of tricuspid regurgitation was regarded as malcoaptation of the tricuspid leaflets. This was either due to annular dilatation with intact leaflets

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<th>Table 1. Patient Characteristics</th>
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<td>Age, y</td>
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<tr>
<td>Previous operations, n</td>
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<tr>
<td>VSD closure</td>
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<td>ASD closure</td>
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<tr>
<td>Concomitant procedures, n</td>
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<tr>
<td>Conduit/baffle revision</td>
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<td>VSD closure</td>
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<tr>
<td>PA banding</td>
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<td>VSD indicates ventricular septal defect; ASD, atrial septal defect; PVR, pulmonary valve replacement; and PA, pulmonary artery.</td>
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or structural abnormalities of the TV other than Ebstein(-like) malformations (n=5). In 11 patients, TVP was initially attempted. In 3 of these 11 cases, this procedure did not result in improved TV competence, and it was decided to perform TVR in the same operation. In 4 patients, the TV leaflets were Ebstein-like or had other important malformations, and in 1 patient, the tricuspid annulus was too severely dilated to perform TVP; thus, TVR was performed immediately in these cases. In 9 patients, concomitant procedures, mainly conduit or baffle revisions (n=6), were performed.

Complications and Survival
All major postoperative complications are summarized in Table 2. One patient who underwent surgery acutely after explantation of an infected implantable cardioverter defibrillator died 109 days after surgery. This patient had a multitude of complications and eventually died of therapy-refractory sepsis. Arrhythmia was the most frequently encountered short-term complication, occurring in 8 patients. In 1 patient, recurrent ventricular fibrillation was observed postoperatively, which required implantation of an implantable cardioverter defibrillator; another 5 patients had supraventricular tachycardia, which was treated with cardioversion in 3 and with mediation in 2. One patient had sinus arrest, and 1 had AV block, both of which were treated with pacemaker insertion. Another complication that occurred frequently was low cardiac output caused by ventricular failure (n=5), which required placement of an intra-aortic balloon pump in 3 patients and inotropic support in 2. One patient underwent surgery for gastric perforation, and in another patient, a rethoracotomy for persistent bleeding was performed. Five patients had renal insufficiency that required temporary dialysis with continuous venovenous hemofiltration.

Fifteen patients were discharged in good condition after a mean hospital stay of 27±28 days. Two patients died during the period after discharge. One patient was resuscitated successfully after an episode of ventricular fibrillation 180 days after surgery but died subsequently of ventricular failure, without signs of recurrent tricuspid regurgitation. The other patient died of recurrent tricuspid regurgitation and cardiac failure 659 days after surgery, before adequate treatment could be initiated. Follow-up of patients ranged from 92 to 3101 days after operation, with a 1-year survival rate of 86.7%. Kaplan-Meier curves (Figure 2) for the composite end point of death or recurrent severe tricuspid regurgitation revealed that TVR patients exhibited better survival than patients who underwent TVP; the difference in survival, however, was not statistically significant (P=0.09).

Operation Benefit
Overall, tricuspid function and functional class improved significantly after operation, and RV function remained

Table 2. Postoperative Complications

<table>
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<tr>
<th>Complications</th>
<th>TVP</th>
<th>TVR</th>
<th>Total</th>
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<tr>
<td>None</td>
<td>4 (25)</td>
<td>1 (6)</td>
<td>5 (31)</td>
</tr>
<tr>
<td>Arrhythmia</td>
<td>3 (19)</td>
<td>5 (31)</td>
<td>8 (50)</td>
</tr>
<tr>
<td>Low cardiac output</td>
<td>2 (13)</td>
<td>3 (19)</td>
<td>5 (31)</td>
</tr>
<tr>
<td>Renal insufficiency</td>
<td>2 (13)</td>
<td>3 (19)</td>
<td>5 (31)</td>
</tr>
<tr>
<td>Infection</td>
<td>1 (6)</td>
<td>1 (6)</td>
<td>2 (13)</td>
</tr>
<tr>
<td>Rethoracotomy</td>
<td>1 (6)</td>
<td>1 (6)</td>
<td>2 (13)</td>
</tr>
<tr>
<td>Death</td>
<td>0</td>
<td>1 (6)</td>
<td>1 (6)</td>
</tr>
</tbody>
</table>

Values are n (%).
stable. In Figure 3, it can be readily seen that tricuspid competence improved significantly after TVP and, as could be expected, after TVR. After TVP, however, tricuspid regurgitation was not substantially reduced in 3 patients (2 ccTGA, 1 aTGA) on postoperative echocardiographic evaluation. Therefore, the overall improvement in tricuspid function was less prominent than with TVR. Furthermore, 3 patients developed recurrent TV regurgitation that resulted in grade 2.6±0.5 regurgitation at 10.7±1.9 months of follow-up after TVP. After TVR, on the other hand, the improvement of tricuspid competence was maintained at 10.7±1.9 months of follow-up, as could be expected. RV function, which was classified as normal (grade 1) to severely depressed (grade 4), remained unchanged postoperatively and at 10.7±1.9 months’ follow-up (Figure 3). Preoperatively, RV function was classified as 2.2±0.7, and at maximum follow-up (3.2±1.9 years), it was 2.0±0.9. Moreover, no differences in RV function were noted preoperatively or during follow-up between patients who underwent TVP and those who underwent TVR. Overall, NYHA class improved from 2.7±0.6 to 2.1±0.8 (P=0.007) 3.2±1.7 months after surgery. At 3.2±1.9 years of follow-up, functional class was 2.0±0.9. Again, no difference in NYHA class was found between TVP and TVR preoperatively or during follow-up.

Discussion

In the present study, we systematically evaluated the complications, survival, and benefit of TV surgery for substantial regurgitation in patients with a dysfunctional RV in the systemic position. Key findings were that a high incidence of postoperative complications was observed, and overall TV competence and functional class improved after surgery. Finally, the survival characteristics of TVP appeared less favorable than those of TVR.

The incidence of complications related to the procedure was high in the present study population; however, all complications were treated successfully in those patients who underwent elective surgery, and the presence of complications was not related to worse outcome. In the largest study on TV operations to date by van Son et al, comprising 40 patients with ccTGA, similar types of postoperative complications were described (arrhythmia, bleeding, and infection), and no relation between postoperative complications and adverse outcome was reported. In another report by Beauchesne et al on patients with
ccTGA, postoperative complications were again not related to postoperative outcome. Therefore, provided that surgical treatment of TV regurgitation in patients with an RV in the systemic position is performed exclusively in specialized centers, the complication rate should generally not be a reason to refrain from these operations.

Three patients, all with previous Mustard corrections, died in the present study population, which resulted in a 1-year survival rate of 86.7%. In 1 of these patients, the cause of death was TV incompetence and associated systemic ventricular failure after TVP. In the other 2 patients, TV incompetence and systemic ventricular dysfunction were present but not indubitably related to the cause of death (sepsis and ventricular arrhythmia). In the previously mentioned study by van Son et al, a survival rate of 78.0% at 5 years and 60.7% at 10 years was reported. In other, smaller studies on patients with ccTGA in which survival characteristics of TV surgery were reported as part of a larger study, 1-year survival rates ranged from 63% to 100%. To the best of our knowledge, no studies are available that report data solely on TV operations in patients with acTGA. Incidentally, survival characteristics are reported as unfavorable in small groups of patients (n=3), with 1-year survival rates of 25% to 33%, which is much lower than the 1-year survival rate in the present study group of acTGA patients (71.4%).

In general, TVP was intended in all patients; however, in patients with structural leaflet malformations or severe leaflet tethering, which was noted either preoperatively or during surgery, it was decided to perform TVR. In these patients, the risk for recurrent tricuspid regurgitation after TVP was regarded as higher than for patients with intact tricuspid leaflets and without signs of tethering. In the present study population, tricuspid competence demonstrated a significant improvement after surgery; however, the results of TVP were less promising than the results of TVR. After TVP, the improvement was less prominent than after TVR, and the rate of recurrence of regurgitation was relatively high after TVP (37%). Therefore, when the survival curves of the TVP procedure are compared with the TVR procedure, there appears to be a disadvantage for TVP, although it is not statistically significant.

In the present study population, RV function remained stable during follow-up, and no differences in RV function were noted between patients who underwent TVP and those who underwent TVR. In most patients, the preoperative degree of RV dysfunction was generally mild (n=12), and it can be questioned whether an important improvement in RV function is expected after TV surgery. The sparse results of earlier studies suggest that TV surgery in the early stages of RV dysfunction is superior to TV surgery in more advanced stages of RV dysfunction. Therefore, the strategy in most patients was to consider TV surgery on the basis of functional impairment, even if only mild RV dysfunction was present. Although no improvement in RV function was observed on echocardiographic evaluation, functional class did improve in the present study population. A mechanism that may underlie this observation is that improved competence of the TV leads to more effective output of the systemic ventricle, whereas the ejection fraction might even decline. Before surgery, stroke volume is partially regurgitated back into the systemic atrium, whereas after surgery, stroke volume is devoted completely to cardiac output, which leads to overall better function. Second, visual assessment of RV function, although generally accepted, may not reveal subtle but important changes in RV function. Other imaging modalities, such as cardiac MRI, may demonstrate improved rather than stable RV function in the long term.

The combination of tricuspid regurgitation and progressive RV dysfunction is a frequent finding in patients with ccTGA or acTGA. It is associated with a rapid decline in functional class and decreased survival, and surgical treatment is usually warranted. Selection of the optimal surgical strategy, specifically in “older” patients (third to fourth decade of life), is a complicated issue. It could be postulated that either relocation of the (morphologically) left ventricle in the systemic position or cardiac transplantation is the surgical treatment of choice in these cases; however, results after relocation of the left ventricle into the systemic position, in terms of survival and benefit, are discouraging in the adult population with transposition of the great arteries. The option of cardiac transplantation is probably the superior option from a functional point of view. Unfortunately, donor organs are scarce, and cardiac transplantation can only be applied in a limited number of patients. Hence, cardiac transplantation as a treatment option in the approach to patients with transposition of the great arteries who have tricuspid regurgitation and a dysfunctional RV remains difficult. The results from the present study population, which was relatively old (34.2±14.5 years in the ccTGA group and 35.0±5.9 years in the acTGA group), demonstrate that TVR and TVP are associated with low mortality and improvement of functional class. Therefore, these procedures could be regarded as a first option in patients with a systemic RV and tricuspid regurgitation. However, survival characteristics tend to be worse after TVP, mainly because of a high recurrence rate of tricuspid regurgitation. Tricuspid leaflet tethering may be the mechanism behind the recurrence of tricuspid regurgitation after TVP.

**Study Limitations**

In the present study, semiquantitative analysis of tricuspid and RV function with echocardiography was used. Cardiac MRI provides a more accurate modality for estimation of RV function; however, an important number of patients have pacemakers and could not undergo MRI. For consistency, we therefore chose echocardiography for comparison of data during follow-up. As another limitation, which is present in most studies of patients with congenital heart disease, the study population was small. Therefore, the results of the present study must be confirmed prospectively in a larger patient population.

**Conclusions**

Mortality is rather low after tricuspid surgery in patients with a systemic RV. Although the incidence of postoperative complications is high, this should not be a reason to refrain from performing these operations. Short-term results demonstrate that TVR in patients with a dysfunctional RV is associated with an improvement of TV function, stable RV function, and improvement of functional class. Valvuloplasty, which is associated with a high recurrence rate, is not advisable in this group of patients, and TVR is therefore the preferred procedure.
Disclosures
None.

References

CLINICAL PERSPECTIVE
Tricuspid regurgitation is a frequent finding in patients with atrially corrected or congenitally corrected transposition of the great arteries and, consequently, a systemic right ventricle. When important tricuspid regurgitation occurs, it is usually associated with rapid deterioration of right ventricular function and functional status. Selection of adequate therapy is a challenging issue, specifically in those patients beyond the second decade of life. Several surgical approaches are possible in cases of important tricuspid regurgitation, and in the present study, tricuspid valvuloplasty or tricuspid valve replacement was investigated in a cohort of 16 adult patients with symptomatic important tricuspid regurgitation and relatively mild right ventricular dysfunction. Mortality was rather low, but the rate of complications was high. All complications were manageable, which makes this a safe operation. The results of this study indicate that tricuspid valve replacement may be superior to tricuspid valvuloplasty. Although the short-term results were equal in terms of right ventricular function and improvement in functional class, a high incidence of recurrent moderate tricuspid regurgitation was observed within 1 year after valvuloplasty. It is concluded that tricuspid valve surgery is safe in patients with transposition of the great arteries and a systemic right ventricle, provided it is performed in specialized centers. Furthermore, tricuspid valve replacement is probably superior to tricuspid valvuloplasty in adult patients with important tricuspid regurgitation and a systemic right ventricle.
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Circulation. 2009;119:1467-1472; originally published online March 9, 2009; doi: 10.1161/CIRCULATIONAHA.108.805135
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

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