

# The quadricuspid truncal valve: Surgical management and outcomes



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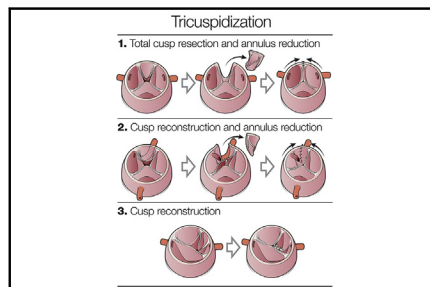
## ABSTRACT

**Objective:** To determine the outcomes of patients with a quadricuspid truncal valve (TV) and durability of TV repair.

**Method:** We reviewed 56 patients with truncus arteriosus and a quadricuspid TV who underwent complete repair between 1979 and 2018.

**Results:** TV insufficiency was present in 39 patients (mild, n = 22; moderate, n = 14; and severe, n = 3). Fourteen patients had concomitant TV surgery. Early mortality in patients who had concomitant TV surgery was 14% (2 out of 14 patients) and overall survival was 77.1% ± 11.7% at 15 years. Freedom from TV reoperation was 30.3% ± 14.6% at 15 years. Early mortality in patients who did not undergo concomitant TV surgery was 9.5% (4 out of 42 patients) and overall survival was 74.9% ± 6.9% at 15 years. Progression of TV insufficiency requiring TV surgery occurred in 16.7% (7 out of 42 patients). Freedom from TV reoperation was 77.1% ± 7.8% at 15 years. The most common method of repair was tricuspidization of the TV. Freedom from TV reoperation was 64.3% ± 21.0% at 10 years after tricuspidization and 0% at 6 years after other types of TV surgery. Overall follow-up was 97.6% (41 out of 42 patients) complete for survivors with median follow-up of 16.6 years. At last follow-up there was no TV insufficiency in 16 patients, mild insufficiency in 24 patients, and moderate insufficiency in 1 patient.

**Conclusions:** More than one-third of patients with a quadricuspid TV require TV surgery. Tricuspidization of the quadricuspid TV appears to be a durable repair option with good long-term outcomes. (J Thorac Cardiovasc Surg 2021;161:368-75)



Techniques of tricuspidization of the truncal valve.

## CENTRAL MESSAGE

More than one-third of children with a quadricuspid truncal valve will require truncal valve surgery. Tricuspidization is a durable repair option in young children.

## PERSPECTIVE

Tricuspidization of the quadricuspid truncal valve has good long-term outcomes. In young children, tricuspidization of the quadricuspid valve is a durable repair option and may help to avoid the need for truncal valve replacement.

See Commentaries on pages 376 and 377.

We recently reviewed the long-term outcomes of surgical repair of truncus arteriosus and the impact of truncal valve (TV) insufficiency.<sup>1-3</sup> Although there was no apparent association between TV surgery and mortality, concomitant TV surgery is a significant challenge during truncus arteriosus repair. We previously reported that most patients who have required TV surgery had a quadricuspid TV.<sup>1-3</sup> Furthermore, most patients with moderate or severe TV insufficiency had a quadricuspid TV.<sup>1-3</sup> In contrast, mild insufficiency of the quadricuspid

TV is rarely progressive.<sup>1-3</sup> Herein, we describe the outcomes of surgery in children with a quadricuspid TV.

## PATIENTS AND METHODS

The Royal Children’s Hospital Human and Research Ethics Committee approved the current study. All consecutive patients (n = 56) with truncus arteriosus and a quadricuspid TV who underwent operation at The Royal Children’s Hospital between 1979 and 2018 were reviewed. Data were obtained by review of medical records from initial presentation until final cardiology follow-up. Patient characteristics are summarized in Table 1.

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### Abbreviations and Acronyms

TTE = transthoracic echocardiogram  
TV = truncal valve

### Definitions

Early mortality was defined as deaths occurring within 30 days of surgery or before hospital discharge. All other deaths were considered late. Subsequent TV surgery was defined as the first TV surgery done after initial truncus arteriosus repair. The presence of TV insufficiency was evaluated by preoperative transthoracic echocardiography (TTE) and color Doppler imaging using standard echocardiographic criteria,<sup>4-6</sup> including M-mode assessment of left ventricular function, Doppler assessment of pressure half-time (mild [ $>500$  ms], moderate [500-200 ms], and severe [ $<200$  ms]), diastolic flow reversal in the descending aorta (mild [brief, early diastolic reversal], moderate [intermediate], and severe [holodiastolic flow reversal]), color flow regurgitant jet size (mild [small central jet  $<25\%$  left ventricular outflow tract], moderate [intermediate jet greater than mild but no signs of severe], and severe [large central jet  $>65\%$  left ventricular outflow tract]), and measurements of the truncal regurgitant jet width to annulus ratio (mild [ $<25$ ], moderate [25-64], and severe [ $\geq 65$ ]). TV insufficiency was graded qualitatively from none/trivial to severe. All patients underwent preoperative TTE. Generally, TV surgery was performed at truncus arteriosus repair when preoperative TV insufficiency was graded as moderate or severe. Lesser degrees of TV insufficiency were repaired only when intraoperative decision making dictated that TV repair was necessary.

### Data Analysis

Data were analyzed using Stata version 14 (StataCorp LP, College Station, Tex). Descriptive statistics for continuous data are expressed as median (range). Mann-Whitney *U*-test and  $\chi^2$  test were used where appropriate. Categorical data were summarized as frequencies and percentages. Kaplan-Meier actuarial survival ( $\pm$  standard error) was used to analyze and plot time-related end points. Log-rank test was used to compare survivor functions.

## RESULTS

Median age at truncus arteriosus repair was 53 days (range, 1 day-3.1 years) and median weight was 3.6 kg (range, 1.7-12 kg). Median age decreased over time. Median age was 69.5 days between 1979 and 1988 ( $n = 14$ ); 70 days between 1989 and 1998 ( $n = 23$ ); 32 days between 1999 and 2008 ( $n = 9$ ); and 8 days between 2009 and 2018 ( $n = 10$ ). Concomitant anomalies were present in 27 patients (Table 1), including associated aortic arch obstruction in 7 patients. Preoperative TTE determined TV insufficiency in 39 patients. There were 17 patients with no TV insufficiency, 22 with mild insufficiency, 14 with moderate insufficiency, and 3 with severe insufficiency. Patient outcomes are summarized in Figure 1. Overall early mortality in patients with a quadricuspid TV was 10.7% (6 out of 56 patients) and there were 8 late deaths indicating an overall survival of  $75.4\% \pm 6.0\%$  (95% confidence interval [CI], 61.2%-85.1%) at 15 years. Overall survival in patients with mild or less TV insufficiency was  $75.8\% \pm 7.1\%$  (95% CI, 58.5%-86.6%) at 15 years compared with  $76.0\% \pm 10.5\%$  (95% CI, 48.0%-

90.3%) at 15 years ( $P = .93$ ) (Figure 2, A). Overall follow-up was 97.6% (41 out of 42 patients) complete for survivors with median follow-up of 16.6 years (range, 1-38 years). At final follow-up, all patients were found to be in New York Heart Association functional class I or II.

### Concomitant TV Surgery

Concomitant TV surgery was undertaken in 14 patients (Figure 1). All but 1 patient had at least moderate TV insufficiency on preoperative TTE. One patient with mild TV insufficiency on initial preoperative imaging was deemed to have moderate insufficiency on follow-up imaging. Early mortality in patients who underwent concomitant TV surgery was 14% (2 out of 14 patients), both deaths occurred in neonates. The first early death was due to cardiac arrest, and the second early death was due to a persistently low cardiac output state in a patient who could not be weaned off extracorporeal membrane oxygenation. There was 1 late death due to acute-on-chronic respiratory failure secondary to bronchiolitis. Overall survival in patients who required concomitant TV surgery was  $77.1\% \pm 11.7\%$  (95% CI, 44.2%-92.1%) at 15 years (Figure 2, B).

Surgery was performed via median sternotomy with the use of cardiopulmonary bypass in all patients. There were 3 surgeons (C.P.B., Y.D., I.E.K.) who performed TV surgery for quadricuspid valve during the study period. The truncus arteriosus was transected and the TV directly inspected. Characteristics and outcomes of patients who underwent concomitant TV repair are summarized in Table 2. As shown in Figure 3, A, tricuspidization was achieved by total cusp resection and annulus reduction, cusp reconstruction and annulus reduction, or cusp reconstruction. Often, the smallest or most dysplastic leaflet was identified and resected, along with the corresponding part of the arterial wall. The truncal wall was reconstructed by direct suture and this suture line continued to close the defect between the 2 adjacent valve leaflets (Figure 3, A-1). This method was used in 4 patients concomitantly at truncus repair. In 2 patients, the left coronary artery was arising from the sinus of the redundant valve leaflet, and the leaflet was partially resected and reconstructed in a similar fashion (Figure 3, A-2). In the remaining patient, the raphe between 2 rudimentary cusps was resected and both cusps were sutured together to form a single, good-quality cusp (Figure 3, A-3). Competence of the TV was achieved by nontricuspidization in 7 patients (Figure 3, B).

Eight patients underwent 11 TV reoperations (Figure 1). Median time to TV reoperation was 248 days (2.4 years; range, 12 hours-6.1 years). Four reoperations occurred within the first year following initial TV surgery. Overall freedom from TV reoperation was  $30.3\% \pm 14.6\%$  (95% CI, 7.2%-58.2%) at 15 years. Freedom from reoperation in the 7 patients who underwent concomitant tricuspidization of their TV was  $64.3\% \pm 21.0\%$  (95% CI, 15.2%-90.2%) at 10 years compared with 0% at 6 years for any

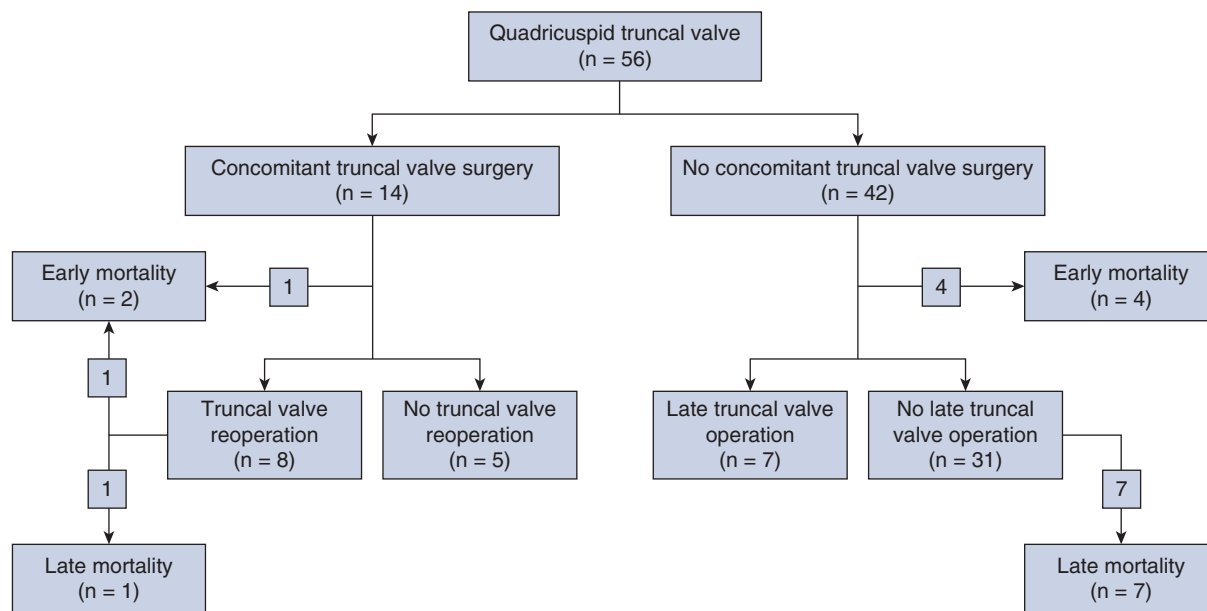
**TABLE 1. Patient characteristics**

Characteristic	Overall (n = 56)	Concomitant TV surgery (n = 14)	No concomitant TV surgery (n = 42)
Age (d)	53 (1-1123)	12.5 (1-1123)	66 (4-682)
Weight (kg)	3.6 (1.7-12)	3.1 (1.7-12.0)	3.7 (2.0-8.2)
Neonate at time of repair	35.7 (20/56)	57.1 (8/14)	28.6 (12/42)
CPB time (min)	126 (60-241)	206 (87-241)	112 (60-227)
Crossclamp time (min)	63 (40-263)	120 (80-263)	59 (40-158)
<b>Degree of TV insufficiency</b>			
None	30.4 (17/56)	0	40.5 (17/42)
Mild	39.3 (22/56)	7.1 (1/14)	50.0 (21/42)
Moderate	25.0 (14/56)	71.4 (10/14)	9.5 (4/42)
Severe	5.4 (3/56)	21.4 (3/14)	0
<b>Degree of TV stenosis</b>			
None	82.1 (46/56)	64.3 (9/14)	88.1 (37/42)
Mild	12.5 (7/56)	21.4 (3/14)	9.5 (4/42)
Moderate	5.4 (3/56)	14.3 (2/14)	2.4 (1/42)
Severe TV stenosis	0	0	0
<b>Concomitant anomalies</b>			
Interrupted aortic arch	10.7 (6/56)	7.1 (1/14)	11.9 (5/42)
Coronary artery anomaly	7.1 (4/56)	14.3 (2/14)	4.8 (2/42)
MAPCA	7.1 (4/56)	14.3 (2/14)	4.8 (2/42)
Coarctation	1.8 (1/56)	7.1 (1/14)	0
DiGeorge syndrome	10.7 (6/56)	21.4 (3/14)	7.1 (3/42)
Scimitar syndrome	1.8 (1/56)	7.1 (1/14)	0

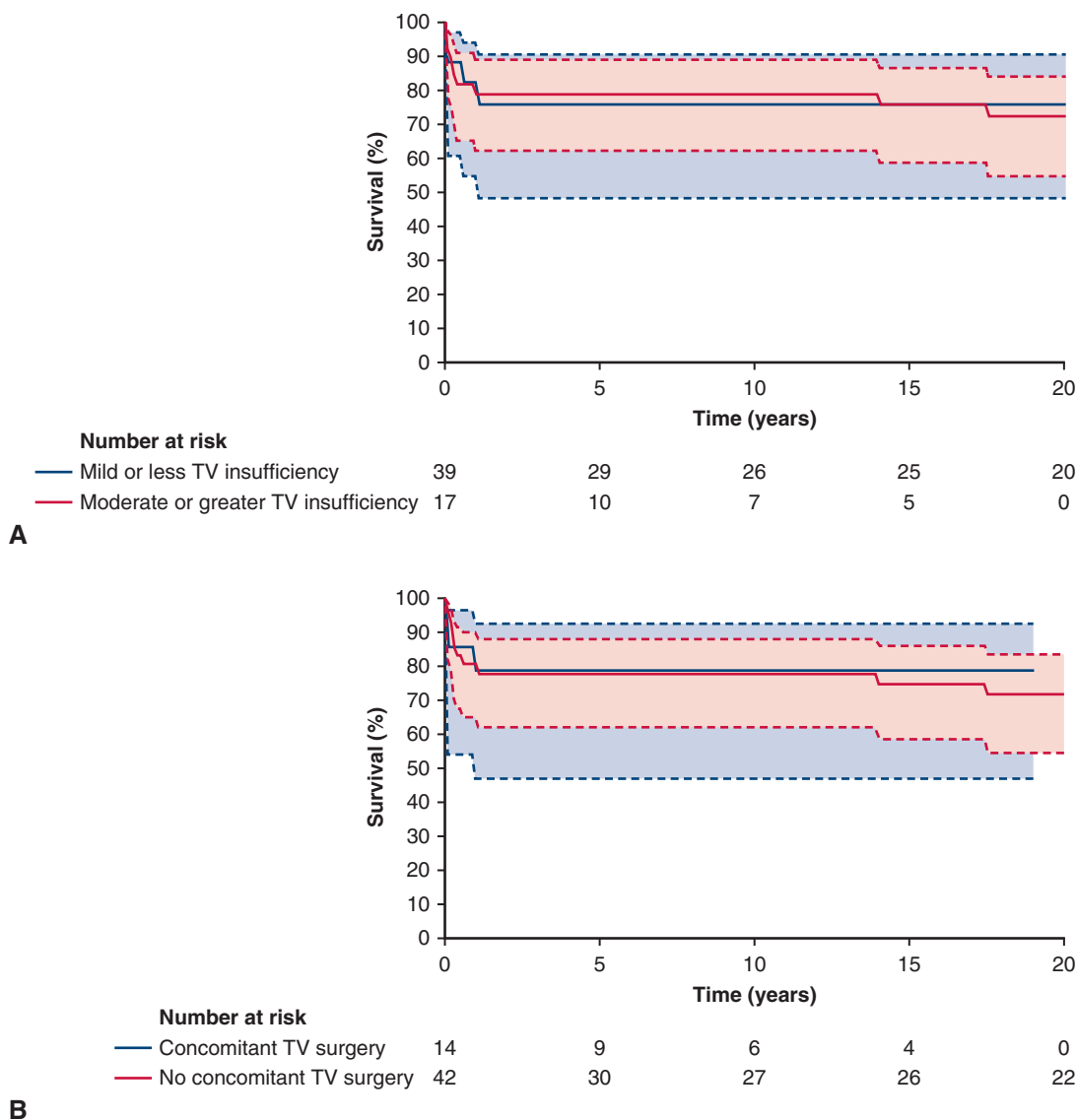
Values are presented as % (n/N) or median (range). TV, Truncal valve; CPB, cardiopulmonary bypass; MAPCA, major aortopulmonary collateral artery.

other repair method ( $P = .04$ ). Two patients who underwent tricuspidization of their TV required TV reoperation at 2 months and 6 years. Of the remaining 6 patients who required TV reoperation, 2 underwent tricuspidization of their previously repaired TV. The first patient initially underwent subcommissural annuloplasty and plication of the sinotubular junction, then underwent tricuspidization at

2.8 years, followed by replacement with a 24-mm aortic homograft 8 years later due to endocarditis with root abscess. The patient is currently well at 13 years. The second patient initially underwent resection of accessory tissue from the valve cusps, then underwent tricuspidization at 2.6 years. The patient is currently well at 2.7 years. After median follow-up time of 16.1 years (10.2 years; range, 1.2-



**FIGURE 1.** Diagram showing the timing and distribution of truncal valve surgery, mortality, and reoperation.



**FIGURE 2.** Survival in children with a quadricuspid truncal valve (TV). A, Mild or less TV insufficiency versus moderate or greater TV insufficiency. B, Concomitant TV surgery versus no concomitant TV surgery.

19 years), there was TV insufficiency in 5 patients, and mild insufficiency in 6 patients after concomitant TV surgery.

**No Concomitant TV Surgery**

There were 42 patients who did not undergo concomitant TV surgery (Figure 1). Most patients who did not require concomitant TV surgery had mild (n = 21) or no (n = 17) TV insufficiency (90.5%; 38 out of 42). Early mortality in patients who did not require concomitant TV surgery was 9.5% (4 out of 42). Two early deaths were due to acute-on-chronic respiratory failure secondary to severe tracheobronchomalacia; 1 early death was due to cardiac arrest; and 1 early death was due to sepsis. There were 7 late deaths, 5 of which were within the first year following truncus repair and overall survival

of 74.9% ± 6.9% (95% CI, 58.3%-85.7%) at 15 years (Figure 2, B).

Seven out of 42 patients (16.7%) required subsequent TV surgery (Figure 1). Median time to subsequent TV surgery was 5.2 years (range, 7 days-13.3 years). Of these patients, 1 initially had no TV insufficiency, 4 had mild insufficiency, and 2 had moderate insufficiency. Freedom from subsequent TV surgery was 77.1% ± 7.8% (95% CI, 57.3%-88.5%) at 15 years. Subsequent TV operations are summarized in Table 3. Five patients underwent TV replacement. Two patients underwent TV repair with tricuspization of their TV at 7 days and 5.2 years. Of these 2 patients, the first required TV replacement at 15.6 years and is currently well at 21.7 years, whereas the second is free from TV reoperation and well at 15.3 years. At final

TABLE 2. Concomitant truncal valve (TV) surgery

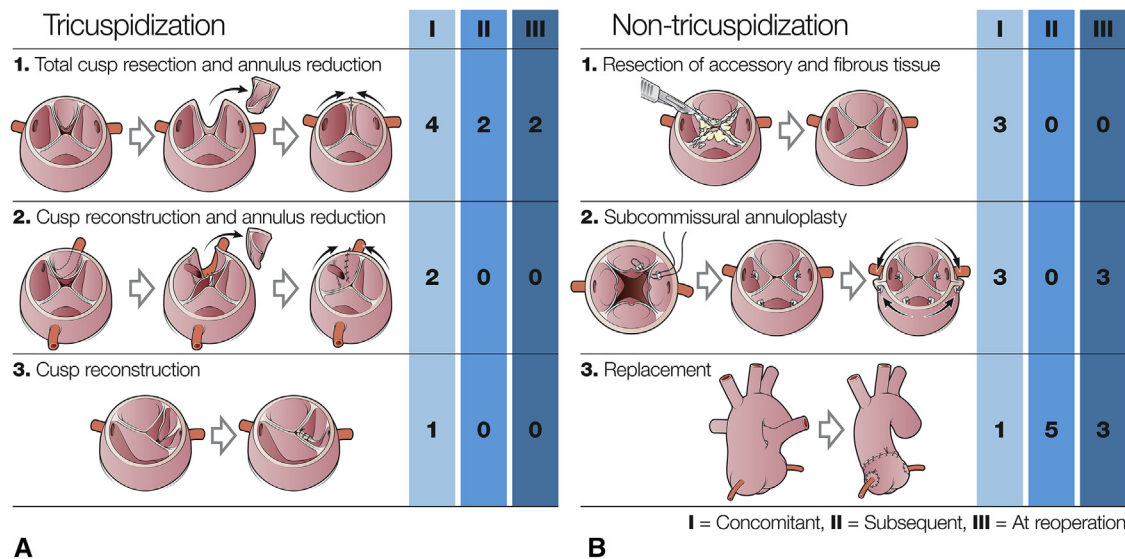
Pt	TV insufficiency	Additional diagnoses	Age (d)	Weight (kg)	TV surgery type	Other surgery	Time to TV reoperation	TV reoperation	Follow-up time
1	Moderate	None	1	1.7	Commissurotomy and resection of accessory tissue	None	–	None	Death (early)
2	Mild	None	80	4.8	Tricuspidization (cusp reconstruction)	None	–	None	19.0 y
3	Moderate	Coronary anomaly	33	2.9	Tricuspidization (cusp reconstruction and annulus reduction)	None	–	None	19.0 y
4	Moderate	None	164	5.3	Tricuspidization (cusp resection and annulus reduction)	None	–	None	16.4 y
5	Moderate	DiGeorge syndrome	41	3.1	Subcommissural annuloplasty and STJ plication	None	2.8 y	Tricuspidization (cusp resection and annulus reduction)*	13.0 y
6	Severe	DiGeorge syndrome	10	2.8	Tricuspidization (cusp resection and annulus reduction)	None	0.2 y	Konno procedure and replacement with 19-mm Carbomedics aortic valve	Death (late)
7	Severe	Scimitar syndrome; MAPCA	7	3.0	Subcommissural annuloplasty	MAPCA ligation; right pneumonectomy	1 d	Replaced with 14-mm aortic homograft	Death (early)
8	Moderate	None	2	3.5	Replacement with 10-mm aortic homograft	None	0.6 y	Replaced with 17-mm St Jude aortic valve	11.7 y
9	Moderate	None	1123	12.0	Subcommissural annuloplasty	None	6.1 y	Subcommissural annuloplasty and STJ plication	9.8 y
10	Moderate	CoA	1	3.1	Commissurotomy and resection of accessory tissue	CoA repair	0.7 y	Commissuroplasty†	8.6 y
11	Moderate	DiGeorge syndrome	79	3.8	Tricuspidization (cusp resection and annulus reduction)	None	6.0 y	Commissuroplasty	8.2 y
12	Moderate	ASD	15	3.0	Resection of nodules from valve cusps	None	2.6 y	Tricuspidization (cusp resection and annulus reduction)	2.7 y
13	Moderate	Coronary anomaly	4	3.0	Tricuspidization (cusp reconstruction and annulus reduction)	Coronary unroofing	–	None	1.0 y
14	Severe	IAA	3	3.2	Tricuspidization (cusp resection and annulus reduction)	AA reconstruction	–	None	1.0 y

St Jude: St Jude Medical, St Paul, Minn; Carbomedics: LivaNova, London, UK; On-X: On-X Life Technologies, Austin, Tex. *Pt*, Patient; *TV*, truncal valve; *STJ*, sinotubular junction; *MAPCA*, major aortopulmonary collateral artery; *CoA*, coarctation; *ASD*, atrial septal defect; *IAA*, interrupted aortic arch; *AA*, aortic arch. \*Replaced 8 years later with a 24-mm aortic homograft due to endocarditis with root abscess. †Replaced 4.2 years later with a 19-mm On-X aortic valve then again 1 year later with a 21-mm On-X aortic valve due to endocarditis.

follow-up, in patients who did not have concomitant TV surgery, there was no TV insufficiency in 11 patients, mild insufficiency in 17 patients, and moderate insufficiency in 1 patient. Furthermore, the TV status was unchanged in patients who had late deaths.

**Tricuspidization**

There was a total of 11 patients who underwent tricuspidization throughout the study period. The first concomitant tricuspidization was performed in 2000 (the second patient in this cohort to undergo concomitant



**FIGURE 3.** Truncal valve (TV) surgical techniques. A, Tricuspidization: 1 = cusp resection and annulus reduction, 2 = cusp reconstruction and annulus reduction, and 3 = cusp reconstruction. B, Nontricuspidization: 1 = resection of accessory and fibrous tissue, 2 = subcommissural annuloplasty, 3 = replacement. I, Concomitant TV surgery; II, subsequent TV surgery; III, procedure performed at reoperation.

TV surgery). There was no association between tricuspidization and era or surgery. Freedom from TV reoperation following tricuspidization was 64.9% ± 16.7% (95% CI, 24.9%-87.4%) at 10 and 15 years (Figure 4, A and B). In comparison, freedom from TV reoperation following any other repair method was 0% at 6 years (Figure 4, A). When including patients younger than age 6 years who underwent any other TV surgery, including replacement, freedom from TV reoperation was 12.7% ± 11.9% (95% CI, 0.7%-42.7%) at 15 years (P = .05) (Figure 4, B).

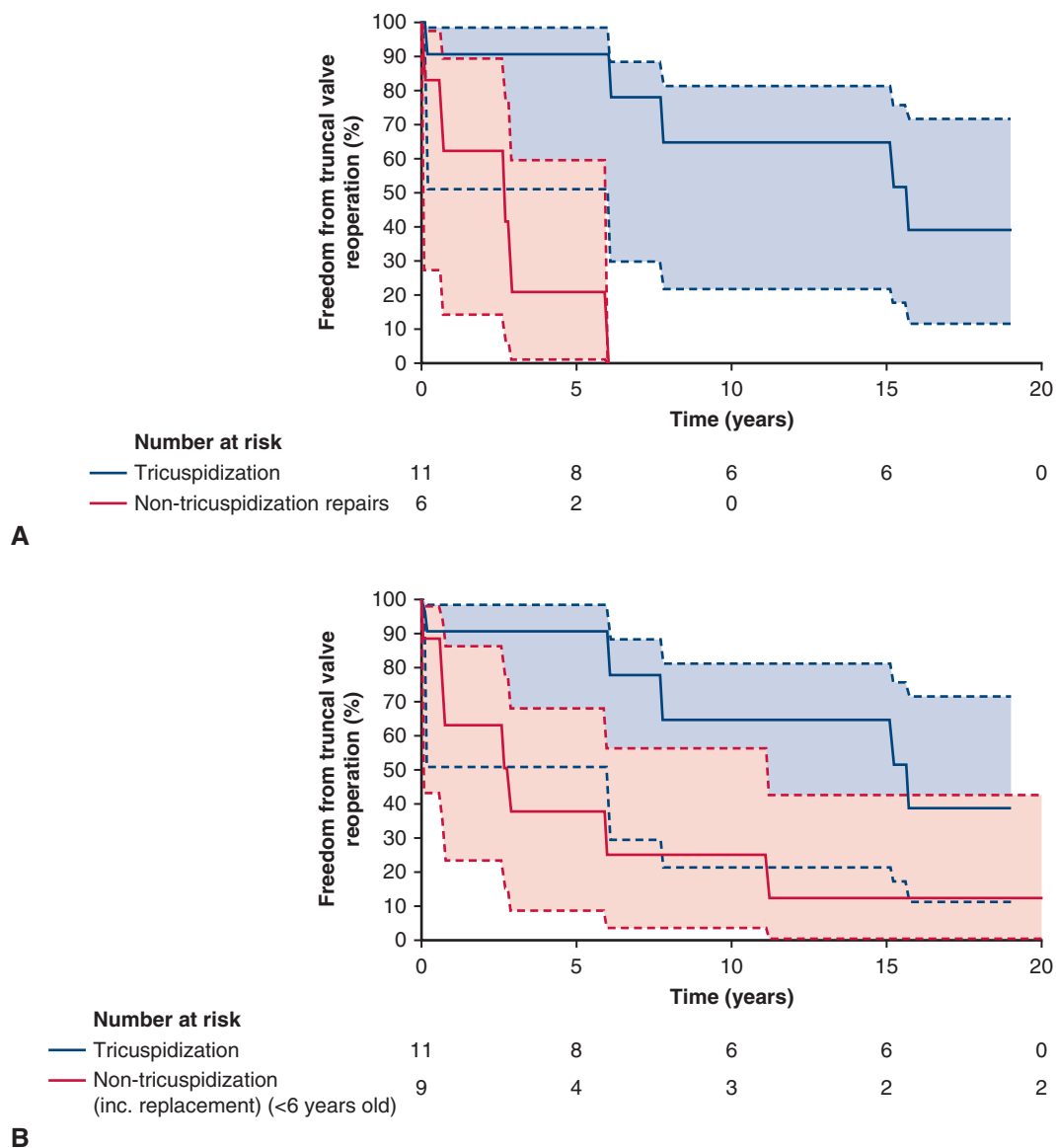
**Neonates**

Twenty neonates were included among the study population (35.7%; 20 out of 56). Overall early mortality in neonates was 15% and overall survival was 75.0% ± 9.7% (95% CI, 50.0%-88.8%) at 15 years. Of the neonates, 8 underwent concomitant TV surgery (40%) with an early mortality of 25% and overall survival of 62.5% ± 17.1% (95% CI, 22.9%-86.1%) at 10 years. The outcomes of concomitant TV surgery in neonates can be seen in Table 2. Three neonates underwent tricuspidization. Of the remaining 12 neonates, early mortality was 8%

**TABLE 3. Subsequent truncal valve (TV) surgery**

Pt	TV insufficiency	Additional diagnoses	Age (d)	Weight (kg)	Late TV surgery type	Late TV surgery time	Time to TV reoperation	TV reoperation	Follow-up time
1	Mild	None	68	3.9	Replaced with 25-mm St Jude aortic valve	3.7 y	–	None	38.1 y
2	Mild	IAA	12	2.9	Replaced with 25-mm St Jude aortic valve	7.7 y	–	None	28.3 y
3	Moderate	DiGeorge syndrome	13	3.2	Replaced with 16-mm Carbomedics aortic valve	18 d	11.2 y	Replaced with 21-mm On-X aortic valve	25.1 y
4	Moderate	None	12	2.6	Replaced with 25-mm St Jude aortic valve	13.3 y	–	None	22.8 y
5	Mild	None	217	4.7	Tricuspidization (cusp resection and annulus reduction)	7 d	15.6 y	Replaced with 23-mm On-X aortic valve	21.7 y
6	None	None	32	4.0	Tricuspidization (cusp resection and annulus reduction)	5.2 y	–	None	15.3 y
7	Mild	None	17	3.3	Replaced with 21-mm On-X aortic valve	6.5 y	–	None	8.6 y

St Jude: St Jude Medical, St Paul, Minn; Carbomedics: LivaNova, London, UK; On-X: On-X Life Technologies, Austin, Tex. Pt, Patient; TV, truncal valve; IAA, interrupted aortic arch.



**FIGURE 4.** Freedom from truncal valve (TV) reoperation. A, Tricuspidization (blue) compared with other types of repair by nontricuspidization techniques (red) performed at any stage. B, Tricuspidization (blue) compared with other types of nontricuspidization, including TV replacement, in children younger than age 6 years (red).

(1 out of 12) and overall survival was  $83.3\% \pm 10.8\%$  (95% CI, 48.2%-95.6%) at 10 years. Four neonates who did not have concomitant TV surgery required subsequent TV surgery (Table 3). Of these patients, 2 initially had mild insufficiency, and 2 had moderate insufficiency.

**DISCUSSION**

TV insufficiency is among the most important factors influencing outcomes of patients with truncus arteriosus.<sup>1,2,7-10</sup> We previously reported no apparent association between the degree of TV insufficiency or concomitant TV surgery with mortality.<sup>2,3</sup> However, our previous analysis<sup>3</sup> was limited by a small number of deaths. Despite the apparent lack of association with mortality,

most surgeons would agree that severe TV insufficiency, particularly during the neonatal period, increases mortality.

The current study demonstrated that moderate or greater truncal insufficiency in the setting of quadricuspid morphology could be dealt with effectively by valve repair or replacement, so that overall survival was not different from those children with mild or less TV insufficiency (Figure 2, A). Additionally, concomitant TV surgery was effective, so that children who required concomitant TV surgery had similar survival to those who did not require concomitant TV surgery (Figure 2, B).

Previous studies have demonstrated that the most significant risk factors for death are moderate or greater TV insufficiency and an associated interrupted aortic arch.<sup>7,11,12</sup>

There were 6 patients in our cohort with an interrupted aortic arch, 5 of whom had mild TV insufficiency. There was 1 patient with an interrupted aortic arch and severe TV insufficiency who underwent concomitant TV and aortic arch repair. This patient is currently well at 1 year of follow-up.

We previously reported that patients with a quadricuspid TV and moderate or greater truncal insufficiency, were most likely to require TV surgery at some stage throughout their lives.<sup>2</sup> In contrast, most patients with a quadricuspid TV and mild or less TV insufficiency are free from long-term TV surgery. However, in the current study, 16% of patients with mild or less insufficiency had progressive TV insufficiency that required TV surgery. Furthermore, there were 4 patients with moderate TV insufficiency who did not undergo concomitant TV surgery, of whom 2 have required subsequent TV surgery, 1 has died, and 1 was lost to follow-up after 1 year.

We have used several techniques to concomitantly repair the TV. Our preferred method of repair, that appears to give best long-term results, is tricuspidization of the quadricuspid valve with reduction of the annulus. This can be achieved by either resection of a leaflet and annulus reduction, which was first described by Imamura and colleagues in 1999,<sup>13</sup> cusp reconstruction and annulus reduction, or cusp reconstruction. We have used this technique concomitantly at truncus arteriosus repair in 7 patients; subsequently in an additional 2 patients who did not have initial TV surgery; and 2 patients who required reoperation following nontricuspidization. There were only 3 reoperations in 11 patients who underwent tricuspidization, compared with 4 reoperations in 6 patients who underwent repair by nontricuspidization. Freedom from reoperation in patients who underwent concomitant tricuspidization was 64% at 10 years compared with 0% at 6 years in patients who underwent concomitant TV repair by nontricuspidization. Thus, tricuspidization appeared to provide superior results compared with other techniques of repair (Figure 4, A). Furthermore, tricuspidization provided better long-term outcomes even if the nontricuspidization group included younger children (younger than age 6 years), in whom TV replacement was performed (Figure 4, B). Clearly, in older children, TV replacement can be performed with an adult-sized mechanical prosthesis that may potentially last for life. In fact, 4 patients, aged 4 to 13 years, had TV replacement with an adult-sized mechanical prosthesis that has so far lasted without reoperation from 8.6 to 38 years (Table 3). However, such replacement with a large prosthesis may not be feasible in younger children. Besides, a mechanical prosthesis requires lifelong anticoagulation therapy. Thus, tricuspidization of the quadricuspid TV is desirable whenever possible. Although the current study is somewhat limited due to a small number of patients, it appears that

tricuspidization of the TV is the most durable repair option in patients with a quadricuspid TV.

### Limitations

This study is subject to the usual limitations of a retrospective study. Continuous echocardiographic data was not available, thus accurate timing for the progression of TV insufficiency was unable to be determined. Due to the overall limited sample size, some variables contained a relatively small number of patients, thus limiting statistical analysis.

### CONCLUSIONS

More than one-third of patients with a quadricuspid TV require TV surgery. Tricuspidization of the quadricuspid TV appears to be a durable repair option with good long-term outcomes.

### Conflict of Interest Statement

Dr d'Udekem has received personal fees from Actelion and Berlin Heart. All other authors have nothing to disclose with regard to commercial support.

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