

Mechanical tricuspid valve replacement in hypoplastic left heart syndrome: An institutional experience



Mehar Hoda, MD,^a Robert Douglas Benjamin Jaquiss, MD,^b Lorraine James, MD,^c and Poonam Punjwani Thankavel, MD^d

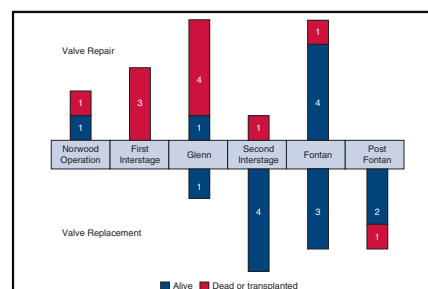
ABSTRACT

Objective: Atrioventricular valvar regurgitation in patients with single ventricles is associated with worse outcomes. Valve repair or replacement has been undertaken in an attempt to reduce mortality and morbidity. Current data on valve replacement in single ventricle patients are limited and derived from heterogeneous populations. We describe our experience with repair and replacement of the tricuspid valve in children undergoing single ventricle palliation for hypoplastic left heart syndrome.

Methods: We included 27 patients with hypoplastic left heart syndrome with at least moderate tricuspid regurgitation who underwent valve intervention between 2007 and 2021 at our institution; charts were retrospectively reviewed for data.

Results: Eleven patients (73% male) underwent valve replacement (median age, 3 years). Preoperative ventricular systolic function was normal in 10 patients (91%). Median follow-up postoperatively was 4 years with no early mortality, 1 (9%) late mortality, and 1 heart transplant (9%). Morbidity consisted of complete heart block in 1 patient (9%), with no important hemorrhagic or thrombotic events. Among survivors of replacement (n = 9), ventricular function was preserved in all (n = 8) who had normal function preoperatively and improved in the remaining patient. Sixteen patients underwent valve repair (median age, 4 months) with no early mortality, 8 (50%) midterm mortalities, and 2 heart transplants (12%).

Conclusions: Tricuspid valve replacement is a feasible option in hypoplastic left heart syndrome with significant tricuspid regurgitation, with favorable outcomes in the intermediate follow-up. When undertaken in the setting of normal function, ventricular function may be preserved in up to 80% of patients. Long-term follow-up is needed. (JTCVS Open 2022;11:363-72)



Timing and outcomes for tricuspid valve surgery in patients with HLHS.

CENTRAL MESSAGE

Tricuspid valve replacement is feasible in HLHS with significant TR with favorable outcomes in the intermediate follow-up period.

PERSPECTIVE

AVVR in patients with single ventricles is associated with significant morbidity and mortality. We observed favorable outcomes with mechanical tricuspid valve replacement in patients with HLHS in the intermediate follow-up period. When undertaken in the setting of normal function, ventricular function can be preserved in up to 80% of patients.

Hypoplastic left heart syndrome (HLHS) requires a 3-staged surgical approach to achieve sustainable single ventricle

From the ^aDivision of Pediatric Cardiology, Department of Pediatrics, University of Texas Southwestern, Dallas, Tex; ^bDivision of Pediatric and Congenital Cardiothoracic Surgery, Department of Cardiovascular and Thoracic Surgery, University of Texas Southwestern, Dallas, Tex; ^cDepartment of Pediatrics, Children's Hospital of Los Angeles, Los Angeles, Calif; and ^dDivision of Congenital Heart Surgery, Department of Pediatrics, Medical City Children's Hospital Congenital Heart Center, Dallas, Tex.

Institutional Review Board Number: STU-2021-0451, approved 6/28/2021.

Received for publication March 13, 2022; revisions received May 24, 2022; accepted for publication June 14, 2022; available ahead of print July 16, 2022.

Address for reprints: Mehar Hoda, MD, Division of Pediatric Cardiology, Department of Pediatrics, University of Texas Southwestern, 5323 Harry Hines Blvd, Dallas, TX 75390 (E-mail: meharh76@gmail.com).

2666-2736

Published by Elsevier Inc. on behalf of The American Association for Thoracic Surgery. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

<https://doi.org/10.1016/j.jxjon.2022.06.015>

circulation. Competency of the tricuspid valve is of paramount importance throughout, with significant regurgitation associated with increased risk of mortality. Up to one-third of patients at varying stages of single ventricle palliation have been reported to have significant (moderate or worse) atrioventricular valve regurgitation (AVVR).¹⁻³ Patients with moderate to severe AVVR have decreased survival after stage 1 palliation when compared with patients with mild or no insufficiency.⁴⁻⁶ The same pattern has been observed in patients after cavopulmonary Glenn shunt^{7,8} and Fontan surgery.⁸⁻¹¹ This has led to much focus on surgical approaches to reduce AVVR, although the optimal intervention and timing have not been well defined.

The mechanism of tricuspid regurgitation (TR) in HLHS is multifactorial. Stamm and colleagues¹² found that 35% of all tricuspid valves in hypoplastic left hearts

Abbreviations and Acronyms

- AVVR = atrioventricular valve regurgitation
- HLHS = hypoplastic left heart syndrome
- RV = right ventricle
- TR = tricuspid regurgitation

have moderate to severe dysplasia, including thickening, nodularity, and short chordae tendinae. These baseline anatomic abnormalities may be exacerbated by chronic volume overload, dilation of the tricuspid valve annulus, ventricular dysfunction, leaflet prolapse, papillary muscle displacement, and leaflet tethering.¹³⁻¹⁵ Intervention on the AVV before development of heart failure is considered appropriate to conserve ventricular systolic function and improves outcomes in patients with HLHS,^{2,16-19} but predicting the onset of ventricular dysfunction is difficult. In patients of any age, repair of an incompetent AVV is always preferable, but this is especially true in children because of issues of growth and the particular difficulties of managing anticoagulant medications in this population. Few data are available on mechanical AVVR in patients with single ventricles in general and HLHS in particular.^{2,20-23} Although our institutional preference has been and remains in favor of repair when feasible and successful, we have increasingly been willing to consider tricuspid valve replacement in this population because of somewhat disappointing results with repair. We sought to describe our institutional experience with tricuspid valve surgery

in patients with HLHS, with particular focus on mechanical tricuspid valve replacement, over a 14-year period (Figure 1).

MATERIALS AND METHODS

A retrospective chart review of patients presenting to the Children’s Medical Center of Dallas between 2007 and 2021 was performed. Patients with HLHS who had undergone tricuspid valve repair or replacement were identified from our surgical database. The study was approved by the Institutional Review Boards for Human Research (IRB STU-2021-0451 approved June 28, 2021) at the University of Texas Southwestern Medical Center and Children’s Medical Center.

The medical records of patients were reviewed for age, gender, and medical and surgical history, including anticoagulation regimen (as well as associated events of significant bleeding, thrombosis, or embolism). Echocardiograms, cardiac catheterizations, and surgical reports were obtained. Estimates of TR and right ventricular systolic function were recorded from the echocardiogram performed most immediately before the tricuspid valve surgery. Additional measurements and assessments were made on the echocardiogram during the period of data collection if quantification of TR or right ventricular (RV) function were not provided in the original report. During the study period, the decision of whether and when to intervene on the tricuspid valve was individualized and not driven by protocol or defined threshold. In general, patients with moderate or greater tricuspid valve regurgitation and mild or no right ventricular dysfunction underwent tricuspid intervention. Patients with both significant TR and significant right ventricular dysfunction were diverted to the transplant pathway. For the purposes of analysis, patients were grouped by the type of definitive tricuspid operation; specifically, patients who underwent tricuspid repair and later or contemporaneously underwent replacement were considered in the replacement group. Because the repair and replacement groups were thought to be fundamentally dissimilar, no attempt was made to compare the groups statistically. Only summary descriptive statistics are provided for each group. Timepoints for outcomes were defined as early (≤ 30 days), midterm (31 days to 5 years) and late (>5 years).

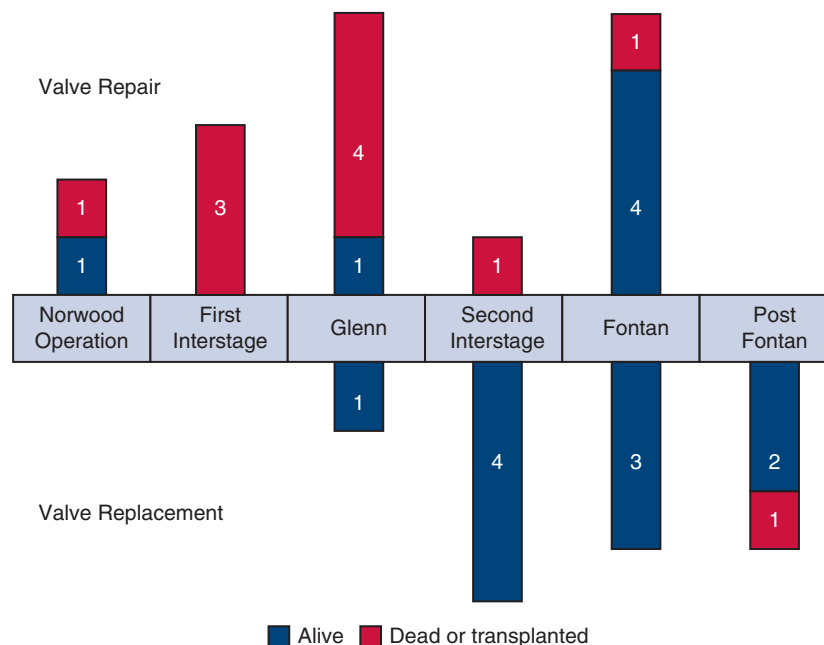


FIGURE 1. Timing and outcomes for tricuspid valve surgery in patients with HLHS.

RESULTS

Patients (n = 27) underwent intended definitive tricuspid valve intervention at 1 of 6 time points: at Norwood operation, in the first interstage, at Glenn operation, in the second interstage, at Fontan operation, or after the Fontan operation (Figure 1). Repair was the only approach undertaken through the Glenn operation, and thereafter, replacement was the predominant approach, although a number of repairs were also done in the post-Glenn stages. Time-related freedom from death or transplant after definitive operation is presented in Figure 2.

Tricuspid Valve Replacement

Eleven patients (73% male) underwent mechanical tricuspid valve replacement (Table 1). The median age at development of moderate or greater TR was 1 years (range, in utero to 8 years), with median time to valve replacement of 2 years (range, 5 months to 5 years). All patients were medically managed with diuretics or an afterload reducing agent before valve replacement. Despite medical management, all 11 patients had at least moderate TR at the time of surgery, and 9 patients (82%) had tricuspid annular dilation (Z score ≥ 2). All patients had evidence of prolapse on preoperative echocardiogram of varying degrees. In addition, valvular thickening, dysplasia, leaflet restriction, and chordal elongation were also identified preoperatively. Before valve replacement, RV systolic function was preserved in 10 patients (91%), and the remaining patient had mildly diminished systolic function.

Preoperative cardiac catheterization demonstrated elevated right ventricular end-diastolic pressure (≥ 10 mm Hg) in 6 patients (55%). Only 1 patient had a prior surgery for valve repair, which was accomplished in the second interstage (after Glenn operation), and this patient was analyzed as a replacement rather than a repair. The repair was an Alfieri repair with Kaye annuloplasty of the antero-septal commissure in a child with moderate to severe TR

secondary to a deep central cleft in the anterior leaflet, a deficient thin and pliable septal leaflet, and no posterior leaflet. There was residual mild TR postoperatively that progressed over the next 9 months requiring valve replacement at the time of the Fontan operation, at age 4 years.

The median age at time of mechanical valve replacement was 3 years (range, 5 months to 9 years). The median cardiopulmonary bypass time was 91 minutes (range, 45-338 minutes), and median crossclamp time was 44 minutes (range, 28-134 minutes) (Table 2). All patients were extubated within the first postoperative day and required inotropic support for a median of 1 day (range, 1-4 days). The median duration of intensive care unit stay was 5 days (range, 2-39), and median hospital stay was 14 days (range, 8-46).

In the entire cohort, 7 patients (64%) had concomitant cardiac procedures at the time of valve replacement. These procedures included unsuccessful attempts at tricuspid valve repair (n = 2), Fontan operation (n = 3), Fontan revision, pacemaker insertion, Maze procedure, and right atrial reduction. Eight patients had valve replacement as a procedure separate from the Fontan: 5 (63%) before Fontan and 3 (37%) after. As expected, patients undergoing additional cardiac procedures had a longer median cardiopulmonary bypass time (132 vs 48 minutes) and crossclamp time (69 vs 33 minutes). There was no difference in the duration of mechanical ventilation and inotropy requirement. Patients undergoing additional cardiac procedures had a longer median intensive care unit stay by 4 days.

Regarding anticoagulation, 8 patients were started on intravenous heparin postoperatively (range, 1-3 days) with transition to aspirin and coumadin 1 to 4 days later. The international normalized ratio was monitored with a therapeutic goal value range of 2.5 to 3.5. The remaining 3 patients (aged 2-3 years) were started on subcutaneous enoxaparin sodium (Lovenox, Sanofi) postoperatively and switched to Coumadin on an outpatient basis. One patient

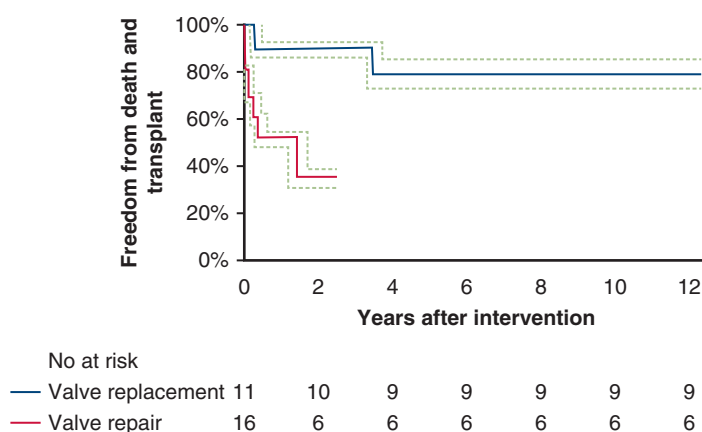


FIGURE 2. Kaplan–Meier curve depicting freedom from death and transplant in patients undergoing tricuspid valve repair (red, n = 16) and tricuspid valve replacement (blue, n = 11). Because the groups were dissimilar, no statistical comparison was made.

TABLE 1. Tricuspid valve replacement

Patient	Age at diagnosis of moderate TR	Grade of TR before replacement	Mechanism of TR on echocardiogram	RVEDp	RV systolic function	Age at replacement	Valve used	Additional intraoperative findings	Concomitant surgery	RV function on last echocardiogram	TR on last echocardiogram
1	In utero	Severe	Prolapse, annular dilation	10	Normal	5 mo	21-mm St Jude	Dysplasia, binding of septal leaflet to septum with abnormal chordae, elongated chordae of anterior leaflet	Glenn	Normal	None
2	1 y	Severe	Prolapse and thickening of anterior leaflet, annular dilation	11	Normal	3 y	25-mm St Jude	None	None	Normal	Trivial
3	5 mo	Severe	Prolapse of anterior leaflet, restricted septal leaflet, annular dilation	8	Normal	2 y	23-mm St Jude	Extreme dysplasia and restriction with multiple secondary chordae and papillary muscle fusion to the underside of the septal leaflet.	None	Normal	None
4	5 mo	Severe	Prolapse of anterior leaflet, restricted septal leaflet, annular dilation	12	Normal	4 y	29-mm St Jude	None	Alfieri repair	Moderately depressed	None
5	1 y	Severe	Prolapse	9	Normal	2 y	23-mm St Jude	None	None	Severely depressed, transplanted	N/A
6	2 y	Moderate to severe	Prolapse of anterior leaflet, annular dilation	9-11	Normal	3 y	21-mm St Jude	Extreme dysplasia Multiple deep scallops in all leaflets	Fontan	Normal	Trivial
7	4 y	Severe	Prolapse, dysplasia, annular dilation	Not available	Normal	4 y	25-mm St Jude	Myxomatous valve, cleft, prolapse	Fontan, cleft closure, commissuroplasty	Normal	None
8*	2 y	Moderate	Annular dilation	12	Mildly depressed	5 y	27-mm St Jude	Prolapse	Fontan, pacemaker insertion	Moderately depressed	None

(Continued)

TABLE 1. Continued

Patient	Age at diagnosis of moderate TR	Grade of TR before replacement	Mechanism of TR on echocardiogram	RVEDp	RV systolic function	Age at replacement	Valve used	Additional intraoperative findings	Concomitant surgery	RV function on last echocardiogram	TR on last echocardiogram
9	2 wk	Moderate to severe	Dysplasia, prolapse of anterior and septal leaflet	7	Normal	5 y	23-mm St Jude	None	None	Normal	None
10	8 y	Severe	Annular dilation, prolapse of anterior leaflet	8	Normal	9 y	31-mm ATS Medical, Inc	None	Maze, right atrial reduction, Fontan revision	Normal	None
11	2 y	Moderate to severe	Prolapse of anterior and septal leaflet, annular dilation	10	Normal	5 y	25-mm St Jude	None	Resection of membrane at ostium of right pulmonary vein	Moderately depressed [†]	Trivial

TR, Tricuspid regurgitation; RVEDp, right ventricular end-diastolic pressure; RV, right ventricle; NA, not available. *Patient underwent tricuspid valve repair at 3 years after presenting with severe TR and depressed RV function. Underwent optimization with oral heart failure medications with improvement in systolic function and TR. †Tricuspid valve replaced 2 years later due to worsening TR. ‡Patient deceased.

was transitioned to Coumadin after 2 years, and 2 after 2 months. Anti-Xa levels were monitored with a therapeutic goal value of 0.5 to 1. No major or minor bleeding or thromboembolic events were reported during the early or midterm follow-up period.

The patients were followed for a median of 4 years (range, 1-12). There was no early mortality in this cohort. One patient (9%) died 4 years after valve replacement. This patient was diagnosed with moderate TR at 2 years old and underwent replacement 4 years later, after Fontan completion. Preoperative assessment demonstrated low normal right ventricular systolic function, borderline elevated right ventricular end-diastolic pressure of 10 mm Hg, and moderate to severe TR with leaflet prolapse and annular dilation. The patient had moderately depressed ventricular function postoperatively that worsened over the next 4 years, leading to referral for transplantation. Unfortunately, there was hemodynamic decompensation after pretransplant catheterization, culminating in cardiac arrest, and the patient ultimately died.

One patient in the cohort received a heart transplant 5 months after valve replacement. This patient was diagnosed with severe TR at 1 year and underwent valve replacement at 2 years. Similar to the previously mentioned patient, preoperative assessment demonstrated low normal right ventricular function, borderline elevated right ventricular end-diastolic pressure of 10 mm Hg and severe TR with leaflet prolapse, annular dilation, and excessive chordal elongation. He developed moderately depressed ventricular function postoperatively that worsened over the next 3 months, and he received a donor heart shortly after being listed.

Of the remaining 9 patients, 8 had normal right ventricular systolic function on last echocardiogram. One patient had moderately depressed function at 1-year follow-up. This patient had undergone unsuccessful tricuspid valve repair at 3 years of age followed by a valve replacement at 5 years with the Fontan operation. Right ventricular systolic function was mildly depressed before valve replacement, and surgery was complicated by complete heart block necessitating a permanent pacemaker. This patient's RV function progressed over the next year to moderately depressed. With initiation of oral congestive heart failure medications, RV systolic function improved to low normal at 3 years follow-up.

Tricuspid Valve Repair

Sixteen patients (69% male) underwent tricuspid valve repair (Table 3). The median age at development of at least moderate TR was 4 months (range, 8 days to 4 years). Before valve replacement, RV systolic function was preserved in 13 patients (81%), and function was mildly depressed in 2 patients (12%) and moderately depressed in 1 patient (6%). These patients underwent repair at a median age of 4 months (range, 8 days to 4 years) and median

TABLE 2. Operative details

	Valve replacement median (range)	Valve repair median (range)
Age at surgery (y)	3 (0.5-9)	0.3 (0.02-3)
Weight at surgery (kg)	15.2 (7-25)	5.8 (3-24.2)
Cardiopulmonary bypass time (min)	91 (45-338)	107 (45-272)
Clamp time (min)	44 (28-134)	49 (21-164)
	Number (n = 11)	Number (n = 16)
Additional intraoperative valvar findings		
Dysplasia	4	2
Prolapse	2	7
Cleft	1	2
Restriction	1	3
Dilation	0	4
Type of mechanical valve		
St Jude 21 mm	2	
St Jude 23 mm	3	
St Jude 25 mm	3	
St Jude 27 mm	1	
St Jude 29 mm	1	
ATS Medical, Inc 31 mm	1	
Concomitant surgery		
Attempt at valve repair	1	N/A
Norwood	0	2
Fontan	3	5
Permanent pacemaker insertion	1	0
Maze procedure with right atrial plication	1	0
Fontan revision	1	0
Glenn	1	5
Resection of pulmonary venous obstruction	1	1
None	2	2
Intraoperative/postoperative complications		
Sinus bradycardia	1	0
First-degree AV block (transient)	2	0
First-degree AV block (persistent)	1	0
Second-degree block (transient)	1	0
Junctional rhythm (+accelerated)	4	3
Complete heart block requiring pacemaker	1	1
Need for extracorporeal support	0	1

AV, Atrioventricular.

size of 5.8 kg (range, 3-24.2). Fourteen patients (88%) underwent repair during a concomitant operation (Table 3).

These patients were followed for a median duration of 5 months (range, 2 weeks to 3 years). There was no early mortality, there were 9 (56%) midterm mortalities, and 1 patient received a heart transplant (6%) (Figure 3). Of the remaining 6 patients, 4 had normal RV systolic function on last echocardiogram and 2 had mildly depressed function. All 6 had residual TR; 3 had mild and 3 had moderate TR. Of the 9 mortalities, 6 were not considered candidates for bailout tricuspid valve replacement secondary to poor ventricular function that was thought highly likely to be exacerbated by a "suddenly competent" tricuspid valve. The remaining 3 patients were considered too small to undergo valve replacement.

DISCUSSION

Survival in patients with single ventricles undergoing surgical palliation has greatly improved, with current Fontan survival of 80% at 30 years.²⁴ This decrease in mortality rate over the years is due to a combination of factors including improved surgical techniques, better perioperative care, and more judicious patient selection. However, presence of significant AVVR remains an important independent risk factor for both early and late outcomes post-Fontan,²⁵ and a great deal of focus has been given to surgical repair of the tricuspid valve as a way to improve Fontan candidacy/survival in these children. Because of the complex etiology of tricuspid valve dysfunction, surgical repair is not always straightforward or reproducible and patients may develop worsening regurgitation at any point after repair.^{26,27} In the present study, the results of repair, in a group too young or too small to undergo replacement, were disappointing, with a high rate of attrition or diversion to the transplant pathway.

Replacement of the AVV with a mechanical valve is generally avoided in the pediatric population due to absence of growth potential, lack of available prostheses in very small sizes, risk of injury to the conduction system, and the difficulties of chronic anticoagulation in children.² Mahle and colleagues² reported on 17 patients with mixed variations of single ventricle physiology who underwent AV valve replacement, 44% of whom developed complete heart block. There were 4 late deaths (24%) and 1 patient (6%) required heart transplant in their series, but overall improvement in mortality and morbidity was achieved. Sughimoto and associates²¹ reported on 56 patients undergoing valve replacement, half of whom had depressed ventricular function, and reported a 34% 3-year mortality. Recently, Stephens and Dearani²⁸ have advocated for a lower threshold to proceed with valve replacements, especially in patients with a tricuspid valve.

TABLE 3. Tricuspid valve repair

Patient	Age at diagnosis of moderate TR	Grade of TR before replacement	Mechanism of TR on preoperative echocardiogram	RVEDp	RV systolic function	Age at repair	Technique used	TR on postoperative echocardiogram	Concomitant surgery	RV function on last echocardiogram	TR on last echocardiogram
1	25 d	Moderate to severe	Prolapse, restriction, annular dilation	Unavailable	Normal	25 d	Annuloplasty	Trivial	Norwood	Patient deceased	N/A
2	8 d	Moderate	Data unavailable	Unavailable	Normal	8 d	Kaye-Wooler commisuroplasty	Mild to moderate	Norwood	Normal	Moderate
3	2 mo	Moderate	Annular dilation	11	Normal	2 mo	Kay Wooler commisuroplasty	Severe	Sutureless repair of right pulmonary vein	Patient deceased	N/A
4	3 mo	Severe	Flail leaflet, annular dilation	Unavailable	Normal	3 mo	Dacron posterior annuloplasty	Trivial	Extracorporeal membrane oxygenation decannulation	Patient deceased	N/A
5	2 wk	Moderate to severe	Flail leaflet secondary to Sano stent migration, annular dilation	12	Normal	3 wk	DeVega annuloplasty type stitches at the anterior septal commissure and the posterior septal commissure	Moderate to severe	None	Patient deceased	N/A
6	2 mo	Moderate to severe	Annular dilation, restriction	13	Mildly depressed	3 mo	PTFE bridge	Moderate	Glenn	Patient deceased	N/A
7	3 mo	Severe	Prolapse of anterior leaflet, restriction of septal leaflet, annular dilation	12	Normal	4 mo	Anteroseptal commisuroplasty	Mild	Glenn	Transplanted	N/A
8	4 mo	Moderate	Prolapse, mild dysplasia, annular dilation	9	Normal	4 mo	Anteroseptal commisuroplasty	Mild	Glenn	Normal	Mild
9	4 mo	Moderate	Cleft, prolapse, annular dilation	10	Normal	5 mo	Septal-posterior and anteroseptal commisuroplasty and annuloplasty	Mild	Glenn	Patient deceased	N/A

(Continued)

TABLE 3. Continued

Patient	Age at diagnosis of moderate TR	Grade of TR before replacement	Mechanism of TR on preoperative echocardiogram	RVEDp	RV systolic function	Age at repair	Technique used	TR on postoperative echocardiogram	Concomitant surgery	RV function on last echocardiogram	TR on last echocardiogram
10	2 mo	Severe	Data unavailable	10	Normal	4 mo	Commisuroplasty, anuloplasty	Trivial	Glenn	Patient deceased	N/A
11	2 y	Moderate	Restriction, mild dysplasia, annular dilation	10	Normal	3 y	Anteroseptal commisuroplasty, anuloplasty	Mild	None	Transplanted	N/A
12	3 y	Moderate	Cleft, annular dilation	8	Normal	3 y	Alfieri repair	Mild	Fontan	Normal	Mild to moderate
13	6 mo	Severe	Prolapse of anterior leaflet, restriction of septal leaflet, annular dilation	11	Normal	2 y	Anteroseptal commisuroplasty	Mild	Fontan	Patient deceased	N/A
14	3 y	Moderate to severe	Prolapse, annular dilation	7-9	Moderately depressed	4 y	Edge to edge anteroseptal and Kaye-Wooler posteroseptal commisuroplasty	Mild	Fontan	Mildly depressed	Mild
15	4 y	Moderate	Annular dilation	9	Mildly depressed	4 y	DeVega anuloplasty with strip of CardioCel partially around anterior and posterior leaflets	Mild	Fontan	Mildly depressed	Mild
16	3 y	Moderate to severe	Prolapse, annular dilation	11	Normal	3 y	Alfieri stitch, Kay anuloplasty	Mild	Fontan	Normal	Mild to moderate

TR, Tricuspid regurgitation; RVEDp, right ventricular end-diastolic pressure; RV, right ventricle; NA, not available; PTFE, polytetrafluoroethylene.

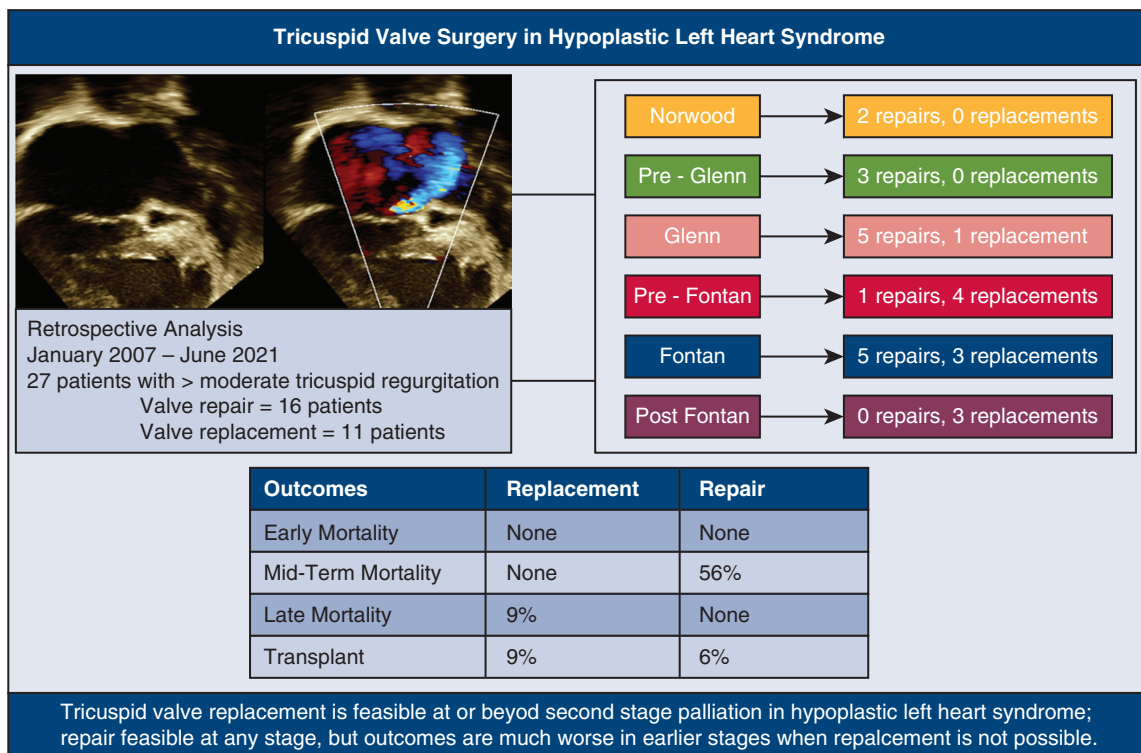


FIGURE 3. Mechanical replacement of the tricuspid valve in HLHS is feasible at or beyond second-stage palliation in HLHS; repair is feasible at any stage, but outcomes are worse in earlier stages when replacement is not possible.

Our patient cohort was homogenous, consisting of those with HLHS and anatomic tricuspid valves. Overall outcomes for tricuspid valve replacement were 0% early mortality and 9% late mortality, with complete heart block in 1 patient (9%). Heart block was less common in our cohort than in the report by Mahle and colleagues,² perhaps explained by exclusion of heterotaxy and atrioventricular discordance in our series. Our patients had no adverse hemorrhagic or thrombotic outcomes with anticoagulation despite a young age at replacement (median 3 years). This is influenced by a dedicated outpatient cardiology pharmacy program that allows for close monitoring and adjustments of anticoagulation regimens, but also likely reflects a still relatively short median follow-up.

Right ventricular systolic function was preserved in 8 of the 9 transplant-free survivors at a median follow-up of 4 years. One patient with mild systolic dysfunction before surgery initially developed moderate dysfunction after surgery, perhaps related to surgical heart block and the need for pacemaker. With goal-directed heart failure therapy, function has returned to low normal on most recent evaluation. These data suggest that normal RV systolic function before AVV replacement may allow for preserved ventricular function in 80% of patients.

Study Limitations

There are several limitations of the present study, including a sample size that is relatively small, as would be expected from a single-center study. Moreover, the period of study is relatively long, during which time the approach to such patients has inevitably evolved. In addition, as mentioned there was no process of standardized decision making for timing and type of intervention for the repair or replacement group. Perhaps most important, the study should not be interpreted as a comparison of repair and replacement because the techniques are reasonable alternatives, nor should it be viewed as an attempt to advocate for the superiority of replacement versus repair. We continue to believe that, other things being equal, repair is superior to replacement.

CONCLUSIONS

If necessary, mechanical replacement of the tricuspid valve can be accomplished safely with acceptable and encouraging early and midterm results, despite the complexity of associated anticoagulant therapy. When undertaken in the setting of normal right ventricular systolic function, early data suggest ventricular function may be preserved in most patients.

Conflict of Interest Statement

The authors reported no conflicts of interest.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling or reviewing manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

References

1. Reyes A II, Bove EL, Mosca RS, Kulik TJ, Ludomirsky A. Tricuspid valve repair in children with hypoplastic left heart syndrome during staged surgical reconstruction. *Circulation*. 1997;96(9 Suppl):II-341-3; discussion II-344-5.
2. Mahle WT, Gaynor JW, Spray TL. Atrioventricular valve replacement in patients with a single ventricle. *Ann Thorac Surg*. 2001;72:182-6.
3. Elmi M, Hickey EJ, Williams WG, Van Arsdell G, Caldarone CA, McCrindle BW. Long-term tricuspid valve function after Norwood operation. *J Thorac Cardiovasc Surg*. 2011;142:1341-7.e4.
4. Barber G, Helton JG, Aglira BA, Chin AJ, Murphy JD, Pigott JD, et al. The significance of tricuspid regurgitation in hypoplastic left-heart syndrome. *Am Heart J*. 1988;116(6 Pt 1):1563-7.
5. Gaynor JW, Mahle WT, Cohen MI, Ittenbach RF, DeCampi WM, Steven JM, et al. Risk factors for mortality after the Norwood procedure. *Eur J Cardiothorac Surg*. 2002;22:82-9.
6. Sano S, Huang SC, Kasahara S, Yoshizumi K, Kotani Y, Ishino K. Risk factors for mortality after the Norwood procedure using right ventricle to pulmonary artery shunt. *Ann Thorac Surg*. 2009;87:178-85; discussion 185-6.
7. Lee TM, Aiyagari R, Hirsch JC, Ohye RG, Bove EL, Devaney EJ. Risk factor analysis for second-stage palliation of single ventricle anatomy. *Ann Thorac Surg*. 2012;93:614-8; discussion 619.
8. Imai Y, Takanashi Y, Hoshino S, Terada M, Aoki M, Ohta J. Modified Fontan procedure in ninety-nine cases of atrioventricular valve regurgitation. *J Thorac Cardiovasc Surg*. 1997;113:262-8; discussion 269.
9. Driscoll DJ, Offord KP, Feldt RH, Schaff HV, Puga FJ, Danielson GK. Five- to fifteen-year follow-up after Fontan operation. *Circulation*. 1992;85:469-96.
10. Podzolkov VP, Chiaureli MR, Yurlov IA, Zelenikin MM, Kovalev DV, Dontsova VI, et al. Results of Fontan operation in patients with atrioventricular valve regurgitation. *Eur J Cardiothorac Surg*. 2015;48:308-14; discussion 314-5.
11. Bautista-Hernandez V, Scheurer M, Thiagarajan R, Salvin J, Pigula FA, Emani S, et al. Right ventricle and tricuspid valve function at midterm after the Fontan operation for hypoplastic left heart syndrome: impact of shunt type. *Pediatr Cardiol*. 2011;32:160-6.
12. Stamm C, Anderson RH, Ho SY. The morphologically tricuspid valve in hypoplastic left heart syndrome. *Eur J Cardiothorac Surg*. 1997;12:587-92.
13. Nii M, Guerra V, Roman KS, Macgowan CK, Smallhorn JF. Three-dimensional tricuspid annular function provides insight into the mechanisms of tricuspid valve regurgitation in classic hypoplastic left heart syndrome. *J Am Soc Echocardiogr*. 2006;19:391-402.
14. Takahashi K, Inage A, Rebeyka IM, Ross DB, Thompson RB, Mackie AS, et al. Real-time 3-dimensional echocardiography provides new insight into mechanisms of tricuspid valve regurgitation in patients with hypoplastic left heart syndrome. *Circulation*. 2009;120:1091-8.
15. Kutty S, Colen T, Thompson RB, Tham E, Li L, Vijarnsorn C, et al. Tricuspid regurgitation in hypoplastic left heart syndrome: mechanistic insights from 3-dimensional echocardiography and relationship with outcomes. *Circ Cardiovasc Imaging*. 2014;7:765-72.
16. Sugiura J, Nakano T, Oda S, Usui A, Ueda Y, Kado H. Effects of tricuspid valve surgery on tricuspid regurgitation in patients with hypoplastic left heart syndrome: a non-randomized series comparing surgical and non-surgical cases. *Eur J Cardiothorac Surg*. 2014;46:8-13.
17. Bautista-Hernandez V, Myers PO, Loyola H, Marx GR, Bacha EA, Baird CW, et al. Atrioventricular valve annular remodeling with a bioabsorbable ring in young children. *J Am Coll Cardiol*. 2012;60:2256-8.
18. Ohye RG, Gomez CA, Goldberg CS, Graves HL, Devaney EJ, Bove EL. Tricuspid valve repair in hypoplastic left heart syndrome. *J Thorac Cardiovasc Surg*. 2004;127:465-72.
19. Kwak JG, Park CS, Lee CH, Lee C, Kim SJ, Song JY, et al. Early surgical correction of atrioventricular valvular regurgitation in single-ventricle patients. *Ann Thorac Surg*. 2010;90:1320-3.
20. Wong DJ, Iyengar AJ, Wheaton GR, Ramsay JM, Grigg LE, Horton S, et al. Long-term outcomes after atrioventricular valve operations in patients undergoing single-ventricle palliation. *Ann Thorac Surg*. 2012;94:606-13; discussion 613.
21. Sugimoto K, Hirata Y, Hirahara N, Miyata H, Suzuki T, Murakami A, et al. Mid-term result of atrioventricular valve replacement in patients with a single ventricle. *Interact Cardiovasc Thorac Surg*. 2018;27:895-900.
22. Nakata T, Hoashi T, Shimada M, Ozawa H, Higashida A, Kurosaki K, et al. Systemic atrioventricular valve replacement in patients with functional single ventricle. *Semin Thorac Cardiovasc Surg*. 2019;31:526-34.
23. Alshami N, Sarvestani AL, Thomas AS, St Louis J, Kochilas L, Raghuvier G. Valve replacement in children with single ventricle physiology. *Pediatr Cardiol*. 2020;41:129-33.
24. d'Udekem Y, Iyengar AJ, Galati JC, Forsdick V, Weintraub RG, Wheaton GR, et al. Redefining expectations of long-term survival after the Fontan procedure: twenty-five years of follow-up from the entire population of Australia and New Zealand. *Circulation*. 2014;130(11 Suppl 1):S32-8.
25. Downing TE, Allen KY, Glatz AC, Rogers LS, Ravishankar C, Rychik J, et al. Long-term survival after the Fontan operation: twenty years of experience at a single center. *J Thorac Cardiovasc Surg*. 2017;154:243-53.e2.
26. Muntaner CD, King G, Zannino D, Alphonso N, Finucance K, Winlaw D, et al. Poor late outcomes after tricuspid valve repair in a single ventricle: experience of 103 patients. *Ann Thorac Surg*. 2021;111:987-94.
27. Alsoufi B, Sinha R, McCracken C, Figueroa J, Altin F, Kanter K. Outcomes and risk factors associated with tricuspid valve repair in children with hypoplastic left heart syndrome. *Eur J Cardiothorac Surg*. 2018;54:993-1000.
28. Stephens EH, Dearani JA. Management of the bad atrioventricular valve in Fontan...time for a change. *J Thorac Cardiovasc Surg*. 2019;158:1643-8.

Key Words: anticoagulation, hypoplastic left heart syndrome, single ventricle, tricuspid regurgitation, valve replacement