Success and limitations of right ventricular sinus myectomy for pulmonary atresia with intact ventricular septum

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Objectives: Right ventricular sinus myectomy has been proposed for pulmonary atresia with intact ventricular septum for morphology falling within the uncertain area for eventual biventricular repair. Our objective was to evaluate right ventricular sinus myectomy by characterizing the morphologic spectrum of these patients, determining whether biventricular repair was achieved, ascertaining growth of right-sided structures, and assessing survival.

Methods: We evaluated medical records, all imaging studies, and follow-up data (complete in all but 1 patient) from 43 patients with pulmonary atresia with intact ventricular septum treated from October 1993 to July 2005, 16 of whom underwent right ventricular sinus myectomy. Serial echocardiographic measurements of right-sided cardiac structures were converted to Z values to estimate their growth relative to somatic growth.

Results: Patients undergoing right ventricular sinus myectomy had mild-to-moderate right ventricular size diminution (grade -1.2 ± 3.2) and a tricuspid valve Z value of -4.9 ± 1.9 . Thirteen (87%) of the 16 patients achieved biventricular repair. After right ventricular sinus myectomy, mean right ventricular cavity size grade increased to 1.4 ± 0.66 , but the tricuspid valve Z value did not change appreciably over time. Five-year survival after sinus myectomy was 85%; late deaths were in patients with the smallest tricuspid valves at presentation (Z value < -7).

Conclusions: Right ventricular sinus myectomy in the uncertain area for biventricular repair of pulmonary atresia with intact ventricular septum leads to immediate increase in right ventricular volume. It, in combination with establishing right ventricle–pulmonary trunk continuity, allowed early biventricular repair in 87% of patients. However, tricuspid valve growth in relation to somatic growth was minimal. Thus, small tricuspid valve size might limit the long-term success of biventricular repair achieved by means of right ventricular sinus myectomy.

Pulmonary atresia with intact ventricular septum (PAIVS) is a rare congenital cardiac anomaly with a wide spectrum of morphologic heterogeneity.^{1,2} At one end are patients with severe right ventricular (RV) hypoplasia, often with RV-dependent coronary circulation requiring a single-ventricle solution. At the other end are patients with normal RV size with adequate inflow and outflow portions that can be managed with biventricular repair. In between are patients with

Abbreviations and Acronyms

CHSS	= Congenital Heart Surgeons' Society
IRB	= institutional review board
PAIVS	= pulmonary atresia with intact ventricular septum
RV	= right ventricular

moderate RV hypoplasia whose optimal surgical management and ultimate results remain uncertain.

In managing PAIVS, some surgeons routinely perform a systemic-to-pulmonary artery shunt in nearly all, then perform a cavopulmonary anastomosis, and finally convert to Fontan circulation early in life^{3,4}; others perform a pulmonary valvotomy or transannular patch, with or without a systemic-to-pulmonary-artery shunt, in the hope that with time, RV structures will enlarge sufficiently to support a biventricular circulation.⁵ In 1986, Joshi, Brawn, and Mee⁶ proposed performing RV infundibular resection as part of early palliation of PAIVS when a well-formed RV infundibulum was present, leading to an imperforate pulmonary valve, and RV-dependent circulation was absent. Later, this became sinus resection⁷ and was combined with pulmonary valvotomy and tricuspid valvotomy, if necessary (Figure 1). They believed RV resection would immediately improve RV capacity, promote growth of right-sided cardiac structures, and foster successful biventricular repair.

The purpose of this study was to evaluate results of a treatment program for PAIVS that included RV sinus myectomy. Specifically, we sought to (1) characterize the right-sided morphologic spectrum of patients undergoing RV sinus myectomy, contrasting it with those who were managed with either univentricular or biventricular repair without RV sinus myectomy; (2) determine the likelihood of achieving biventricular repair with RV sinus myectomy; (3) ascertain whether RV sinus myectomy indeed fosters growth of right-sided cardiac structures; and (4) assess survival after RV sinus myectomy.

Materials and Methods**Patients**

From October 1993 to July 2005, 43 patients underwent at least 1 palliative or definitive surgical procedure for PAIVS at Cleveland Clinic Children's Hospital. Sixteen (37%) had undergone previous palliative procedures elsewhere. Surgical intent to treat was designated biventricular when initial palliation included pulmonary valvotomy and univentricular when it included a systemic-to-pulmonary artery shunt alone.

Data

Surgical details were recorded prospectively in the Congenital Heart Surgery Registry, and catheter-based interventions were recorded prospectively into the Pediatric Catheterization Registry. Both have been approved for use in research by the institutional review

board (IRB), with patient or parental consent waived. Details of relevant percutaneous and operative procedures performed elsewhere were obtained from referring cardiologists.

Morphology

With IRB approval, preoperative and serial postoperative transthoracic echocardiograms for each patient were reviewed by a pediatric cardiologist (GLR), a pediatric cardiology fellow (UM), and a cardiac surgery fellow (RB). Diameters of tricuspid valve, pulmonary valve, and branch pulmonary arteries were measured with calipers. These diameters were converted to Daubeny Z values⁸: size equal to mean normal for body surface area was designated Z value = 0, size 1 standard deviation below mean normal was designated Z value = -1, and so forth. RV chamber size was graded qualitatively from -5 to +5, with negative numbers representing degree of RV chamber diminution from extreme (-5) to mild (-1), normal size designated by 0, and positive numbers representing degree of chamber enlargement from mild (+1) to extreme (+5), as defined by the Congenital Heart Surgeons' Society (CHSS) multi-institutional study.⁹

Follow-up

An IRB-approved questionnaire was mailed to each referring cardiologist. In addition, parents, cardiologists, or physicians were contacted directly to ascertain vital status, functional status, current medications, subsequent procedures, echocardiograms (reports and analog or digital media), and catheterization reports. Median follow-up was 1.6 years. Twenty-five percent of living patients were followed more than 6.5 years, and 10% were followed more than 10 years. A total of 137 patient-years of data were available for analysis. Follow-up was complete in all but 1 patient.

Data Analysis

All analyses were performed with SAS (SAS, Inc, Cary, NC) statistical software. Continuous variables were summarized by cumulative distribution functions, as well as means $\pm$ standard deviations and as equivalent 15th, 50th, and 85th percentiles when values were skewed, with comparisons made by the Wilcoxon rank-sum nonparametric test. Categorical data were summarized as frequencies and percentages, with the χ^2 test or Fisher's exact test (when the expected frequency was <5) used for comparison. Temporal trend analyses of RV cavity size grade, pulmonary and tricuspid valve measurements, and corresponding Z values was performed using longitudinal mixed modeling for repeated measures (PROC MIXED; SAS, Inc).¹⁰ Uncertainty of time-related estimates was expressed by asymmetric confidence limits equivalent to ± 1 standard deviation (68%).

Results**RV Sinus Myectomy**

Sixteen patients underwent RV sinus myectomy at a median age of 1.25 years (15% at <0.4 years and 15% at >2.9 years). It had been preceded in 14 (88%) patients by a systemic arterial-to-pulmonary artery shunt and in 13 (81%) by pulmonary valvotomy (Table 1); we deemed that intent to treat among these 13 was biventricular repair (Figure 2). An infundibular chamber was present in all and small in only 1; none had RV-coronary fistulae (Table 1).

Figure 1. Right ventricular myectomy is performed by means of a combined transatrial/transpulmonary approach. **A,** On the *left*, the *dashed line* indicates the intended extent of right ventricular sinus resection. On the *right*, the figure represents the results of sinus resection through the tricuspid valve. **B,** On the *left*, an incision is made in the pulmonary trunk. At *top right*, pulmonary valvotomy is shown. This reveals infundibular muscle (*lower right*) that will be resected (*dashed line*) to complete the right ventricular myectomy. RVT, Right ventricular trabecular sinus.

At the time of RV sinus myectomy, additional procedures included infundibular muscle resection in 14 patients (88%); pulmonary valvotomy, valvuloplasty, or replacement in 13 (81%); and tricuspid valvotomy or valvulo-

plasty in 10 (63%) (Table 2). Thus, this multicomponent procedure has been characterized colloquially as “RV overhaul.” In addition, a systemic-to-pulmonary artery shunt was closed in 11 patients (69%), and an atrial septal

TABLE 1. Patient characteristics

Variable	Intent to treat			P value
	BVR, no RVSM (n = 7)	RVSM (n = 16)	UVR, no RVSM (n = 20)	
	No. (%)	No. (%)	No. (%)	
Demography				
Male sex	7 (100)	9 (60)	8 (40)	.02
RV morphology				
Infundibulum				<.0001
Absent	0 (0)	0 (0)	17 (85)	
Small	0 (0)	1 (6.3)	0 (0)	
Present	3 (43)	5 (31)	2 (10)	
Well formed	4 (57)	10 (63)	1 (5.0)	
Coronary circulation				
RV-to-coronary fistula	0 (0)	0 (0)	6 (30)	.02
RV-dependent circulation	0 (0)	0 (0)	4 (20)	.08
Initial procedures				
Systemic–pulmonary shunt	6 (86)	14 (88)	18 (90)	
Pulmonary valvotomy	7 (100)	13 (81)	0 (0)	
Atrial septal procedure				
Septectomy	1 (14)	0 (0)	8 (40)	
Balloon septostomy	2 (28)	2 (12)	6 (30)	

BVR, Biventricular repair; RVSM, right ventricular sinus myectomy; UVR, univentricular repair; RV, right ventricle.

Figure 2. Flow chart of the 43 patients progressing toward definitive repair. See [Appendix E1](#) for details. *a*, Alive; *BVR*, biventricular repair; *d*, dead; *RVSM*, right ventricular sinus myectomy; *UVR*, univentricular repair; *VR*, ventricular repair.

defect was partially or completely closed in 12 (75%), resulting at that operation in biventricular repair in 11 (69%) patients.

Comparison

Right-sided structures at presentation (earliest echocardiograph) of patients undergoing RV sinus myectomy were similar in size to those of patients undergoing biventricular repair directly ([Table 3](#)), although patients undergoing RV sinus myectomy achieved biventricular repair at an earlier age (median of 1.25 years vs 2.0 years; [Figure E1](#)). Both of these groups had larger right-sided structures, including their tricuspid valves ([Figure 3](#)), than those of patients undergoing univentricular repair ([Table 3](#)).

Biventricular Repair

Of the 16 patients undergoing RV septal myectomy, 13 eventually achieved biventricular repair, 8 of whom were left with a small residual atrial septal defect similar in size to a patent foramen ovale ([Table 2](#)). To achieve biventricular repair, 4 required procedures subsequent to RV sinus myectomy, 1 catheter-based systemic-to-pulmonary shunt closure and 3 atrial septal defect or systemic-pulmonary shunt closure in

TABLE 2. Description of right ventricular sinus myectomy operations

Patient no.	Intent to treat	Additional muscle resection	PV management	Concomitant procedures			Definitive repair	Subsequent procedures	End state
				Systemic-pulmonary shunt closure	ASD closure	TV management			
1	BVR	Infundibulum	Valvotomy	Partial	No	Valvotomy	No	None	Death before definitive repair
2	UVR	Infundibulum	None	Complete*	No	Valvotomy	No	†	One and a half ventricle repair
3	BVR	Infundibulum	Repair	Complete	Complete	None	Yes	None	BVR
4	BVR	Infundibulum	Valvotomy	Complete	Partial	Valvotomy	Yes	None	BVR
5	BVR	None	Valvotomy, monocusp	Complete	Complete	Anuloplasty	Yes	None	BVR
6	UVR	None	Valvotomy	Complete	Partial	Valvuloplasty	Yes	None	BVR
7	BVR	Infundibulum	Valvuloplasty	No‡	Partial	Valvuloplasty	Yes	None	BVR
8	BVR	Infundibulum	Valvotomy	Shunt performed	No	Valvotomy	No	§	BVR
9	BVR	Infundibulum	Valvuloplasty	Complete	Partial	Valvotomy	Yes	None	BVR
10	BVR	Infundibulum	None	Complete	Partial	None	Yes	None	BVR
11	BVR	Infundibulum	Valvotomy	Complete	Complete	None	Yes	None	BVR
12	BVR	Infundibulum	Valvotomy	Complete	Partial	Valvotomy	Yes	None	BVR
13	UVR	Infundibulum	RV-PT conduit	Complete	Complete	None	Yes	¶	BVR
14	BVR	Infundibulum	None	No	No	None	No	None	Alive without definitive repair
15	BVR	Infundibulum	Monocusp	Complete	Complete	None	Yes	None	BVR
16	BVR	Infundibulum	Valvotomy	No	Partial	Valvuloplasty	No	#	BVR

PV, Pulmonary valve; ASD, atrial septal defect; TV, tricuspid valve; BVR, biventricular repair; UVR, univentricular repair; RV, right ventricle; PT, pulmonary trunk.

*Bidirectional Glenn procedure performed. †Right ventricle-pulmonary artery conduit, pulmonary valve replacement, partial atrial septal defect closure, tricuspid valvotomy. ‡Coil closure of patent ductus arteriosus before definitive repair. §Redo right ventricular sinus myectomy, repeat pulmonary valve replacement, shunt and atrial septal defect complete closure, repeat tricuspid valvotomy. ||Bidirectional Glenn taken down. ¶Tricuspid valvuloplasty plus right ventricle-pulmonary trunk conduit replacement. #Catheter-based shunt closure.

TABLE 3. Right-sided heart morphology (on earliest echocardiogram) in patients with pulmonary atresia and intact ventricular septum according to intent to treat

Variable	Intent to treat						<i>P</i> value†
	BVR, no RVSM (n = 7)		RVSM (n = 16)		UVR, no RVSM (n = 20)		
	n*	No. (%) or 15th/50th/85th Percentiles	n*	No. (%) or 15th/50th/85th Percentiles	n*	No. (%) or 15th/50th/85th Percentiles	
Tricuspid valve							
Diameter	7	0.8/0.9/1.1	14	0.85/1.1/1.47	13	0.4/0.68/1.27	.02
Ratio with respect to normal	7	0.56/0.82/0.94	14	0.6/0.75/0.87	13	0.31/0.49/0.78	.009
Area · m ^{−2}	7	1.8/3.39/4.64	14	2.03/3.12/4.14	13	0.52/1.3/2.59	.004
Z value	7	−8.38/−4.45/−2.52	14	−7.56/−4.95/−3.21	13	−15.95/−10.38/−5.96	.003
Degree of regurgitation	7		16		16		.005
None	0 (0)		5 (31)		14 (88)		
Mild	4 (57)		6 (38)		1 (6.3)		
Moderate	2 (29)		4 (25)		1 (6.3)		
Moderately severe	1 (14)		1 (6.3)		0 (0)		
RV cavity	6		9		11		.1
−5	0 (0)		0 (0)		0 (0)		
−4	2 (33)		2 (22)		8 (73)		
−3	3 (50)		2 (22)		1 (9.1)		
−2	0 (0)		2 (22)		0 (0)		
−1	0 (0)		1 (11)		0 (0)		
0	0 (0)		0 (0)		2 (18)		
1	0 (0)		0 (0)		0 (0)		
2	1 (17)		0 (0)		0 (0)		
3	0 (0)		1 (11)		0 (0)		
4	0 (0)		0 (0)		0 (0)		
5	0 (0)		1 (11)		0 (0)		
Pulmonary valve	6		12		9		
Diameter		0.4/0.5/0.85		0.49/0.8/1.02		0.23/0.5/1.4	.2
Z value		−8.14/−4.18/−1.55		−5.52/−2.29/−0.96		−12.19/−6.85/−2.75	.04
Pulmonary artery	4		10		9		
Diameter		0.5/0.6/0.72		0.65/0.95/1.25		0.3/0.63/1.66	
Z value		−4.81/−1.96/−0.8		−2.36/−0.48/0.18		−6.86/−2.95/−0.88	.03

BVR, Biventricular repair; RVSM, right ventricular sinus myectomy; UVR, univentricular repair; RV, right ventricular. *Data available. †Kruskal–Wallis test.

combination with additional pulmonary valve and other right-sided procedures. Of the remaining 3 of 16 patients, 1 underwent one and a half ventricle repair, 1 died before definitive repair, and 1 was alive without definitive repair at last follow-up.

Growth of Right-sided Structures

Although tricuspid valve size increased with patient growth after RV sinus myectomy (Figure 4, A), its relation to body size (Z value) increased minimally across time (*P* = .6) and remained small (Figure 4, B). For perspective, the average Z value of −5.2 translates to approximately 72% of normal size and a tricuspid valve area index of 3 cm² · m⁻². In contrast, RV cavity size increased after RV sinus myectomy from an average grade of −1.2 (mild-to-moderate diminution)

to 1.4 (mild-to-moderate enlargement, Figure 5). Similarly, pulmonary valve size increased to near normal (Figure 6).

Survival

To date, 3 patients have died after RV sinus myectomy. Actuarial survival after RV sinus myectomy at 1, 5, and 10 years was 94%, 85%, and 64%, respectively. The earliest death occurred 7 months postoperatively, before definitive repair. Preoperative echocardiographic analysis recorded no tricuspid regurgitation, and postoperative tricuspid valve size was not recorded. Two patients died subsequent to definitive repair. One death occurred 3.4 years after biventricular repair; the postoperative Z value of the tricuspid valve was −7.6, and tricuspid regurgitation was trivial. The other

Figure 3. Cumulative distribution of tricuspid valve (TV) diameter on first echocardiogram, expressed in standardized Z-value format, of patients undergoing right ventricular sinus myectomy (RVSM) contrasted with those undergoing direct biventricular repair without myectomy (BVR) and the remaining patients whose intent to treat was univentricular repair (UVR).

occurred 4.4 years after one and a half ventricle repair; the postoperative Z value of the tricuspid valve at 2.5 years after repair was -8.2 .

Discussion

RV Sinus Myectomy

RV size has been the primary determinant of surgical management of PAIVS.¹¹ For patients with a nearly normal- or large-sized RV and those with severe RV hypoplasia, management decisions are easy.^{12,13} The dilemma is how best to manage patients whose RV size falls between these extremes. Although the conventional wisdom is that 2 ventricles are better than 1, a successful univentricular or one

and a half ventricle repair might yield better long-term results than a poor or marginal biventricular repair.

At the time our patients were treated, RV sinus myectomy for patients in the morphologically uncertain area for biventricular repair was based on the presence or absence of a RV infundibulum^{6,7}. Those with a well-formed infundibulum underwent pulmonary valvotomy and resection of excess muscle bundles within the RV. This approach was based in part on the morphologic RV categorization proposed by Bull and colleagues.¹ The present study suggests that a critically important additional factor when considering sinus myectomy for eventual biventricular repair is the size of the tricuspid valve annulus.

Figure 4. Size of the tricuspid valve (TV) across time after right ventricular sinus myectomy. Closed circles are grouped mean values, unadjusted for repeated measures, to illustrate raw data. The solid line is an estimate of mean TV size adjusted for repeated measures, and dashed lines are confidence limits equivalent to ± 1 standard deviation. A, Diameter. Average size at presentation was 1.18 ± 0.40 cm. B, Z value. Average size at presentation was -4.9 ± 1.9 .

Figure 5. Qualitative right ventricular (RV) size grade across time after RV sinus myectomy. Average size at presentation was -1.2 ± 3.2 . Format is as in Figure 4.

Figure 6. Standardized (Z value) pulmonary valve (PV) "annulus" diameter across time after right ventricular sinus myectomy. Average size at presentation was -0.51 ± 1.31 . Format is as in Figure 4.

Comparison

In this series, patients undergoing RV sinus myectomy all had an infundibular chamber and fell into the upper end of the spectrum of RV hypoplasia. They had correspondingly far less tricuspid valve hypoplasia (and therefore a larger inlet portion of the RV) than patients deemed suitable only for univentricular repair. Some patients undergoing sinus myectomy even overlapped at the upper end of the spectrum of RV and tricuspid valve size, with patients going directly to biventricular repair. Those undergoing RV sinus myectomy, however, achieved biventricular repair earlier in life than those having biventricular repair without RV sinus myectomy. This suggests that early surgical enlargement of the RV cavity is advantageous.

Biventricular Repair

Biventricular repair was achieved in nearly all patients managed by RV sinus myectomy, despite some having a small tricuspid valve, higher than reported in most large multi-institutional studies and close to the 50% suggested by the CHSS.^{9,14-16} Sano and associates¹⁷ used a similar decision-making approach in a cohort of 25 patients with either PAIVS ($n = 19$) or critical pulmonary stenosis ($n = 6$), all with a detectable infundibulum, undergoing initial palliation indicative of intended biventricular repair. Six had RV sinus myectomy; by age 36 months, 2 of these patients had achieved biventricular repair.

Others have suggested using the RV inflow, specifically tricuspid valve size, as the structure for biventricular repair decision-making rather than the presence of a RV infundibulum.⁹ However, the metric for expressing tricuspid valve size has not been uniform. For example, in a CHSS study, a tricuspid Z value of greater than -3 was associated with increased likelihood of safe biventricular repair,⁹ but the

CHSS standardization (Z values) of the tricuspid valve was referenced to an autopsy series,¹⁸ and it is known that autopsy specimens shrink. We have instead used the Daubney standardization of tricuspid valve size based on 2-dimensional echocardiography in infants and children.⁸ There is no simple conversion of Z values between these 2 methods of standardization. Therefore, we have provided additional metrics normalized to body surface area, percentage of mean normal tricuspid valve size, and centimeters per square meter.

Growth of Right-sided Structures

When initially proposed, RV sinus myectomy was thought not only to immediately increase RV capacity but also to promote growth of all right-sided cardiac structures.^{6,7} Although the pulmonary valve increased in size to near normal (after pulmonary valvotomy), tricuspid valve size after sinus myectomy did not accelerate toward normal but only increased in parallel with somatic growth. This left it small for body size, despite interventions aimed at increasing its size. We interpret the information from Mee's Melbourne series⁷ and that of Sano and associates¹⁷ as demonstrating the same disappointing phenomenon we have observed. Thus, tricuspid valve size appears to be a limiting factor that might affect long-term success of biventricular repair and, as such, should be considered in surgical decision-making concerning univentricular versus biventricular versus one and a half ventricle repair.

Survival

Although 85% 5-year survival after RV sinus myectomy is encouraging, the apparent inability of this procedure to stimulate tricuspid valve growth appears to have a detrimental effect when biventricular repair is attempted. In the 2 patients in our series who died 3 and 4 years after biventricular repair,

the Z value of the tricuspid valve remained less than -7 . Thus, tricuspid valve size at birth appears to have important negative implications regarding long-term success of biventricular repair. Severe tricuspid valve hypoplasia recognized at birth, even if a RV infundibulum is present, probably places patients into the univentricular or one and a half ventricle repair portion of the morphologic spectrum rather than the uncertain area.

Limitations

This is a single-institution, nonrandomized, clinical study. Unlike studies by the CHSS, it is not a birth inception cohort, but reflects referral bias. Furthermore, our protocol is biased by the prevailing surgical philosophy at our institution (and that of many programs), which is that biventricular repair is better in the long term than univentricular repair. A third bias was our assumption that RV sinus myectomy would accelerate growth of all right-sided structures to normal size.

In our study, we did not address morphologic subtypes of pulmonary atresia. At our institution, patients with membranous atresia and normal-sized and normally functioning RVs undergo radiofrequency perforation of the pulmonary valve to effect biventricular repair.¹⁹ These patients were not included in this study. We presume their right-sided structures were larger than those of patients presenting for surgical treatment.

Conclusions

RV sinus myectomy in PAIVS for morphology that falls within the uncertain area for eventual biventricular repair leads to immediate increase in RV volume. It, in combination with establishing RV–pulmonary trunk continuity, allowed early biventricular repair in 87% of patients. Thereafter, however, tricuspid valve growth in relation to somatic growth was minimal. Thus, although small tricuspid valve size did not deter early completion of biventricular repair after RV sinus myectomy, it might limit long-term success. The clinical decision-making inference is that the presence of an infundibular chamber should be only one of the morphologic criteria for selecting patients for RV sinus myectomy; in particular, size of the tricuspid valve is an important consideration. If the Daubeney Z value of the tricuspid valve annulus is less than about -5 , it is probably not prudent to pursue biventricular repair either with or without sinus myectomy. If the tricuspid valve annulus is -5 or greater, RV-dependent circulation is absent, and an infundibulum is present with an imperforate pulmonary valve, we recommend sinus myectomy as part of a strategy that can lead rapidly to biventricular repair.

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Appendix E1. Details of 43 surgical patients undergoing pulmonary atresia with intact ventricular septum

Of the 43 patients, biventricular repair was intended in 20 (47%), and univentricular repair was intended in 23 (53%; [Figure 2](#)).

Of the 20 patients for whom biventricular repair was intended, 13 (65%) underwent RV sinus myectomy, 5 with adequate RV size went directly to biventricular repair (25%), and 2 died before any other procedure (10%). Of the 13 patients undergoing RV sinus myectomy, definitive biventricular repair was achieved in 11 (85%), and 1 died before undergoing any other procedure (7.7%).

Of the 23 patients for whom univentricular repair was intended, 8 (35%) went on to completion Fontan, 1 (4.3%) went on to one and a half ventricle repair, 3 (13%) went on to RV sinus myectomy, 1 (4.3%) went on directly to biventricular repair, and 7 (33%) died before definitive repair; 3 (13%) were alive without definitive repair at last follow-up. Of the 3 patients who underwent RV sinus myectomy, 2 had definitive biventricular repair, and 1 had a one and a half ventricle repair.

Thus, of 43 patients, 19 (44%) achieved biventricular repair, 2 (4.7%) achieved a one and a half ventricle repair, and 8 (19%) achieved a univentricular repair.

Figure E1. Nonparametric competing-risks estimates of end states after the presentation described in Figure 2. A, Biventricular repair strategy. B, Univentricular repair strategy.