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ORIGINAL ARTICLE

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Achieving biventricular circulation in patients with moderate hypoplastic right ventricle in pulmonary atresia intact ventricular septum after transcatheter pulmonary valve perforation

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Abstract

Objective: Transcatheter valve perforation for pulmonary atresia intact ventricular septum is the standard of care for patients with mild right ventricular hypoplasia. However, its role in moderate right ventricular hypoplasia has been less well defined. We sought to report the long-term outcome of patients with moderate hypoplastic right ventricle who had undergone the procedure.

Design, Settings, and Patients: We performed a retrospective analysis on patients who had undergone transcatheter pulmonary valve perforation from January 1996 to January 2015 at our institution. The procedures would be carried out irrespective of the right ventricular size, as long as there were no absolute contraindications.

Intervention and Outcome Measures: Demographic and procedural data were correlated with outcome measures. Outcomes analyzed included procedural success, reintervention rates, final circulation type, and functional class. Multivariate analysis and receiver operator curve were used to identify for parameters in predicting biventricular circulation.

Results: The procedural success rate was 92% (33 out of 36) in this group with moderate right ventricular hypoplasia (tricuspid valve z score -4.2 ± 3.0, 69.4% of patients with z score <-2.5). Early reintervention rate was 39%, mostly being insertion of modified Blalock–Taussig shunt. Overall reintervention-free survival was 53%, 30%, and 19% at 1, 6, and 12 months postintervention. Despite no significant catch-up right ventricular growth, majority of survivors (84%) enjoyed a biventricular circulation with good functional status. A tricuspid to mitral valve ratio >0.79 was a good predictor of biventricular outcome. (specificity of 100%, positive predictive value 100%).

Conclusion: Encouraging long-term results with biventricular circulation and functional status were demonstrated with transcatheter pulmonary valve perforation in patients even with moderate hypoplastic right ventricle, which is comparable to that with mild right ventricular hypertrophy. The baseline tricuspid to mitral valve ratio was identified as a potentially useful tool in predicting biventricular circulation.

KEYWORDS
cardiac catheterization, laser, pulmonary atresia with intact ventricular septum, radiofrequency, right ventricular outflow tract, transcatheter pulmonary valve perforation

1 | INTRODUCTION

Pulmonary atresia with intact ventricular septum (PAIVS) is a rare congenital heart disease. The vast variability of right ventricular (RV) and tricuspid valve (TV) morphology has led to diverse management strategies.[1–3] Notwithstanding this diversity, early establishment of right ventricular outflow tract (RVOT) antegrade flow has been advocated for those without contraindication for RV decompression. Since 1990s, being first reported by Qureshi et al using laser energy[4] and subsequently radiofrequency (RF)[5] energy, transcatheter perforation of the atretic pulmonary valve has been widely adopted as the mainstay of treatment for PAIVS patients with mild RV hypoplasia.[6–10] However, the role of transcatheter valve perforation as the initial intervention for patients with moderate RV hypoplasia has been less clearly defined. This was probably the result of imposing an arbitrary cutoff TV annular *z* score, a surrogate for RV size, below which transcatheter management would not be offered. Moreover, there has also been a paucity of data on preintervention predictors for long-term biventricular circulation.[8–10]

We sought to report the long-term outcome of patients after transcatheter perforation of the atretic pulmonary valve as the first intervention in our institute, where an aggressive approach has been adopted. All PAIVS patients, including those with moderate RV hypoplasia, would be considered for transcatheter valve perforation unless there were absolute contraindications. Other than reporting on the procedural success rate, the most important outcome of interest would be the final type of circulation, the degree of catch-up RV growth with time, and the functional status of the patients. We also aimed at identifying predictors of final circulatory status.

2 | METHODS

We performed a retrospective analysis of patients with PAIVS who underwent an attempt of transcatheter pulmonary valve perforation in our center from January 1996 to January 2015. Only those patients who received the procedures as the intended initial interventions were included in the study. Patients who received a modified Blalock–Taussig shunt (mBT shunt) before the transcatheter procedures were excluded from the study. Laser energy was used in the early period from 1996 to 2005; thereafter, radiofrequency (RF) energy has become the preferred means. During the aforementioned period, all patients diagnosed with PAIVS, irrespective of the TV dimension and the body surface area-corrected *z* score, were considered for transcatheter valvotomy unless they met clinical exclusion criteria. These included, unipartite RV, RV dependent coronary circulation, muscular type of pulmonary atresia, or the presence of significant TV anomaly, eg, severe Ebstein anomaly, which precluded a biventricular circulation. Written informed consents were obtained from parents before the procedure.

Cardiac catheterization technique for valve perforation had been described in detail in previous publications.[4,5] In brief, all the procedures were carried out under general anesthesia. The femoral vein was percutaneously cannulated with a femoral sheath. A 4–5 Fr Judkin right (JR) catheter would be advanced to the RVOT, positioned just proximal to the atretic pulmonary valve. Once position was confirmed, a 0.018-inch laser guidewire (Advanced Interventional Systems Inc, Irvine, California), or a Nykanen RF wire (Baylis Medical Company Inc, Montreal, Canada), would be advanced via the JR catheter. Laser or RF energy would then be applied for valve perforation. This would be followed by serial balloon valvuloplasty, usually with small coronary balloons followed by larger balloons. Prostaglandin would then be stopped in the laboratory after the procedure unless saturation remained low (<70%). For patients with persistent desaturation below 70% despite receiving prostaglandin, a mBT shunt or surgical RVOT reconstruction would be considered.

During subsequent follow-up, decisions for further catheter or surgical interventions will be based on the clinical and echocardiographic data. Patient will be directed to the 1.5 ventricular pathway by means of a bidirectional Glenn shunt if persistent desaturation below 80% is observed at 6–12 months of age.

The medical records of all included patients were reviewed, with extraction of the following demographic and peri-procedural data: age, sex, body weight and length at the time of catheterization, comorbidity, and precatheterization echocardiographic parameters. Outcome measures extracted included procedural success, complications, mortality, reintervention, the final type of circulation, and New York Heart Association (NYHA) functional class. Complications were classified into acute catheter-related, in-hospital complication, and late complication. Acute catheter-related complications were defined as complications that were directly related to the procedure, which occurred in the catheterization laboratory or within 24 hours post-catheter intervention; all other complications that took place after the first 24 hours postprocedure and before first hospital discharge were defined as in-hospital complications, while all other complications happened after discharge were classified as late complications. Mortalities were classified as either early or late: early mortality being defined as mortality ≤30 days postprocedure and late mortality being beyond 30 days postprocedure. Reinterventions, transcatheter or surgical, were classified into early which took place ≤30 days, and late reinterventions that took place >30 days after the procedure.

Patients were classified into two circulation types: either biventricular or nonbiventricular circulation. Biventricular circulation was defined as having all the systemic venous blood returning

TABLE 1 Baseline characteristics at the time of intervention ($n = 36$)

Median age at initial catheterization (days) (IQR, range)	3 (IQR 4.75, 1–83)
Sex, male (%)	19(52.8%)
Body weight (kg)	3.11 ± 0.54
Bipartite RV	6/36 (16.7%)
Presence of RV coronary sinusoids (%)	8/36 (22.2%)
TV dimension *z* score	−4.2 ± 3.0
Number of patients with moderate RV hypoplasia[a]	7/36 (19.4%)
Number of patients with severe RV hypoplasia[a]	18/36 (50%)
TV/MV ratio	0.80 ± 0.16
PV dimension *z* score	−4.5 ± 2.5

Abbreviations: MV, mitral valve; PV, pulmonary valve; RV, right ventricular; TV, tricuspid valve.
[a]Moderate RV hypoplasia as defined by TV *z* score from −2.5 to −5; severe RV hypoplasia as defined by TV *z* score <−5.

normally to the right atrium and a SaO_2 of ≥95% in air in the absence of a surgical shunt. Nonbiventricular circulation would include the following patients with: (a) Fontan circulation, (b) bidirectional Glenn Shunt (the so-called "1.5 ventricular state"), and (c) SaO_2 constantly below 95% in air, presumably due to right to left shunt at the atrial level.

All the preintervention echocardiographic parameters extracted from patients' medical record and were verified by two of the authors (RHC and AKTC) after reviewing the index echocardiogram cine to ensure the measurements were obtained correctly. In our institution, the echocardiographic measurements and the *z* score calculations were based on the algorithms published by Daubeney et al.[11] In short, the mitral and tricuspid valve dimensions were obtained from the apical four-chamber view at early diastole, and the pulmonary valve dimensions were obtained at the short-axis view during systole, while all measurements were made from hinge point to hinge point. The RV would be defined as moderately hypoplastic if the TV *z* score fell in the range of −2.5 to −5; and severely hypoplastic should the TV *z* score measured <−5. The precatheterization echocardiogram and angiograms of the catheter procedures were reviewed together by two reviewers (RHC and AKTC) in order to determine the right ventricular morphology (tripartite vs. bipartite).

Follow-up echocardiographic data were extracted for those who had an echocardiogram performed within one year from the point of data collection on January 1, 2017. These parameters were reviewed and analyzed, mainly focusing on parameters pertaining to right heart size and function. In addition to the aforementioned parameters, the right atrial (RA) area *z* score was also calculated from the algorithm by Cantinotti et al.[12] The difference between baseline and the latest TV *z* score, which reflected the change of TV dimension over time, was also used to assess RV growth.

Statistical analysis was performed using SPSS 24 (IBM, Armonk, New York). Continuous variables were expressed as mean ± SD, with the exception of age at initial catheterization procedure which was expressed as median with interquartile range and range. Fisher's exact test was used to analyze binary variables, while two-sided *t* test or Mann-Whitney U test was used to analyze continuous variables in univariate analysis for the association with various outcome measures. Multivariate analysis was performed using the logistic regression model to assess for significant association with the final type of circulation. Receiver-operator curve (ROC) was constructed for baseline parameter to predict biventricular circulation, using the area under the curve to assess the performance and the Yonden index to identify the optimal cutoff point.

3 | RESULTS

From January 1996 to January 2015, 43 patients with PAIVS were brought to the cardiac catheterization laboratory of our center with the intention to perforate the atretic pulmonary valve by transcatheter means. Thirty-six patients (84%) who received the procedure as the initial intervention were included in the study. Seven patients who received a modified Blalock-Taussig shunt (mBT shunt) before the transcatheter procedures were excluded. Baseline characteristics of the 36 patients were shown in Table 1. The mean baseline TV annular *z* score was −4.2 ± 3.0. Twenty-five out of 36 patients (69.4%) had a TV annular *z* score <−2.5, among which 7 out of 36 (19.4%) patients had moderately hypoplastic RVs and 18 out of 36 (50%) had severely hypoplastic RVs. Four patients had TV anomaly, including 3 with mild tricuspid stenosis and 1 with mild Ebstein anomaly. There were no other associated intracardiac lesions in our cohort of patients.

The acute and long-term outcome of the patients was summarized in Figure 1. Acute procedural success was achieved in 33 out of 36 procedures (92%). Acute catheter-related complications occurred in 8 out of 36 patients (22.2%). Three patients (8.3%) suffered from major catheter-related complications. Among them, 2 patients had RVOT perforation resulting in cardiac tamponade, 1 from the laser energy and the other from the RF group. Unfortunately, both patients succumbed during surgical exploration in the operation theatre. One other patient suffered from loss of lower limb pulse postcatheterization that resolved after a short course of systemic heparinization. Five patients (13.8%) had minor complications, all of whom had transient supraventricular tachycardia during the procedure that spontaneously subsided. There were no other catheter-related acute or in-hospital complications.

In terms of mortality, there were 5 early mortality recorded (13.9%), 3 of which took place within 24 hours post procedure. Among the 24-hour mortality, 2 of them were the aforementioned patients who suffered from the RVOT perforation during the catheter procedure. One patient had circulatory collapse secondary to a restrictive patent foramen ovale while awaiting for surgical palliation after an unsuccessful catheter procedure. He succumbed 19 hours after the catheter procedure despite an emergent balloon atrial septostomy and resuscitation. This was not classified as a procedure-related complication. The other two early mortality occurred after successful transcatheter pulmonary

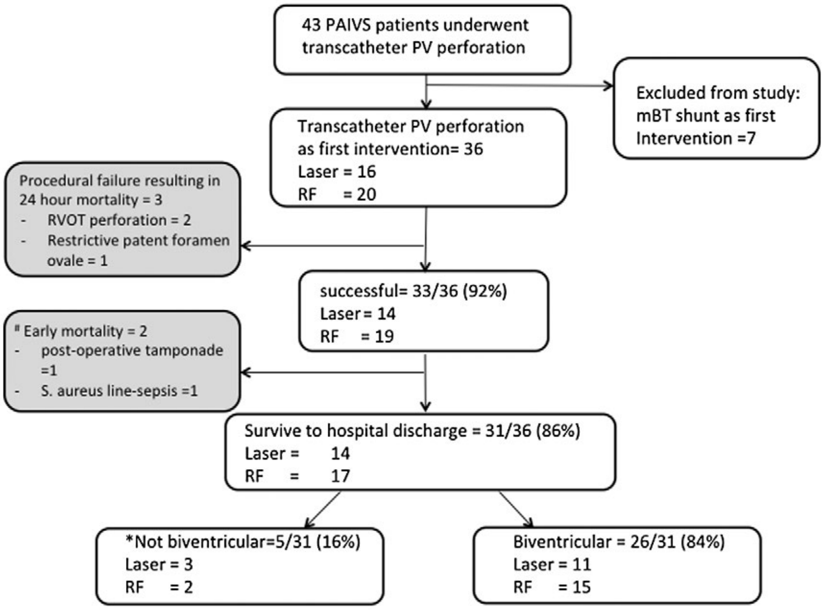

FIGURE 1 Summary of short- and long-term outcomes of patients. Abbreviations: RF, radiofrequency; RVOT right ventricular outflow tract.

*1.5 ventricular repair = 2; Fontan = 1; patients with SaO2 <95% = 4. #Mortality classification: early mortality = mortality took place ≤ 30 days postcatheterization; late mortality = mortality took place >30 days postcatheterization (there were no late mortality).

valve perforation, both unrelated to the catheter procedure. One patient suffered from cardiac tamponade after early reintervention with surgical mBT shunt insertion and atrial septal defect augmentation. The other patient died of uncontrolled line-related *Staphylococcus aureus* septicaemia. There was no late mortality.

Reinterventions were commonly required in our cohort. The overall reintervention-free survival was 53%, 30%, and 19% at 1, 6, and 12 months post–catheter intervention, respectively. Early reinterventions were required in 13 patients (39%). These included 10 mBT shunt insertion, 2 balloon atrioseptostomy, 3 PDA ligation, and 1 percutaneous balloon pulmonary valvuloplasty (PBPV). The use of laser-assisted valve perforation, as compared with RF energy, was the only factor identified to be associated with early reintervention (P = .04). However, the subgroup analysis comparing the two groups using different energy did not show significant differences in terms of the preprocedural or procedural data. For patients requiring a mBT shunt, the only factor showing significant association was a lower pulmonary annular z score (−5.7 ± 2.9 vs −3.8 ± 2, P = .03). There was a tendency of a lower TV z score (−5.4 ± 3 vs −3.6 ± 2.9) and a higher postprocedural PS gradient (36 ± 20 mm Hg vs 25 ± 15 mm Hg) but both did not reach statistical significance.

As at January 1, 2017 when the outcome data were extracted, 31 surviving patients were being reviewed regularly as outpatients. The median follow-up duration was 10.7 years (ranged from 2.3 to 19.8 years). Altogether 45 late reinterventions were required in 26 of the 31 long-term survivors, among them 17 patients required 2 or more procedures. Figure 2 showed the breakdown of the late reinterventions involved. The only factor associated with late reinterventions was a higher residual RVOT gradient from echocardiogram after the initial catheter intervention. (29.1 ± 24.3 mm Hg vs 14.7 ± 6.9 mm Hg; P = .013) Patients with lower pulmonary valve dimension z score (−4.7 ± 2.6 vs −3.3 ± 1.2) had a tendency toward reintervention, though it did not reach statistical significance. All other parameters showed no significant association.

In terms of the circulatory status, 26 out of the 31 survivors (84%) enjoyed a biventricular circulation. Sixteen out of these 26 patients (62%) had moderate or severe RV hypoplasia at baseline. Among these 16 patients, 3 had achieved biventricular circulation without reintervention. Of the 5 patients who had nonbiventricular circulation (5/31, 16%), 2 patients had a 1.5-ventricular circulation and 1 patient underwent a Fontan operation. Among the 20 survivors with moderate RV hypoplasia, 80% (16/20) had achieved biventricular circulation.

Table 2 shows the relationship of baseline patient characteristics and the latest circulatory status. In the biventricular circulation group, the mean TV z score was −3.34 (±2.68) with a range of −9 to +2.49. The wide range of z score is mainly due to two outliers with TV z score of −9, while all others have z score>−6.5. The two outliers had attained biventricular circulation after undergoing multiple

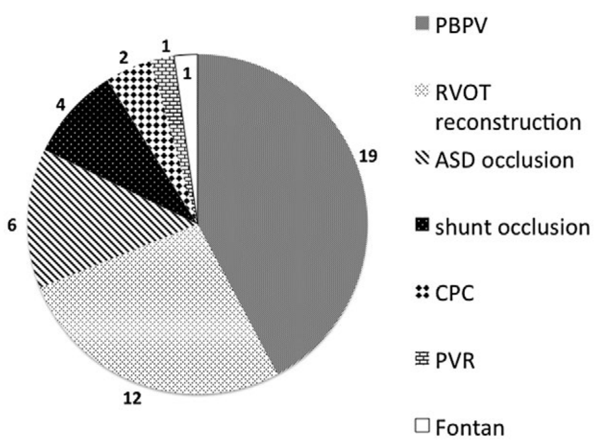

FIGURE 2 Types of late reinterventions. Abbreviations: ASD, atrial septal defect; CPC, cavopulmonary connection; PBPV, percutaneous balloon pulmonary valvuloplasty; PVR, pulmomary valve replacement; RVOT, right ventricular outflow tract

TABLE 2 Baseline patient characteristics and the latest circulatory status

	Biventricular state (26/31, 84%)	Nonbiventricular state (5/31, 16%)	*P* value
Median age at initial catheterization (days) (IQR, range)	5 (IQR 23.5, 1-83)	4 (IQR 25, 2-51)	NS
Weight at procedure (kg)	3.21 ± 0.55	2.80 ± 0.31	NS
TV *z* score	−3.34 ± 2.68	−6.86 ± 3.29	.028
TV/MV ratio	0.86 ± 0.15	0.67 ± 0.14	.017[a]
TV *z* score <−2.5	16/26 (62%)	4/5 (80%)	NS
PV *z* score	−4.21 ± 2.30	−5.59 ± 2.70	NS
Balloon to PV dimension ratio	1.29 ± 0.22	1.39 ± 0.46	NS
Require ASD augmentation	1/26 (3.8%)	2/5 (40%)	.012
Bipartite RV	2/26 (7.7%)	2/5 (40%)	.048

Abbreviations: ASD, atrial septal defect; MV, mitral valve; PV, pulmonary valve; RV, right ventricle; TV, tricuspid valve.
[a]A higher TV/MV ratio is the only significant factor identified with logistic regression model being associated with biventricular circulatory status.

reinterventions. One of them received 2 PBPV, a surgical RVOT reconstruction followed by transcatheter ASD occlusion, while the other patient had required 2 PBPV. Comparing the patient groups with different circulatory status, on univariate analysis, a higher initial TV *z* score, and a higher baseline TV/MV ratio were associated with a biventricular circulation. Bipartite RV and history of balloon atrioseptostomy were associated with a nonbiventricular circulation. Other parameters did not predict the final type of circulation. However, only the baseline TV/MV ratio was significantly associated with the final type of circulation in the logistic regression analysis. ROC demonstrated a baseline TV/MV ratio above 0.79 to be a tool to predict a biventricular circulation with satisfactory performance, with specificity and positive predictive value both being 100% (Figure 3).

Twenty-six patients had an echocardiogram done within one year from the point of data collection, and the results were shown in Table 3. There was only a small degree of catch-up growth of the RV across the whole cohort as reflected by the small increment of the mean TV *z* score over time (0.7 ± 2.80). The increment, however, had a tendency of being larger in the nonbiventricular group as compared with the biventricular group (1.4 ± 2.3 vs 0.4 ± 3.0), though this did not reach statistical significance. RV hypoplasia was still evident in the cohort, though it was more significant in the nonbiventricular group. Right atrial dilatation, restrictive RV physiology, and presence of significant TR and PR were also prevalent in the study group.

At the time of data collection, late complications were observed in 2 patients. One patient with 1.5 ventricular circulation suffers from atrial tachycardia and left ventricular systolic dysfunction and was in NYHA class II. The other patient was wheelchair-bound due to spastic quadriplegia as a result of stroke during postoperative period for RVOT reconstruction at 6 months of age. All patients except the 2 aforementioned were in NYHA class I. There were otherwise no symptomatic arrhythmia or other late complications reported.

4 | DISCUSSION

Our study represented a series with one of the longest postprocedural follow-up reported for patients with PAIVS managed with transcatheter valve perforation, with a median follow-up of 10.7 years.[6–10,13–15] Similar to two other studies, our population comprised of patients with moderate RV hypoplasia as reflected by the overall low TV *z* score, and close to 70% of patients having a TV *z* score <−2.5.[13,15] This was probably as a result of the policy in not imposing a lower limit of TV *z* score, a surrogate of RV dimension, below which a transcatheter valve perforation was not considered. This aggressive approach was a reflection of the belief in the merits of early RV decompression,[16–18] with the understanding that some patients might not sustain a long-term biventricular circulation. This approach on one hand allows the RV a chance to be decompressed and grow to support an eventual biventricular circulation; yet on the other hand, it also does not refute the possibility of switching to a univerntricular repair at a later stage if indicated. Notwithstanding the majority of patients falling into the group of moderate to severe RV hypoplasia in our cohort, we managed to demonstrate a high proportion of patients, 84% of all the survivors, enjoying biventricular circulation at the latest follow-up. Furthermore, among survivors with moderate or severe RV hypoplasia at baseline, up to 80% had achieved biventricular circulation which is comparable to that reported in series with mild RV hypoplasia.[6–9,14] Intriguingly, 2 of our patients with TV *z* score as low as −9 at baseline were able to attain biventricular circulation, albeit that both patients had required multiple reinterventions. The majority of the long-term survivors were capable of maintaining a good functional status with more than 90% of patients falling in NYHA class I. These favorable long-term outcome perhaps suggest that including patients with moderate RV hypoplasia for transcatheter valve perforation is a reasonable approach for patients with PAIVS.

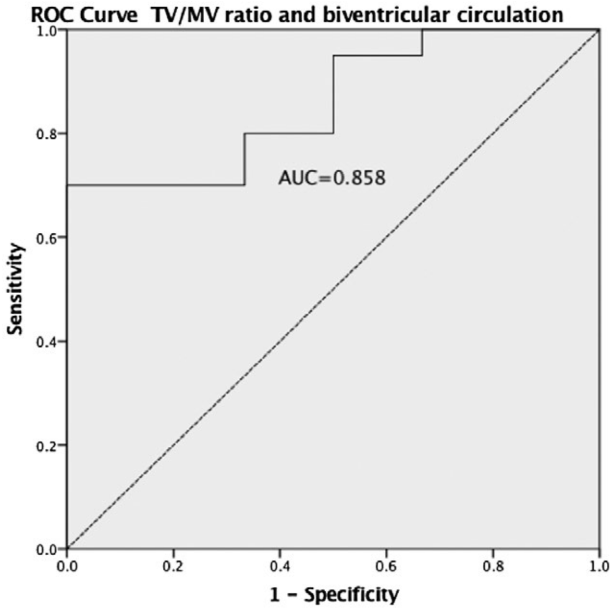

FIGURE 3 Receiver–operator curve for tricuspid to mitral valve (TV/MV) ratio in predicting biventricular circulation. Using the Yonden Index, the optimal cutoff TV/MV ratio identified was 0.79 (specificity 100%, sensitivity 70%, Positive predicted value 100%, negative predictive value 50%)

More intriguingly, the aforementioned satisfactory long-term functional outcome was achieved without demonstrating statistically significant catch-up RV growth, as reflected by the small increment of TV *z* score over time. Controversies still exist as in whether significant RV catch-up growth would be anticipated after restoration of RVOT patency with either transcatheter or surgical means.[8,15–19] However, similar to our findings, reports have demonstrated reasonable medium-term outcome especially pertaining to attainment of biventricular circulation despite the lack of catch-up RV growth.[8,15]

Several factors have been reported to be associated with a nonbiventricular outcome, including a smaller initial PV or TV annular *z* score, smaller TV/MV ratio, lower oxygen saturation at 1 year of age, presence of RV sinusoids, and significant associated TV anomaly.[8–10,15,20] Unfortunately, none of these factors were good predictors of final type of circulation. Moreover, most of these reported factors had limited clinical applicability as they took into account composite adverse outcomes, rather than specifically addressing the issue of final circulation outcome. Minich et al had reported the potential role of TV/MV annular dimension ratio for predicting circulation outcome.[21] However, the suggested cutoff of 0.5, above which a biventricular outcome is predicted, would have included a quite significant proportion of patients with nonbiventricular outcome leading to poor specificity. From our series, we have demonstrated a TV/MV annular dimension ratio of more than 0.79 could confidently predict a biventricular circulation outcome. (specificity and positive predicted value of 100%) This can potentially be useful in predicting patients with biventricular outcome, especially for patients conventionally classified as moderate RV hypoplasia with TV *z* score <−2.5.

TABLE 3 Latest echocardiographic data for different circulatory status

	All patients (*N* = 26)	Biventricular state (21/26)	Nonbiventricular (5/26)	*P* value
RA area *z* score	2.3 ± 1.6	2.1 ± 1.6	2.7 ± 1.8	NS
RV length *z* score	−3.4 ± 1.3	−3.1 ± 1.5	−4.4 ± 1.7	.05
RV area *z* score	−3.0 ± 2.3	−2.2 ± 1.8	−5.8 ± 1.7	.03
TV *z* score	−3.4 ± 2.1	−2.9 ± 1.9	−5.3 ± 2.1	.04
TV to MV ratio	0.93 ± 0.21	1.44 ± 2.06	0.74 ± 0.23	NS
ΔTricuspid valve annular dimension *z* score	0.7 ± 2.8	0.4 ± 3.0	1.4 ± 2.3	NS
PS PG (mm Hg)	19.5 ± 17.4	17.9 ± 17.0	15.3 ± 19.3	NS
TR PG (mm Hg)	31.2 ± 9.8	30.8 ± 9.36	29.5 ± 12.3	NS
TAPSE *z* score	−0.1 ± 2.9	0.04 ± 3.12	−2.99 ± 2.25	.013
RV area change (%)	43.6 ± 12.3	44.1 ± 11.8	39.1 ± 13.9	NS
Restrictive RV physiology[a]	18/26(69.2%)	14/21 (66.7%)	4/5 (80%)	NS
Significant tricuspid regurgitation[b]	17/26 (65.3%)	13/21 (62%)	4/5 (80%)	NS
Significant pulmonary regurgitation[b]	25/26 (96%)	20/21 (95.2%)	5/5 (100%)	NS

Abbreviations: PS PG, pulmonary stenosis Doppler-derived pressure gradient; PV, pulmonary valve; RA, right atrium; RV, right ventricular; TR PG, tricuspid regurgitation jet Doppler-derived pressure gradient; TV, tricuspid valve.
ΔTricuspid valve annular dimension *z* score: change of tricuspid valve *z* score = the latest tricuspid valve annular dimension *z* score minus that measured at the baseline preintervention echocardiogram.
[a]Restrictive physiology defined as detection of diastolic forward flow by Doppler study in pulmonary artery.
[b]Qualitative assessment of valvar regurgitation being equal or worse than moderate degree was regarded as significant.

The long-term functional outcome also appeared to be encouraging from our results, with all except 2 survivors enjoying NYHA functional class I status. It was surprising, however, with prevalent RA dilatation in our group of patients, there was only 1 patient with atrial arrhythmia. Despite this low incidence, it is still prudent to monitor for the development of atrial arrhythmia as patients go into adulthood. The loss of sinus rhythm and hence the "atrial kick" may adversely affect the cardiac output in this group of patient with prevalent restrictive RV physiology.

The rate of early reinterventions reported in various case series ranged from 30% to 47%, mostly comprised of augmentation of pulmonary blood flow in the form of a mBT shunt or ductal stenting as a separate procedure.[6,8–10] This is not unanticipated, as the RV compliance often remained poor immediately postprocedure despite a widely patent RVOT and pulmonary valve.[6,8,22] It's intriguing, however, with comparatively moderate-severe hypoplastic RVs in our series, that the rate of requiring an additional pulmonary blood flow was only 30%, which was similar to that reported in series including only mild RV hypoplasia. Multiple factors have been reported to be associated with early reintervention, namely having a bipartite RV,[9] a lower TV annular *z* score,[7–10,22,23] presence of RV-coronary communication,[24] and a higher baseline RV systolic pressure.[8] From our series, the only factor associated with early reintervention was the use of laser energy, as compared with RF energy. However, on subgroup analysis, there were no significant differences in terms of patient or procedural data comparing the two groups, hence we believe the intergroup differences were probably a result of an era effect. During our early experience, while laser energy was used, physicians might have a lower threshold in going for a "rescue" mBT shunt immediately after catheterization if oxygen saturation remained low. This was in contrast with our more recent approach, in which a period of watchful waiting would be allowed by maintaining ductal patency with prostaglandin. As alluded to earlier on, with the anticipation of a very restrictive RV immediately after establishing RVOT patency, we now believe time should be allowed for the ventricle to adapt before deciding on the need of further palliation.

Pertaining to strategies to mitigate the need of early reintervention, simultaneous duct stenting has been reported as an attractive strategy for patients with moderate RV hypoplasia or the so-called borderline RVs in providing additional source of pulmonary blood flow during the procedure of transcatheter valve perforation.[13,15,25,26] However, the decision for ductal stenting was largely under the discretion of individual operators in some of these studies. On the other hand, some published treatment algorithm for patients with PAIVS recommended ductal stenting be included as part of the primary procedure should patient fall into a group of intermediate or severe hypoplastic RV defined by a cutoff of TV annular *z* score <-2.5.[1] Notwithstanding the possible advantage of ductal stenting, there has been a lack of clear-cut parameter in determining the necessity of ductal stenting as a primary intervention at the same occasion of valve perforation. Taking our cohort as an example, should the aforementioned algorithm be applied, 13 out of the 36 patients (36%) who did not require early intervention would have received an unnecessary ductal stent to provide an additional pulmonary blood source. Our results with a cohort of predominant moderate RV hypoplasia hence supported our current strategy of watchful waiting regarding the need of augmentation of pulmonary blood flow. We believed that many patients are capable of "growing out" of the restrictive RV physiology with time without the need for early pulmonary blood flow augmentation.

There are several limitations to our study. Being a retrospective study, the treatment decisions made for individual patients were not per protocol but rather being dependent on individual physician's discretion. This might be especially true pertaining to the decision for an early "rescue" mBT shunt versus watchful waiting with prostaglandin during the earlier era. Important changes of practice during the period would be another potential source of bias, most notably the change from laser energy to RF energy for valve perforation. Indeed, the use of laser energy was identified as a risk of early reintervention. However, as alluded to earlier on, there were no significant differences in terms of patient or procedural data comparing the two groups, hence we believe the intergroup differences, if any, should represent mainly an era effect while we were at different stages of the learning curve. Finally, despite having reported reasonable late outcome in terms of biventricular survival, morbidity, and NYHA functional class, only a small number of patients had undergone formal cardiopulmonary function tests for objective assessment. We shall have a better understanding of the cohort of patients when these data build up.

5 | CONCLUSION

Our study has demonstrated a satisfactory acute success and an encouraging long-term outcome with transcatheter pulmonary valve perforation in PAIVS patients even with moderate hypoplastic RV. Majority of these patients could attain biventricular circulation with a good functional status. The results reaffirmed our approach in not excluding patients from transcatheter valve perforation based solely on an arbitrary cutoff of moderate negative TV annular *z* score. The baseline TV/MV ratio was identified as a potentially useful tool in predicting a biventricular circulation for patients conventionally classified as moderate RV hypoplasia based on TV *z* score alone.

CONFLICTS OF INTEREST

None.

AUTHOR CONTRIBUTIONS

All authors read and approved the final version of the manuscript

Conceptualized the study, acquisition of data and performed the initial analysis, drafted the manuscript: Chen

Conceptualized the study, critically reviewed and revised the manuscript: Chau, Chow, Cheung, Yung, Lun

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