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Pulmonary Atresia with Intact Interventricular Septum

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Abstract

Background: Pulmonary atresia-intact interventricular septum (PA-IVS) is a rare congenital heart disease. To improve cardiac circulation, the goal now is to carefully select patients to achieve a biventricular repair via Transcatheter Radiofrequency- Assisted Pulmonary Valvotomy and Balloon Valvuloplasty (TRFAPV-BV) and to avoid early open heart surgery. Success rates of >80% have been reported. Due to its rarity, no single institution can provide a consistent interventional guideline. There is much to be learned from a multicentered approach to collecting longitudinal experience to a challenging clinical case.

Objective: We report our procedural and short-term outcomes in our patients who have undergone TRFAPV-BV with an emphasis on possibly identifying the predictors for survival and the need for additional transcatheter Right Ventricular Outflow Tract (RVOT) reintervention.

Methodology: This is a retrospective, descriptive, cohort study of all patients with PA-IVS who underwent TRFAPV-BV from Decemeber 2013 to April 2016. The hospital medical records of each patient was reviewed. Transthoracic two dimensional echocardiogram reports and clips, and cardiac catheterization reports pre and post-TRFAPV-BV were reviewed. In addition, each patient's clinical course through medical records were examined until their most recent clinical follow-up.

Results: There are 29 pediatric patients diagnosed with PA-IVS between December 2013 to April 2016. Out of these 29 patients with PA-IVS, 9 pediatric patients had a tripartite right ventricle underwent TRFAPV-BV, in whom 8 were described as successful, 89% (8/9). This study observed that the following paremeters had a favorable outcome: Tricuspid Valve (TV) annulus z-score > -2.5, tripartite right ventricle, absence of ventricular to coronary connections, tricuspid to mitral valve ratio.

Keywords: Pulmonary; Atresia; Intact interventricular septum; Radiofrequency; Assisted pulmonary valvotomy

Abbreviations

ASD (Atrial Septal Defect); HIE (Hypoxemic Ischemic Encephalopathy);PA-IVS (Pulmonary Atresia-Intact Interventricular Septum); PDA(Patent Ductus Arteriosus);PFO (Patent Foramen Ovale);PPBV (Percutaneous Pulmonary Balloon Valvuloplasty); RVOT (Right Ventricular Outflow Tract);RV (Right Ventricle);TRFAPV-BV (Transcatheter Radiofrequency-Assisted Pulmonary Valvotomy and Balloon Valvuloplasty);TV (Tricuspid Valve); UP-PGH (University of the Philippines-Philippine General Hospital)

Introduction

In Pulmonary Atresia (PA) and Intact Inter Ventricular Septum (IVS), there is an imperforate Right Ventricular Outflow Tract (RVOT), which can be either membranous or represented by a longer segment of muscular atresia. The ventricular septum functionally is intact. The pulmonary blood flow is being supplied by the patent ductus arteriosus. In rare cases, major aortoplulmonary collateral arteries originating from the descending thoracic aorta are the source of the lungs [1].

It has an incidence rate of 0.6/10,000 live births.There is no known sex predisposition, no identified genetic tendency, whereas familial cases have been seen in monozygotic twins.4 In the Philippines, there is no national database available to provide the number of cases per population at risk of this congenital heart disease. In our institution, from 2006 to 2015, we had a total of 125 diagnosed cases in 10 years. The incidence cannot be determined because most of our patients are not born in our institution [2].

Maturational arrest is the cause that probably explains the defect leading to an imperforate "tricuspid" pulmonary valve whose commissures are completely fused and a nearly normal-sized right ventricle. Abnormalities in morphology of the pulmonary valve can result from failure of normal development of the valve itself.

Presence or absence of ventriculocoronary connections and coronary artery abnormalities associated with poor outcomes is an important prognostic feature of this defect. Identification of this abnormality is an absolute contraindication to decompressing the right ventricle by surgery or transcatheter methods [3]. The coronary abnormality can be detected by

echocardiography but angiography is required because ventriculocoronary connections have been observed even in patients with a normal-sized right ventricle not detected in echocardiography [4].

To improve cardiac circulation, the goal now is to carefully select patients to achieve a biventricular or so-called one-and-one-half ventricle repair and to avoid the single ventricle repair or total cavopulmonary (Fontan) circulation. Historically, surgical pulmonary valvotomy under cardiopulmonary bypass with or without a systemic-to-pulmonary artery shunt was the initial procedure in the neonatal period to achieve biventricular circulation. However, the mortality rates were high in this surgical procedure. Over the last decade, an alternative procedure of Transcatheter Radiofrequency-Assisted Pulmonary Valvotomy and Balloon Valvuloplasty (TRFAPV- BV) was more preferred. A favorable anatomy is a tripartite right ventricle of near normal size with valvar pulmonary atresia and a well-developed pulmonary arterial circulation [5].

The success of the radiofrequency procedure has been established in >80% of the patients reported. It may avoid or delay the use of cardiopulmonary bypass in the newborn period; however, the long-term morbidity or mortality is largely unknown. There are still discussions whether the right ventricle and the outflow tract grows over time. In reviewing the literature, it has to be observed whether there is an absolute increase in the size of an anatomic structure from that of the indexed size. Nevertheless, the indexed Z value does not appear to change significantly.

In a non-compliant ventricular chamber, the right ventriclular pressure is at systemic levels or higher, and the end-diastolic pressure may be unusually elevated. In patients with a subsystemic right ventricular pressure, the right-to-left ventricular pressure ratio is <1. This is congruent with a globally disadvantaged right ventricle [6]. It is often associated with a thinned ventricle, and a severe tricuspid regurgitation.

The right ventricle can enlarge if it is adequately decompressed. Still, no single institution has a sufficient number of treated patients in recent times to ascertain this therapeutic recommendation.

In maintaining systemic to pulmonary arterial blood flow, the ductus arteriosus is kept open with prostaglandin infusion. The question now is the timing when to create a systemic-to-pulmonary artery shunt, or implant a stent in the ductus arteriosus after radiofrequency valvotomy. This occurs in 33% to 58% of patients. With a relatively small number of patients affected by this condition, there is much to be learned from a multicentered approach to collecting a longitudinal experience to a challenging clinical problem. The long-term advantage of this circulation remains to be seen [7].

Materials and Methods

In the largest series reported to date and adds to the growing literature regarding the outcomes after catheter-based interventions in neonates with PA-IVS by Schwartz et al in 2013, from January 2000 to July 2011, they had 23 patients with PA-IVS who underwent radiofrequency valvotomy. Studies showed that gradient across the pulmonary valve (9.9mmHg +/- 8.4 *vs.* 19.1mmHg +/- 10.4, p=0.05) after TRFAPV-BV may be predictive of the need for subsequent RVOT reintervention. Tricuspid valve hypoplasia (<-0.7) might be predictive of the need for an additional source of pulmonary blood flow.16 Risk factors for the need for subsequent reintervention after TRFAPV-BV are incompletely defined in this patient population [8].

In 2000, Minich et al had proposed using the ratio of tricuspid valve annulus to mitral valve annulus diameters as a predictor of late outcome. The ratio is helpful in determining right ventricle hypoplasia and therefore is also helpful in determining single versus biventricular repair. A tricuspid/mitral ratio >0.5 has been shown to be an excellent predictor of successful biventricular repair in PA-IVS patients [9].

Satou et al in 2000 correlated Tricuspid Valve (TV) diameter z-score with right ventricle size and has long been a standard in helping to determine whether a patient is a candidate for single versus biventricular repair. A TV z-score less than -3 correlates with the presence of coronary sinusoids and TV z-score greater than -3 has been associated with successful biventricular repair. 2,TVscore of less than - 2.5 predicts poor clinical outcomes [10].

TV Z-scores greater than -2.5 have membranous atresia and are usually the tripartite right ventricle type. They have no major sinusoids and with variable tricuspid regurgitation. The initial treatment is TRFAPV-BV and the long- term prognosis is usually good. Balloon dilation of pulmonary valve restenosis, RVOT reconstruction or tricuspid valve repair may occasionally be required.

As to duration of prostaglandin infusion, that it could not be discontinued in approximately 1-2 weeks. If pulmonary valve annulus is inadequate, several experts advise the placement of a Blalock-Taussig shunt or stenting of the ductus arteriosus. According to authors' experience, patient may be observed as long as 4-6 weeks for the oxygen saturation to improve back to satisfactory levels. Low systemic saturation, even after 4-6 weeks, will warrant a re- evaluation of adequacy of pulmonary blood flow. The choices are to re- catheterization to re-dilate the pulmonary valve or create a systemic to pulmonary arterial shunt by stenting the ductus arteriosus or surgical Blalock-Taussig shunt [11].

Our experience in the Philippines started in December 2013 at the University of the Philippines-Philippine General Hospital (UP-PGH) and we report our procedural and short-term outcomes in patients who underwent TRFAPV-BV with an emphasis on possibly identifying the predictors for survival and the need for additional transcatheter RVOT reintervention.

Outcome measures include the following:

1. Baseline and procedural characteristics
2. Technical success rate: Successful perforation and balloon dilatation of the atretic pulmonary valve with reduction in right ventricular systolic pressure
3. Per procedural complications

4. Changes in hemodynamic measurements (oxygen saturations and pressures)
5. Clinical course
6. Characteristics between patients who did or did not require RVOT reintervention after TRFAPV-BV.

This is a retrospective, descriptive, cohort study wherein all patients with PA-IVS who underwent TRFAPV-BV from December 2013 to April 2016 were included in the study. The medical records of each patient were reviewed, and relevant demographic characteristics such weight (kg), height (cm), gestational age (weeks), age at TRFAPV-BV (days), sex, comorbidities, and ECG abnormalities of the ST-T segments were extracted [12].

The transthoracic two dimensional echocardiogram reports and clips was reviewed, and the following measurements were extracted: Tricuspid valve annulus in apical four-chamber view (mm), Tricuspid valve annulus z-score, Tricuspid to Mitral valve ratio, Tricuspid regurgitation (mmHg), Pulmonary valve annulus (mm), and Pulmonary valve annulus z-score.

Cardiac catheterization reports were reviewed such as maximum balloon diameter (mm), maximum balloon diameter/ pulmonary valve annulus and right ventricular angiograms to assess presence of right ventricle to coronary artery connections, RVOT and pulmonary valve. Pre TRFAPV-BV Right Ventricle (RV) systolic pressure (mmHg) and RV systolic pressure/aortic systolic pressure ratio were recorded. Changes in hemodynamic measurements pre and post-TRFAPV-BV such as oxygen saturations and pressures RV systolic pressure/aortic systolic pressure ratio and gradient across pulmonary valve (mmHg)] were recorded [13].

In addition, patient's clinical course through medical records were examined to document technical success rate, per procedural complications, mortality, other cardiac procedures done, pre-TRFAPV-BV oxygen saturations on room air, duration of prostaglandin infusion post-TRFAPV-BV, post-TRFAPV-BV oxygen saturations upon discharge, procedures after TRFAPV-BV done, post-intervention follow up (months), oxygen saturations on follow up (%), direction of atrial level shunting and circulation type at most recent clinical follow-up [14].

For patients who underwent RVOT reintervention after TRFAPV-BV, demographic characteristics such weight (kg), height (cm), age at reintervention, timing of reintervention, oxygen saturations, the same transthoracic two-dimensional echocardiogram and pre and post RVOT reintervention pressure measurements [RV systolic pressure (mmHg), RV systolic pressure/aortic systolic pressure ratio and gradient across pulmonary valve (mmHg)] were extracted.

Categorical data were described as frequencies and percentages while Continuous variables were summarized as means, standard deviations and median. T-test, Chi-square or Fisher's exact test whenever is applicable was used to test for the statistical significance of the variables grouped according to patients who did or did not require RVOT reintervention after TRFAPV-BV. P value less than or equal to 0.05 was considered significant. The collected data collected were encoded in Microsoft excel. Checking for inconsistencies and completeness of the data was done prior to analysis using Stata statistical software [15].

The study was submitted to University of the Philippines Manila Research Ethics Board (UPMREB) panel prior to the start of the study and was conducted only upon approval.

Results

There were 29 pediatric patients diagnosed with PA-IVS between December 2013 to April 2016. Out of these 29 patients with PA-IVS, 9 pediatric patients had a tripartite right ventricle and all (100%) underwent TRFAPV-BV procedures. The range of age of patients at the time of the procedure was at 1 day old to 8 years (median 6 days) and the weight was at 1.97 kg to 23.5 kg (median 3.41 kg). Five (56%) of which were males and eight (89%) were born fullterm. One patient was born pre-term at 33 weeks and received surfactant for lung prematurity. The baby was kept on prostaglandin for 2 weeks until the baby weighed almost 2 kg for the procedure. Another patient was born to mother with gestational diabetes and was diagnosed to have biventricular hypertrophy on ECG and 2D-echocardiogram [16].

Initial echocardiographic measurements of the patients showed mean tricuspid valve annulus z-score, mean tricuspid to mitral valve ratio of 1 [range 0.62 to 1.34], mean tricuspid regurgitation was 128.67 mmHg [range 80 to 260 mmHg], and the mean pulmonary valve annulus z-score was -2.52 [range -3.58 to -0.38]. Pre-TRFAPV-BV echocardiogram and angiogram showed all patients had a tripartite right ventricle and none had right ventricle-to-coronary connection.

In the cardiac catheterization measurements, the mean right ventricular systolic pressure/aortic systolic pressure ratio was 1.32 [range 1 to 1.9] and the mean maximum balloon diameter/ pulmonary valve annulus was 1.4 [range 1.14 to 1.6]. In the post-TRFAPV-BV measurements, the mean right ventricular systolic/ aortic pressure ratio decreased to 0.53 [range 0.47 to 0.60] and the mean gradient across the pulmonary valve was 28.63 mmHg [range 5 to 60 mmHg] [17].

Out of 9 patients, we had successfully performed TRFAPV-BV in 8 patients. This gave us a technical success rate of 89% in perforating and dilating the atretic pulmonary valve with a decrease in the RV pressures. The unsuccessful attempt was in a one month old 4.1 kg full-term infant with a tricuspid valve annulus Z score of 0.32, tricuspid to mitral valve ratio of 1 and pulmonary annulus z score of 0.38. There was perforation of the RVOT, which required immediate pericardiocentesis. Balloon atrial septostomy and PDA stenting were performed during stabilization. The baby went into ventricular tachycardia leading to its demise from severe metabolic acidosis.

Another patient who had periprocedural complication during TRFAPV-BV is a case of a 14 day old 3.7 kg fullterm infant who went into bradycardia from acute severe hypoxemia (oxygen saturtation in the 40%), probably due to spasm of ductus arteriosus during catheter manipulation pre-TRFAPV procedure [18]. Stenting of the PDA was then first performed to reestablish pulmonary blood flow. The first attempt to cross the pulmonary

valve resulted in perforation the RVOT necessitating pericardiocentesis to address the cardiac tamponade. Upon stabilization, the pulmonary valve was perforated and balloon dilated. The baby was subsequently brought to the intensive care unit in a stable condition with O_2 saturation of 92%. After 4 days, patient had multiorgan dysfunction syndrome (acute kidney injury prerenal, hypoxemic ischemic encephalopathy, hemorrhagic disease of the newborn) and expired [19].

Changes in hemodynamic measurements (oxygen saturations and pressures) listed in (**Table 1).**

	Pre TRFAPV-BV	Post TRFAPV-BV
Oxygen saturations (%)	68.67 (8.77) [59-81]	91.25% (6.31%) [81-98.00]
RV systolic pressure (mmHg)	105.13 (21.64) [71.00-125.00]	48.57 (20.30) [27.00-90.00]
RV systolic pressure/ aortic systolic pressure ratio	1.32 (0.29) [1.00-1.90]	0.53 (0.05) [0.47-0.60]
Gradient across pulmonary valve (mmHg)	N/A	28.63 (19.35) [5.00-60.00]

Table 1: Changes of hemodynamic measurements pre and post TRFAPV-BV.

Changes in hemodynamic measurements pre and post-TRFAPV-BV showed that oxygen saturations increased from a mean of 68.67% (range 59 to 81%) to a mean of 91.25% (range of 81 to 98%). Initial RV systolic pressures with mean of 105.13mmHg (range 71 to 125mmHg) decreased to a mean of 48.57mmHg (range 27 to 90mmHg). Initial RV systolic pressure/ aortic systolic pressure ratio showed a mean of 1.32 (range of 1 to 1.9) decreased to a mean of 0.53 (range 0.47 to 0.60]. Gradient across pulmonary valve post TRFAPV-BV had a mean of 28.63mmHg (range 5 to 60mmHg).

Summary of clinical course of patients who underwent TRFAPV-BV listed in (**Table 2).**

Variables	No. (%) or Mean (SD) [range]
Success of procedure	8 (89%)
Periprocedural complications	2 (22%)
Mortality	3 (33%)
Other cardiac procedures done	2 (22%)
Pre-TRFAPV-BV oxygen saturations (%) on room air	68.67 (8.77) [59-81]
Need for prostaglandin after TRFAPV-BV (days)	1.67 (1.63) [1.00-5.00]
Oxygen saturations (%) prior to discharge	91.25% (6.31%) [81.00-98.00]
Post-intervention follow up (months)	12 (6.54) [4.00-23.00]
O2 saturations on follow up (%)	96.50 (3.21) [92.00-99.00]
Procedures after TRFAPV-BV done	4 (67%)
PFO/ASD flow bidirectional	4 (67%)
PFO/ASD flow intact	2 (33%)
Circulation type Biventricular	6 (100%)

Table 2: Summary of clinical course of patients with PA-IVS who underwent TRFAPV-BV.

As mentioned above, we had a technical procedural success rate of 89%. We had 3 deaths (33%), two of which as described above had a precipitating cardiac tamponade from RVOT perforation. The third mortality occurred in a 2-day-old, 3.75 kg full-term, born to a mother with gestational diabetes mellitus and had biventricular hypertrophy on electrocardiogram and echocardiography. The pulmonary valve was successfully perforated and dilated without any periprocedural complications. The patient was maintained on prostaglandin infusion for 1 day, baby was observed for 3 days off prostaglandin and then discharged from the hospital with oxygen saturations of 85%. After a week from discharge, the patient expired due to hypercyanosis at a tertiary hospital as claimed by the parents [20].

Other procedures such as balloon atrial septostomy, stenting of the ductus and pericadiocentesis were done in 2 (22%) of our patients. This is to address the above- mentioned complications. The mean pre-TRFAPV-BV oxygen saturations on room air were 68.67% with a range of 59 to 81%. The mean duration time for the need of prostaglandin infusion is 1.67 days with the range of 1 to 5 days. At room air without prostaglandin infusion, the patients were discharged with mean oxygen saturations of 91.25% [range 81 to 98%].

Six of our patients had a mean follow up of 12 months [range 4 to 23 months]. On follow up, mean oxygen saturations were at 96.5% [range 92 to 99%]. Four (67%) had additional procedures of RVOT re-intervention (percutaneous pulmonary balloon valvuloplasty). Four (67%) had bidirectional flow across the patent foramen ovale and two (33%) had intact interatrial septum. All 6 (100%) patients had a biventricular type of circulation **(Table 3).**

Variables	No RVOT intervention after RFV	Any RVOT intervention after RFV
	(N=2)	(N=4)
Demographic		
Weight (kg)	7.4 {7-7.8}	12 {6.50-24.20}
Height (cm)	69.5 {65-74}	79.5 {67-145}
Age (months)	5.5 {4-7}	15.5 {90-98}
Timing of reintervention after TRFAPV-BV(months)	N/A	6.5 {4-16}
Oxygen saturations %	99 (0) [99-99]	94.25 (3.86) [90-98]
Residual gradient across pulmonary valve (mmHg)	8 [5-11]	43.5 {28-60}
Pulmonary valve annulus z-score	-2.8 [-2.8--2.8]	-2.75{-3.52--1.12}
Echocardiographic measurements		

Tricuspid to Mitral valve ratio	0.80 [0.8-0.8]	1.01 (0.43) [0.68-1.50]
Tricuspid regurgitation (mmHg)	29 (8.49) [23-35]	65.25 (32.98) [28-103]
Pulmonary valve annulus (mm)	10.47 (0.76) [9.93-11.0]	9.48 (3.51) [5.5-13.8]
Pulmonary valve annulus z-score	-0.08 (0.28) [-0.27-+0.12]	-2.45 (1.17) [-3.97--1.39]
Gradient across pulmonary valve (mmHg)	14.5 (4.95) [11-18]	64 {61-111}
Cardiac catheterization		
RV systolic pressure	N/A	84.33 (21.08) [60-97]
Gradient across pulmonary valve (mmHg)	N/A	68.50 (22.35) [38-88]
Maximum balloon diameter/pulmonary valve annulus	N/A	1.32 (0.07) [1.27-1.40]
RV systolic pressure/ aortic systolic pressure	N/A	0.54 (0.31) [0.32-0.89]
Gradient across pulmonary valve (mmHg)	N/A	17.33 (4.62) [12-20]

Table 3: RVOT re-intervention after TRFAPV-BV.

Due to the rarity of the disease and the procedure, limited number of patients in this study cannot establish whether there is a statistical significance between groups. There are also lacking data in tricuspid annulus z-score and tricuspid to mitral value ratio. The timing of the reintervention after the initial TRFAPV-BV had a range of 4 to 16 months with a median duration of 6.5 months. Nevertheless, it was observed that patients who underwent RVOT reintervention had a higher residual gradient across pulmonary valve (43.5 mmHg [28-60 mmHg] *vs* 8 mmHg [5-11mmHg]) after initial TRFAPV-BV. The four patients who had redilation of the pulmonary valve had progression of residual pulmonary valve stenosis to at least moderate degree based on follow-up serial echocardiogram as the indication for the re-intervention. The mean gradient of the across the pulmonary valve was 68 mmHg.

Discussion

The ability to successfully perforate an atretic pulmonary valve by TRFAPV- BV is dependent on several characteristics as noted by many investigators. Our technical success rate of 89% (8 successful cases in 9 attempts) was not limited by the degree of hypoplasia of the pulmonary valve as six of our patients with a pulmonary valve z-score of -3.58 to -2.8 were done.

All our patients who survived the procedure had a right ventricular systolic pressure/aortic pressure ratio more than one and had a lower degree of tricuspid regurgitation. This is consistent with the finding of Nykanen, who described a RV systolic/aortic systolic pressure of less than one as a disadvantaged right ventricle.8 Our data revealed only a single patient who succumbed one week after intervention who had a subsystemic right ventricular pressure. Heart failure in subsystemic right ventricle can be probably due to poor ventricular compliance leading to decreased antegrade flow to the pulmonary arteries and low cardiac output.

All patients required the maintenance of systemic to pulmonary arterial blood flow by keeping the ductus arteriosus open with prostaglandin infusion. Schmidt in 1992 suggested that prostaglandin should be continued for at least 1-2 weeks post TRFAPV-BV with some cases needing prostaglandin for as long as 4 to 6 weeks. However, in our cases, prostaglandin infusion post TRFAPV-BV was only maintained for a mean duration of 1.67 days with a range of 1 to 5 days. All our patients were discharged with acceptable oxygen saturations that range from 81%-98%. Our strategy was to avoid stenting of the ductus arteriosus after a successful TRFAPV-BV as long as we can document that there is forward flow across the pulmonary valve after the intervention with concomitant increase in oxygen saturations.

In our cases, none of our patients who survived needed additional systemic to pulmonary arterial blood flow either by stenting the ductus arteriosus or surgical Blalock Taussig Shunt. Nykanen reported that stenting of the ductus arteriosus occurred in 33 to 58% of their cases.8 Furthermore, several experts advise the creation of additional pulmonary blood flow shunt if the pulmonary valve annulus or the valve orifice is inadequate. In retrospect, the only case that may have benefited from a stenting of the ductus arteriosus was the patient who died a week after discharge with biventricular hypertrophy. This patient had a right-to-left ventricular pressure ratio of<1, severe tricuspid regurgitation, and a markedly reduced tricuspid valve annulus z- score of -2.83. Cheatham in 1998, concluded that tricuspid valve z-score of less than -2.5 predicts poor clinical outcomes.

All our successful cases with an adequate pulmonary blood flow and oxygen saturations, the tricuspid valve annulus z-scores were greater than -2.5, which was much smaller than, who mentioned that tricuspid valve hypoplasia with a z score of less than -0.7 might be predictive of the need for an additional source of pulmonary blood flow.16 Although none of our surviving patients needed additional pulmonary blood flow despite very small tricuspid valve annulus, it is warranted that they be monitored carefully for this eventual need.

All our patients had a tripartite right ventricle with no right ventricle to coronary connections. Both findings are consistent with previous reports as good predictors of survival. All had a tricuspid valve annulus z-score of greater than -2.5 except for one with -2.83. Majority were consistent with the that tricuspid z-scores greater than -2.5 have tripartite right ventricle type with no major sinusoids.24 Furthermore, all cases that achieved a successful biventricular circulation had a tricuspid to mitral valve ratio that ranged from 0.62 to1.34. This is in agreement with that a tricuspid/mitral ratio >0.5 has been shown to be an excellent predictor of successful biventricular repair in PA-IVS patients.

There were three deaths in our case series, two of which were periprocedural. Perforation of the right ventricular outflow tract

leading to hemopericardium is a life- threatening complication reported in this procedure high-risk procedure. Although we were able to successfully do a pericardiocentesis, the circulatory collapse and ventricular injury sustained by these sick patients put them at further risk. Attention to detail in terms of proper catheter position and careful manipulation cannot be overemphasized.

Due to a limited number of successful patients, statistical significance to describe characteristic differences between cases requiring or not requiring RVOT reintervention after TRFAPV-BV cannot be determined. The timing of the reintervention after the initial TRFAPV-BV had a range of 4 to 16 months with a median duration of 6.5 months. Reintervention is dependent on a higher residual gradient across pulmonary valve and more negative pulmonary valve annulus z-score.

Conclusion

Our retrospective review shows that TRFAPV-BV as a treatment in properly selected cases of PA-IVS is associated with good immediate success. Our review is congruent with conclusion of many investigators that have observed that tricuspid valve annulus z-score greater than -2.5, tripartite right ventricle, absence of ventricular to coronary connections, tricuspid to mitral valve ratio greater than 0.5, and right-to-left ventricular pressure ratio of more than 1, are positive predictors of survival in patients undergoing TRFAPV-BV.

Moreover, parameters after TRFAPV-BV of higher residual gradient across pulmonary valve and lower pulmonary valve annulus z-scores may be predictive of the need for subsequent RVOT intervention.

Declarations

The University of the Philippines Manila Research Ethics Board approved the study. There was no conflict of interest from financial, proprietary considerations of the author or the study site. Strict confidentiality was imposed and the anonymity was protected.

Limitation of the Study and Recommendations

Due to the rarity of the disease and procedure, there is a limited sample size to compute if the parameters are statistically significant or not. In the absence of a protocol in what parameters to measure, there are some data lacking in the review of patients records, and management was based on the discretion of the attending physician. A systematic institutional procedural guideline on how to screen and manage these high-risk patients is being drafted based on this pilot study.

Based on the findings of this review, a 10-year prospective study that utilizes the criteria that predicts successful outcomes may be used in the selection of patients diagnosed with PA-IVS who will undergo TRFAPV-BV. This is to assess survival, morbidity, timing of trans catheter intervention or open-heart surgery and quality of life.

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