Journal of the American College of Cardiology
© 2000 by the American College of Cardiology
Published by Elsevier Science Inc.

Vol. 35, No. 2, 2000
ISSN 0735-1097/00/$20.00
PII S0735-1097(99)00549-5

Pulmonary Atresia With Intact Ventricular Septum Percutaneous Radiofrequency-Assisted Valvotomy and Balloon Dilation Versus Surgical Valvotomy and Blalock Taussig Shunt

Mazeni Alwi, MRCP,* Kandhavello Geetha, MRCP,* Abdul A. Bilkis, MD,* Miin K. Lim, MRCP,* Samion Hasri, MD,* Abdul L. Haifa, MD,* Ahmad Sallehudin, FRCS,† Robaayah Zambahari, FRCP, FACC*

Kuala Lumpur, Malaysia

OBJECTIVE We compared the result of radiofrequency (RF)-assisted valvotomy and balloon dilation with closed surgical valvotomy and Blalock Taussig (BT) shunt as primary treatment in selected patients with pulmonary atresia and intact ventricular septum (PA-IVS).

BACKGROUND Patients with PA-IVS who have mild to moderate hypoplasia of the right ventricle (RV) and patent infundiblum have the greatest potential for complete biventricular circulation. The use of RF or laser wires to perforate the atretic valve followed by balloon dilation provides an alternative to surgery.

METHODS Between May 1990 and March 1998, 33 selected patients underwent either percutaneous RF valvotomy and balloon dilation (group 1, n = 21; two crossed over to group 2) or surgical valvotomy with concomitant BT shunt (group 2, n = 14). Second RV decompression by balloon dilation or right ventricular outflow tract (RVOT) reconstruction were performed if necessary. Patients who remained cyanosed were subjected to transcatheter trial closure of the interatrial communication. Partial biventricular repair was offered to those with inadequate growth of the RV.

RESULTS The primary procedure was successful in 19 patients in group 1. There was one in-hospital death and two late deaths. Of the remaining 16 survivors, 12 achieved complete biventricular circulation, 7 of whom required no further interventions. Two patients required repeat balloon dilation, 1 RVOT reconstruction and 2 transcatheter closure of interatrial communication. Two patients underwent partial biventricular repair. In group 2, there were 3 in-hospital deaths after the primary procedure and 1 patient died four months later. All survivors (n = 10) required a second RV decompression, 8 by balloon dilation and 2 by RVOT reconstruction, after which, two patients died. Of the final 8 survivors, 7 achieved complete biventricular circulation, 5 after coil occlusion of the BT shunt and 2 after closure of interatrial communication.

CONCLUSIONS Radiofrequency valvotomy and balloon dilation is more efficacious and safe compared with closed pulmonary valvotomy and BT shunt in selected patients with PA-IVS. (J Am Coll Cardiol 2000;35:468–76) © 2000 by the American College of Cardiology

Pulmonary atresia with intact ventricular septum (PA-IVS) is a relatively uncommon form of congenital heart disease and is remarkable in its morphologic diversity, characterized by varying degrees of right ventricle (RV) hypoplasia and associated with tricuspid valve dysplasia and sinusoidal right ventriculo-coronary connections (1–3). A single uniform approach in the management of this disease is therefore impractical.

It has increasingly been recognized that there are two principal forms of this disease, those with mild to moderate hypoplasia of the RV who generally have a patent infundibulum, and those with severe hypoplasia of the RV who positively correlate with major RV-coronary connections (4). For patients in the first group, who incidentally form a majority in most of the reported series, many authors believe that the treatment goal is to eventually establish a complete biventricular circulation with RV decompression as the initial palliative procedure (5). On the other hand, patients

From the *Department of Cardiology and †Department of Cardiothoracic Surgery, Institut Jantung Negara (National Heart Institute), Kuala Lumpur, Malaysia. This work was supported and funded by the Hospital Research Committee.

Manuscript received October 19, 1998; revised manuscript received September 27, 1999, accepted October 18, 1999.

Abbreviations and Acronyms

ASO	=	Amplatzer Septal Occluder
BT	=	Blalock Taussig
CVP	=	central venous pressure
PA-IVS	=	pulmonary atresia with intact ventricular septum
PDA	=	patent ductus arteriosus
PFO	=	patent foramen ovale
PGE1	=	prostaglandin E1
RF	=	radiofrequency
RV	=	right ventricle
RVOT	=	right ventricular outflow tract
2DE	=	two-dimensional echocardiography

with severely hypoplastic, diminutive RV are generally assigned towards a univentricular repair, and in the difficult subset of patients with RV-dependent coronary circulation, cardiac transplantation may be an option. Right ventricle decompression may be achieved by simple pulmonary valvotomy without cardiopulmonary bypass, or by a more radical transannular patch of the right ventricular outflow tract (RVOT) in the neonatal period with or without a concomitant systemic-pulmonary shunt. More recently, the use of laser wire and radiofrequency (RF)-assisted valvotomy and balloon dilation has made possible primary treatment of this group of patients to be undertaken in the catheterization laboratory (6,7).

This study reviews the outcome of percutaneous RF valvotomy and balloon dilation versus closed surgical valvotomy and Blalock Taussig (BT) shunt in a selected group of PA-IVS patients with mild to moderate RV hypoplasia and patent infundibulum.

METHODS

Patient selection. We reviewed all patients with PA-IVS who were referred to our institution from May 1990 to March 1998. All patients underwent detailed echocardiographic examination by two principal investigators (M.A. and G.K.), paying particular attention to the size of the RV and tricuspid valve compared with that of the LV and mitral valve, respectively, presence or absence of RV infundibulum and whether right ventriculo-coronary connections were present. The degree of tricuspid regurgitation was assessed by color flow in the apical four-chamber view based on the ratio of maximum regurgitant area to the total right atrial area as described by Fisher and Goldman (8). Grade 1+ to 2+ regurgitation was defined as mild, grade 3+ as moderate and grade 4+ as severe. Apart from visual assessment of the two-dimensional echocardiography (2DE) images, RV hypoplasia was defined as mild if the tricuspid/mitral diameter ratio in the four-chamber view was >0.75; a ratio of 0.50 to 0.75 was considered moderate and a ratio of <0.50 severe. The diameter of tricuspid valve measured on 2DE and the body surface area were used to compute the Z value using

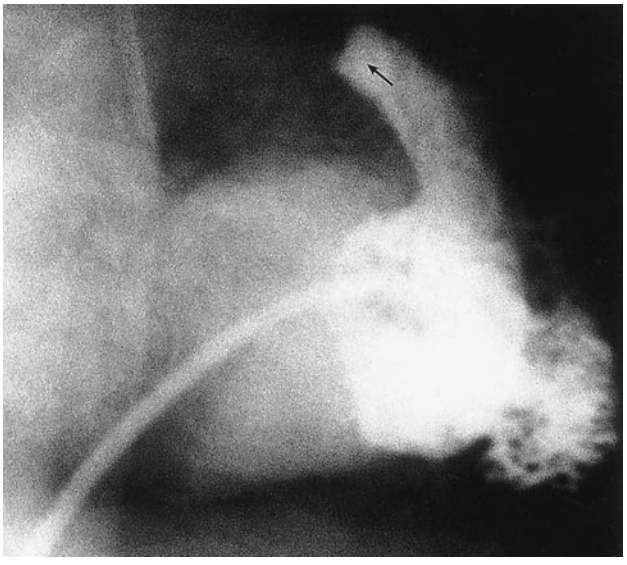

Figure 1. Right ventricular angiogram in the lateral projection showing a well-developed trabecular component, patent infundibulum and atretic pulmonary valve (**arrow**). Moderate tricuspid regurgitation.

the normogram of the Congenital Heart Surgeon Study Group Protocol (CHSS) (9) from January 1996 onwards. Thirty-three patients who had RV anatomy deemed favorable for biventricular circulation, i.e., mild or at most moderate hypoplasia and patent infundibulum, were subjected to percutaneous RF valvotomy and balloon dilation (group 1: n = 21, two of whom crossed over to group 2) or to closed surgical valvotomy without cardiopulmonary bypass with a concomitant BT shunt (group 2: n = 14) as the primary procedure in an unrandomized fashion (Fig. 1). Group 2 essentially formed a historical control as they underwent the surgical procedure before the introduction of the transcatheter technique, except for the two patients who crossed over after unsuccessful attempt at percutaneous RF valvotomy. Patients with diminutive, severely hypoplastic RV and tricuspid valve as well as absence of RV infundibulum were excluded from the present study and were assigned for eventual single-ventricle repair. The rare patients with gross dilation of the RV with Ebstein's malformation of the tricuspid valve and severe tricuspid regurgitation were also excluded.

With the unusual exception of one patient who presented at eight years of age, the majority (27/33, 81%) presented in the newborn period. Three patients presented at three months of age, 1 at seven months and 1 at one year. Twenty-seven (81%) patients received prostaglandin E1 (PGE1) infusion before and during the primary procedure. One patient had a small, restrictive ventricular septal defect but was nevertheless included in the study, as the clinical presentation and other echocardiographic features were otherwise consistent with the diagnosis of PA-IVS.

JACC Vol. 35, No. 2, 2000
February 2000:468–76

Transcatheter primary procedure. Twenty-one patients were subjected to RF valvotomy and balloon dilation, of whom, two patients were later included in the surgical group after unsuccessful procedure (group 1). All patients in group 1 underwent cardiac catheterization under general anesthesia. The femoral artery and vein were cannulated percutaneously using 4F and 5F sheaths, respectively. Right ventricle-dependent coronary circulation was first excluded in all patients. Radiofrequency valvotomy was performed using the HAT 300 generator and 0.018-in. cereblate wire (Sulzer Osypka GmBH, Grenzach-Wyhlen, Germany) via a 5F Judkins right catheter. After successful valvotomy, a 0.014-in. Flex coronary guidewire (ACS Hi-Torque; Floppy II, Temeluca, California), and in the later cases Crosswire (Terumo Corp., Tokyo, Japan), was passed antegradely into the pulmonary artery. Procedural failure is defined as inability to perforate the atretic valve with the RF wire, and subsequent balloon dilation thus could not be performed.

Earlier in the series, the flex coronary wire was routinely snared in the main pulmonary artery and brought down into the descending aorta for better support during balloon dilation. Balloon dilation was performed sequentially using 2-mm and 4-mm coronary balloons (Schneider [Europe] GmBH, Bulach, Switzerland) and finally 8- to 10-mm Tyshak (NuMED; Cornwall, Ontario, Canada) valvuloplasty balloon. The right ventricular-to-aortic pressure ratio was used as a hemodynamic index of the severity of RV hypertension. Prostoglandin infusion was then gradually weaned off after 48 h, and if the patient remained duct dependent two weeks after the procedure or showed evidence of ductal closure despite PGE1 infusion, a modified BT shunt or stenting of the patent ductus arteriosus (PDA) was put forward (adjunct procedure).

Surgical valvotomy. Fourteen patients, including the two who had unsuccessful RF valvotomy, underwent closed pulmonary valvotomy with a concomitant BT shunt via left thoracotomy (group 2). Prostoglandin infusion was discontinued 24 h after the surgery. None of these patients underwent preliminary cardiac catheterization studies, except the two patients who crossed over.

Echocardiographic examination chiefly looking at the degree of residual RVOT obstruction was performed at 24 h after the primary procedure and repeated as necessary during the initial hospitalization.

Morbidity and mortality. The duration of hospital stay, requirement for ventilatory and inotropic support, complications and mortality related to the primary procedure between two groups were compared. Mortality within the initial hospital stay was considered as in-hospital death, and late death were patients who died during subsequent admissions.

Follow-up. On follow-up, the degree of cyanosis, presence of RV failure and echocardiographic observations on RV growth, residual pulmonary stenosis and tricuspid regurgitation were noted. Complete biventricular circulation was considered to have been achieved if there was no clinical cyanosis ($SaO_2 > 96\%$) and echocardiograhic features of at most mild RV hypoplasia and absence of predominantly right to left shunt across the interatrial communication on color Doppler if it has not been closed surgically or by a device.

Subsequent procedures. All survivors from both groups were subjected to cardiac catheterization four to six months after the primary procedure if significant RV outflow obstruction remained on Doppler echo (peak systolic gradient $>$ 25 mm Hg). Balloon dilation was performed if the stenosis was at valvar level, and likewise for RVOT reconstruction for subvalvar obstruction. All survivors in group 2 who were deemed to have achieved complete biventricular circulation underwent further cardiac catheterization at 18 to 24 months (mean 21 months) of age for coil occlusion of the BT shunt. In both groups, for patients who remained cyanosed after the primary procedure despite having no significant RVOT obstruction (peak systolic gradient $<$ 25 mm Hg) but continued to have mild to moderate RV hypoplasia, a trial closure of the interatrial communication/patent foramen ovale (PFO) using a balloon catheter and followed by Amplatzer Septal Occluder (ASO) device (AGA Medical Corporation, Golden Valley, Minnesota) was performed. If the systemic pressure dropped by more than 20% of the baseline and/or central venous pressure (CVP) increased to $>$15 mm Hg, the RV was considered inadequate for complete biventricular circulation. The ASO device was then removed and the patient was subjected to partial biventricular repair, i.e., creation of bidirectional Glenn's shunt and closure of PFO. In patients who could tolerate closure of the PFO using the ASO device, a dobutamine challenge was then performed at incremental doses of 5 and 10 μg/kg/min for 10 min each. If the hemodynamics remained stable, the device was then released permanently and the patient was considered to have achieved complete biventricular circulation.

Statistical analysis. The nonparametric Wilcoxon rank sum test and the Mann-Whitney *U* test were used to compare the continuous variables. Comparison of dichotomous variables was made using the Fisher exact test. Statistical significance was assumed at 5% level.

RESULTS

Table 1 summarizes patient characteristics and preinterventional echocardiographic findings of both groups. The median weight for patients in group 1 was 3.4 kg (range 2.0 to 18.0 kg) and 3.3 kg in group 2 (range 1.7 to 6.0 kg). None of the patients had Ebstein's anomaly, and tricuspid regurgitation varied from 0 in 5 to severe in 12. Two patients in both groups, respectively, had right ventriculo-coronary

Table 1. Patient Data and Preinterventional Echocardiographic Findings

Patient Data	Group 1	Group 2	p
No. of patients	19	14	
Median age at diagnosis in days (range)	6 (1–8 yrs)	1 (1–48)	0.09
Gender			
Female	8	6	NS
Male	11	8	
Median weight in kg (range)	3.4 (2–18)	3.3 (1.7–6)	0.3
Echocardiography			
Ebstein's malformation			
No	19	14	NS
Yes	0	0	
Tricuspid regurgitation			
Nil	2	3	NS
Mild	5	0	
Moderate	3	8	
Severe	9	3	
R ventriculo-coronary connections			
Absent	17	12	NS
Present	2	2	
RV hypoplasia			
Mild	11	8	NS
Moderate	6	6	
Severe	2	0	

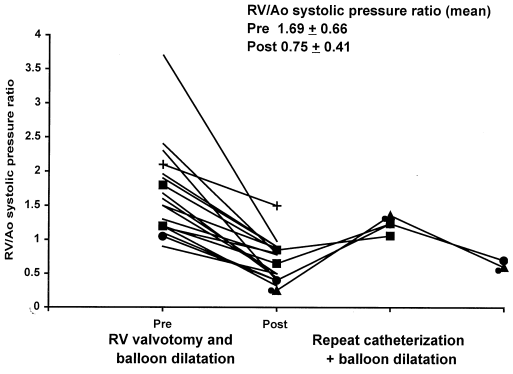

Figure 2. Right ventricular-aorta systolic pressure ratio before and immediately after RF valvotomy and balloon dilation, and during subsequent cardiac catheterization in 19 patients from group 1. One patient was in critical condition at the beginning of the procedure and the initial RV pressure was not available (**triangles**). Patients with subvalvar stenosis (**squares**). Patients who underwent repeat balloon dilation (**circles**). Transvalvar Doppler gradient dropped to < 25 mm Hg at 10 weeks of follow-up (**plus signs**).

connections noted on echocardiogram. There was no significant difference in the characteristics of preinterventional echocardiogram. Dimensions of the tricuspid valve were measured in 17 of the 19 patients in group 1. The mean tricuspid valve diameter was 11 ± 2.6 mm (range 6 to 15 mm) and the mean Z value was −0.6 ± 1.6 (range −4.0 to +1.5). Three patients had Z values of -3.0 and less. The patients in group 2, however, did not have detailed measurement of the tricuspid valve diameter and Z value.

Group 1. Percutaneous RF valvotomy was unsuccessful in two patients (9%), and they were subjected to the standard surgical procedure electively two and four days later. There was no noticeable deterioration in their clinical condition or hemodynamic status after cardiac catheterization. Marked subvalvar muscular hypertrophy was noted in these two patients, and the catheter tip could not be placed beneath the atretic valve but instead was misdirected into the intertrabecular recesses. In the remaining 19 patients in whom the procedure was successful, the RV/aortic systolic pressure ratio dropped from a mean of 1.69 ± 0.66 (0.9 to 3.7) to 0.75 ± 0.41 (0.3 to 1.9) (Fig. 2). The mean procedure time was 126.1 ± 34.8 min (range 60 to 190 min). Two patients remained on prostaglandin infusion two weeks after the primary procedure. One of them received a right modified BT shunt. The other patient became severely cyanosed due to ductal closure despite PGE1 infusion, and she underwent PDA stenting using a 3.5-mm Bard XT stent (C.R. Bard Ireland Ltd., Galway, Ireland). The stent was found to be thrombosed three months later despite maintenance of aspirin, but the patient remained pink as the RV compliance had improved by then. In all survivors of the primary procedure, the transvalvar Doppler gradient dropped to <25 mm Hg and the RVOT appeared wide open at a mean of 10 weeks of follow-up, except in one patient whose transvalvar gradient remained high (80 mm Hg). Repeat cardiac catheterization four months later revealed subvalvar stenosis and a small pulmonary annulus. She later underwent RVOT reconstruction and closure of restrictive VSD. Three patients who attained adequate RV decompression at the initial procedure, however, developed restenosis. In two patients, restenosis was valvar, and both underwent successful balloon dilation six months later. In one patient, the restenosis was subvalvar, but the patient died suddenly 24 h after an otherwise uneventful diagnostic cardiac catheterization.

COMPLICATIONS AND MORTALITY. The total mortality in group 1 was 15.7%. (Table 2). One patient died 3 h after a successful RF valvotomy and RV decompression due to uncontrolled retroperitoneal hemorrhage. This was presumed to have originated from iliac artery trauma during the arterial cannulation. Perforation of the RV outflow tract by the RF wire was seen in three patients, resulting in self-limiting small hemopericardium. Supraventricular tachycardia with hypotension occurred in two patients during the procedure, and both responded well to direct current cardioversion. One patient presented with femoral vein thrombosis one week after a successful procedure, which resolved with heparin infusion. Late deaths were seen in two patients. One death was due to fulminant staphylococcal sepsis four months after a successful primary procedure. Another had an unexplained sudden death 24 h after an uneventful

Table 2. Morbidity and Mortality

	Group 1	Group 2	p
Median days of hospital stay (range)	9 (2–24)	20 (6–47)	0.009
Median days of ventilation (range)	0 (0–4)	5 (1–36)	0.0002
Median days of inotropic support (range)	0 (0–2)	0 (0–30)	0.07
Deaths			
In hospital	1	3	
Late	2	3	
Total (%)	3 (15.7)	6 (42.8)	0.18

diagnostic cardiac catheterization. The patient had subvalvar RV outflow obstruction with a RV systolic pressure of 100 mm Hg.

FOLLOW-UP AND LATE OUTCOME. All the survivors (16/19) were followed up for a minimum of six months after the primary procedure, with a mean of 18 months (6 to 36 months). Twelve patients achieved complete biventricular circulation, seven of whom required no further interventions. Of the remaining five patients, two underwent repeat balloon dilation for restenosis and another underwent RVOT reconstruction with concomitant closure of restrictive ventricular septal defect. These three patients were considered to have attained complete biventricular circulation. Another two patients attained complete biventricular circulation after closure of PFO using the ASO device, 14 and 34 months, respectively, after the primary procedure. This eliminated the cyanosis without impairment of the cardiac output. One of these patients underwent coil occlusion of the ductus arteriosus as well.

Two other patients attained reasonable RV growth (Z +0.6 and +0.1, respectively), albeit with mild cyanosis, but the parents are hesitant for transcatheter closure of the PFO.

Two of the 16 patients (12.5%) in group 1 failed to achieve complete biventricular circulation. The Z values of the tricuspid valve were -3.5 and -4.0 at initial presentation. They both received bidirectional Glenn's shunt and closure of the PFO at six and eight months after the primary procedure.

In one patient, who was severely cyanosed at eight months post-RF valvotomy due to poor RV growth (Z = −3.4), trial closure of PFO eliminated the cyanosis but resulted in hypotension and elevated CVP after 10 min, and the device was then removed. However, at 18 months, despite surgical closure of the PFO at the time of the Glenn's shunt construction, he still had bidirectional shunting through a residual interatrial communication. It was successfully occluded with an ASO device (Fig. 3). In the other patient, trial closure was not undertaken, as the RV was deemed too small (Z = −6) to support the pulmonary circulation (Fig. 4).

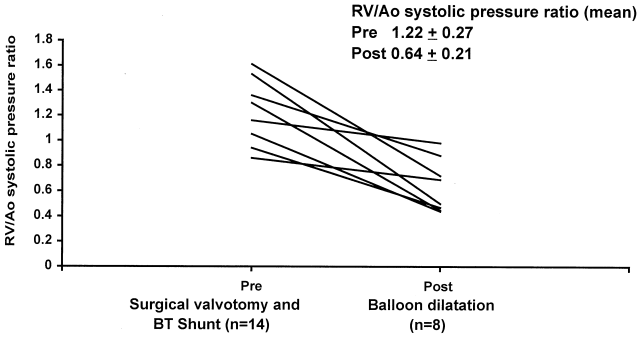

Figure 3. Right ventricular-aortic systolic pressure ratio before and after second RV decompression by balloon dilation in eight patients from group 2.

Group 2. COMPLICATIONS AND MORTALITY. The overall mortality was significantly higher in group 2 (42.8%; p = 0.04) (Table 2). Of the 14 patients in the surgical group, there were three in-hospital deaths after the primary procedure, two due to RV failure at 13 and 36 postoperative days. Both of these patients crossed over from group 1 after failure of percutaneous procedure due to the presence of marked subvalvar muscular hypertrophy. One patient died from pneumonia at the fifth postoperative day. One patient died four months later from RV failure. All 10 survivors required a second RV decompression procedure at 6 to 12 months after the primary surgical procedure due to inadequate relief of RVOT obstruction. Eight were by balloon dilation and two by RVOT reconstruction. The mean RV/aortic systolic pressure ratio before the second procedure was 1.22 ± 0.27 (Fig. 5). Another two patients died from RV failure after the second RV decompression procedure, one soon after surgery and the other 4 h after balloon

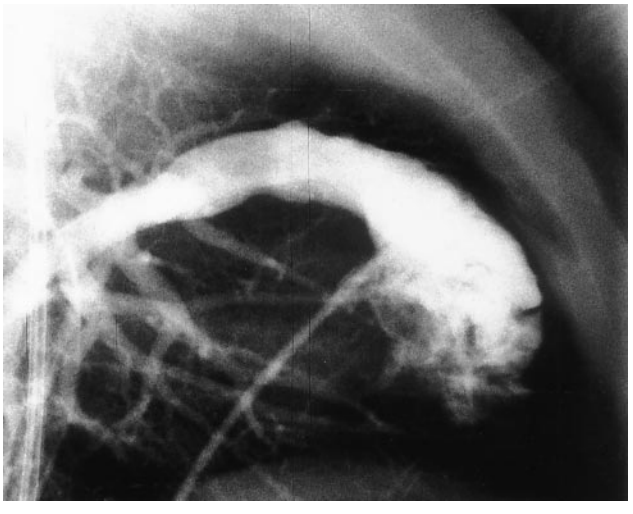

Figure 4. Right ventricular angiogram in the lateral projection eight months after RF valvotomy and balloon dilation. Unobstructed RV outflow tract but relatively small right ventricle (Z value −3.4). Closure of interatrial communication with ASO eliminated the cyanosis with no deterioration of hemodynamics.

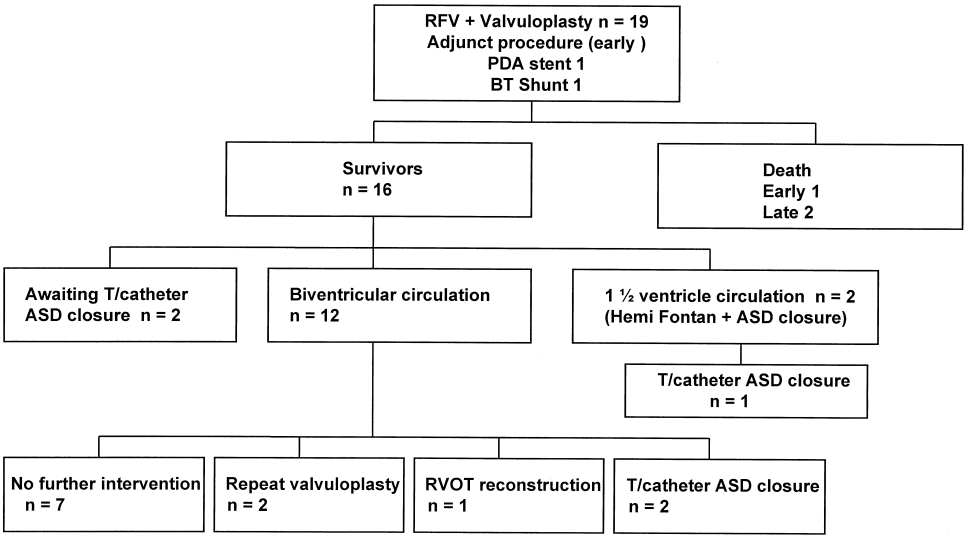

Figure 5. Flow chart summarizing the procedures and outcome of patients in group 1.

dilation. Both of these patients presented in critical condition and the procedures were performed as an emergency.

Eight patients developed postoperative complications of diaphragmatic paralysis, blocked shunt, wound infection and pneumonia. One patient developed shunt-related left pulmonary artery stenosis, and this was treated by placement of a Palmaz stent (Johnson and Johnson, Interventional Systems, Warren, New Jersey) intraoperatively 22 months after the primary procedure. Hospital stay and requirements for ventilatory support were significantly higher in group 2 ($p = 0.009$ and $p = 0.0002$, respectively, Table 2).

FOLLOW-UP AND LATE OUTCOME. The survivors ($n = 8$) in this group had been followed up for a mean of 51 ± 20.3 months (33 to 96 months). Seven patients who were deemed to have complete biventricular circulation had their BT shunt occluded with Gianturco coils. Five patients did not require any further intervention. Two patients underwent transcatheter closure of the PFO at three and eight years of follow-up. The remaining patient is awaiting BT shunt occlusion and trial closure of PFO (Fig. 6)

DISCUSSION

The few reports on long-term survival of patients with PA-IVS have shown that the outlook is relatively poor (10,11). However, recent reports have emphasized improved operative results in infants with this disease (12–14). This has been attributed in part to a better understanding of right ventricular morphology and its use in the selection of

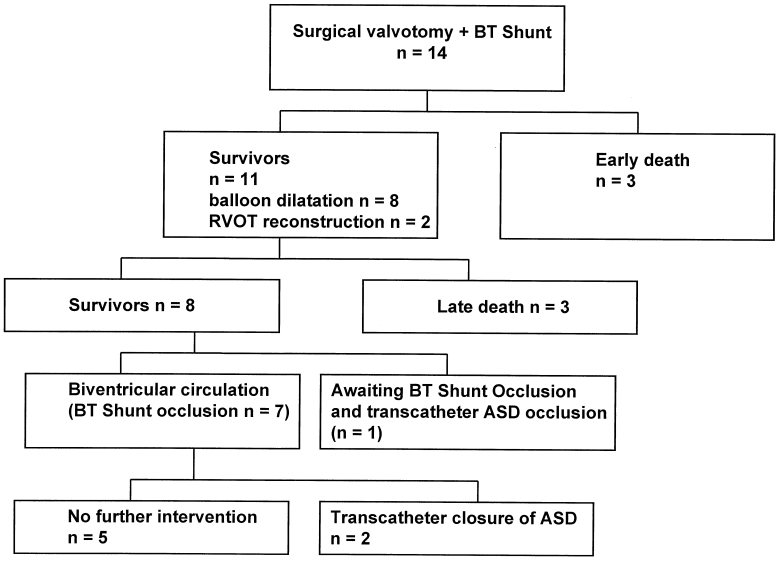

Figure 6. Flow chart summarizing the procedures and outcome of patients in group 2.

JACC Vol. 35, No. 2, 2000
February 2000:468–76

management strategy (5,15). Many authors would now agree that patients with mild-to-moderate RV hypoplasia and patent infundibulum are potential candidates for eventual biventricular circulation. In these patients, the goal of the initial palliation is to decompress the RV and establish forward flow, which, in turn, will promote growth of the RV (4,14). Controversy still exists on the optimal form of RV decompression and whether a systemic-pulmonary shunt should be an integral part of the initial palliative procedure (8,16–20).

While transannular patching provides the greatest possibility of unobstructed forward flow from the RV, the proponents for pulmonary valvotomy have argued that it produces less ventricular damage and pulmonary regurgitation (14). It can also be performed safely without cardiopulmonary bypass. Although it may not completely relieve pulmonary outflow obstruction, a more radical RV outflow reconstruction can be deferred to late infancy when the morbidity and mortality risks can be expected to be lower. Furthermore, in those patients whose residual obstruction is chiefly at valvular level, nonsurgical reintervention by balloon dilation offers a safe and effective alternative (21). This has been our approach in eight patients in group 2.

Transcatheter primary procedure. Transcatheter valvotomy and balloon dilation offer a promising alternative as the primary therapy in selected patients with PA-IVS. Perforation of the atretic pulmonary valve can be achieved using laser guidewire or RF wire, and balloon dilation can then be performed in the conventional way (6,7,22). The sharp, stiff end of a coronary guidewire has been successfully used to perforate the atretic pulmonary valve (23), but this technique has been rendered largely obsolete with the availability of laser and RF wires. The use of laser-based methods has the advantage of using the same guidewire to track the balloon over once the valve has been perforated. In our experience, however, once valvotomy has been achieved with RF wire, the tiny perforation could be recrossed with another guidewire for subsequent balloon dilation in all cases. Furthermore, the risk to staff, limited portability and considerable capital expense favor the use of the RF method (7). Since its introduction, the technical skills for this demanding procedure have improved, the indications have become more precise and more centers have begun to use this approach (24–26). Although this technique has been used in patients with infundibular atresia and even those with ventricular septal defect (27,28), it is clear that only patients with membranous valvular atresia and a patent infundibulum, in the setting of a non-RV-dependent coronary circulation, should be considered for this procedure to achieve the best results and avoid serious complications (26). This subgroup of patients generally has a reasonably well-developed RV and therefore the greatest potential for biventricular circulation (4). The one early death in our series was a result of a rare but nevertheless known complication of vessel cannulation in a newborn rather than directly related to the procedure itself.

Assessment of RV size. There continues to be much debate on the best method of assessing right ventricular size and morphology. There are proponents of using complex scoring of right ventricular size (16), a simple measurement of tricuspid annulus diameter (16) or merely visual assessment of how well formed the RV appears to be (17,26). Although we began measuring the Z value of the tricuspid valve in the later part of the study, we concur with Gibbs et al. (26) that the latter, simplest approach is probably sufficient for clinical decision making. Of all the survivors from both groups, the RV has not appeared to grow despite virtual elimination of outflow obstruction in two patients, both of whom underwent percutaneous RF valvotomy as the primary procedure. We speculate that this may have been in part attributable to their late presentation (three and seven months), and hence delayed decompression of the RV. Their tricuspid valve diameter z values were -3.5 and -4.0 at presentation, respectively. However, all is not lost, as the RV can still be incorporated into the circulation, albeit with a reduced volume load, by performing a bidirectional Glenn's Shunt and closure of interatrial communication (partial biventricular repair) (18).

Closure of interatrial communication. We have found the ASO, which is retrievable, to be a useful device in determining the potential for complete biventricular circulation in patients who continue to remain cynosed due to inadequate growth of the RV. In patients destined for partial biventricular repair, closure of the PFO would lead to an increase in the CVP and concomitant lowering of the systemic arterial pressure. On the other hand, in those patients whose RV is capable of supporting the entire pulmonary circulation, cyanosis would be eliminated without increasing the CVP and compromising the cardiac output. The device can then be safely deployed permanently. However, we feel that it is important to stretch the capability of the RV by dobutamine stress before deployment.

Subgroup with concomitant subvalvar obstruction. Fixed, subvalvular stenosis is part of the anatomic spectrum of PA-IVS (19). In our experience, this was a cause of failure of perforation of the valve in two patients due to malpositioning of the guiding catheter and RF wire in the intertrabecular recesses. These two patients ($Z = -1.1$ and $+0.5$), who were then subjected to the standard surgical procedure, subsequently died of RV failure at 13 and 36 postoperative days. In retrospect, a more radical RVOT reconstruction, albeit with a higher operative mortality risk, would have been more appropriate. No doubt, the prolonged requirement for inotropic and ventilatory support and eventual death due to inadequate RV decompression have negatively skewed the surgical results significantly. However, in another patient, the procedure was successful,

and this has allowed the patient to grow to a reasonable size when a radical RVOT reconstruction could be performed at a lower risk.

Study limitations. As this study is partly retrospective, the Z value of the tricuspid valve as a measure of the RV size was not available in the historical, surgical control group. Nevertheless, the other preintervention echocardiography findings were similar in the two groups, and all patients underwent initial evaluation and selection into the study by the two principal investigators. Second, the majority of patients in the surgical group did not undergo prior cardiac catheterization to exclude RV-dependent coronary circulation. Nonetheless, the two patients in whom right ventriculo-coronary connections were noted on echocardiography were among the survivors. Finally, the patients in the surgical control group received their treatment in a slightly earlier era, and in two patients who died early of RV failure due to inadequate RV decompression, the choice of surgical treatment may not have been appropriate. Therefore, the morbidity and mortality rates and final outcome may not be representative of current surgical results in established institutions (16,18).

Conclusions. From our small series, we conclude that primary treatment with RF valvotomy and balloon dilation compared with surgical valvotomy in this selected group is more efficacious in decompressing the hypertensive RV in that only a small number required a second decompression procedure. It is also associated with lower morbidity, as indicated by lower requirement for ventilation and freedom from potential complications of surgery. Stenting of the PDA in patients requiring prolonged prostaglandin infusion despite absence of significant residual RVOT obstruction, coil occlusion of PDA and transcatheter closure of interatrial communication later in those patients who have attained adequate RV growth offers the exciting prospect that a small number of these patients may be treated entirely in the catheterization laboratory.

The infrequency with which this anomaly occurs makes it difficult to draw strong conclusions from a single institution, but our results argue for further investigation and application of this technique.

Acknowledgment

We greatly appreciate the help of Muhaneeza Hanafiah in the preparation of the manuscript.

Reprint requests and correspondence: Dr. Mazeni Alwi, Department of Cardiology, Institut Jantung Negara, 145, Jalan Tun Razak, 50400 Kuala Lumpur, Malaysia. E-mail: mazeni@ijn.com.my.

REFERENCES

1. Patel RG, Freedom RM, Moes CAF, et al. Right ventricular volume determinations in 18 patients with pulmonary atresia and intact ventricular septum: analysis of factors influencing right ventricular growth. Circulation 1980;61:428–40.
2. Lewis AB, Well W, Lindesmith GC. Evaluation and surgical treatment of pulmonary atresia and intact ventricular septum in infancy. Circulation 1983;67:1318–23.
3. Calder AL, Co EE, Sage WD. Coronary arterial abnormalities in pulmonary atresia with intact ventricular septum. Am J Cardiol 1987;59:436–42.
4. Mainwaring RD, Lamberti JL. Pulmonary atresia with intact ventricular septum: surgical approach based on ventricular size and coronary anatomy. J Thorac Cardiovasc Surg 1993;106:733–8.
5. De Leval M, Bull C, Stark J, Anderson RH, Macartney FJ. Pulmonary atresia and intact ventricular septum: surgical management based on revised classification. Circulation 1982;66:272–80.
6. Qureshi SA, Rosenthal E, Tynan M, Anjos R, Baker E. Transcatheter laser assisted pulmonic valve dilation in pulmonic valve atresia. Am J Cardiol 1991;67:428–31.
7. Rosenthal E, Qureshi SA, Chan KC, et al. Radiofrequency-assisted balloon dilation in patients with pulmonary valve atresia and on intact ventricular septum. Br Heart J 1993;69:347–51.
8. Fisher EA, Goldman ME. Simple, rapid method for quantification of tricuspid regurgitation by two-dimensional echocardiography. Am J Cardiol 1989;63:1375–8.
9. Coles JG, Freedom RM, Lightfoot NE, et al. Long term results in neonates with pulmonary atresia and intact ventricular septum. Ann Thorac Surg 1989;47:213–7.
10. Parsons JM, Rees MR, Gibbs JL. Percutaneous laser valvotomy with balloon dilation of the pulmonary valve as primary treatment for pulmonary atresia. Br Heart J 1991;66:36–8.
11. Cobanoglu A, Metzdorff MT, Pinson CV, Grun GL, Sunderland CO, Starr A. Valvotomy for pulmonary atresia with intact ventricular septum. J Thorac Vasc Surg 1985;89:482–90.
12. Joshi SV, Brawn WJ, Mee RBB. Pulmonary atresia with intact ventricular septum. J Thorac Cardiovasc Surg 1986;91:92–9.
13. Foker JE, Braunlin FA, St Cyr JA, Hunter D, Moller JH, Ring WS. Management of pulmonary atresia with intact ventricular septum. J Thorac Cardiovasc Surg 1986;92:706–15.
14. Hawkins JA, Thorne JK, Boucek MM, et al. Early and late results in pulmonary atresia and intact ventricular septum. J Thorac Cardiovasc Surg 1990;100:492–7.
15. Freedom RM, Wilson G, Trusler GA, Williams WG, Rowe RD. Pulmonary atresia and intact ventricular septum: a review of the anatomy, myocardium and factors influencing right ventricular growth and guidelines for surgical intervention. Scand J Thorac Cardiovasc Surg 1983;17:1–28.
16. Hanley FL, Sade RM, Blackstone EH, Kirklin JW, Freedom RM, Nanda NC. Outcomes in neonatal pulmonary atresia with intact ventricular septum: a multi-institutional study. J Thorac Cardiovasc Surg 1993;105:406–27.
17. Alboliras ET, Julsrud PR, Danielson GK, et al. Definitive operation for pulmonary atresia with intact ventricular septum: results in twenty patients. J Thorac Cardiovasc Surg 1987;93:454–64.
18. Pawade A, Capuani A, Penny DJ, Karl TR, Mee RB. Pulmonary atresia with intact ventricular septum: surgical management based on right ventricular infundibulum. J Cardiac Surg 1993;8:371–83.
19. Reddy VM, Mc Elhinney DB, Silverman NH, Marianeschi SM, Hanley FL. Partial biventricular repair for complex congenital heart defects: an intermediate option for complicated anatomy or functionally borderline right complex heart. J Thorac Cardiovasc Surg 1998; 116:21–7.
20. Freedom RM, Perrin D. The right ventricle: morphological consideration. In: Freedom RM, ed. Pulmonary Atresia With Intact Ventricular Septum. New York: Futura Publishing Compay Inc., 1989; 53–76.
21. Kanter KR, Pennington DG, Nouri S, Chen SC, Jureidine S, Balfor I. Concomitant valvotomy and subclavian-main pulmonary artery shunt in neonates with pulmonary atresia and intact ventricular septum. Ann Thorac Surg 1987;43:490–4.
22. Leung MP, Lo RNS, Cheung H, Lee J, Mok CK. Balloon valvuloplasty after pulmonary valvotomy for babies with pulmonary atresia and intact ventricular septum. Ann Thorac Surg 1992;53:864–70.
23. Gournay V, Piechaud JF, Delogu A, Sidi D, Kachaner J. Balloon valvotomy for critical stenosis or atresia of pulmonary valve in newborns. J Am Coll Cardiol 1995;26:1725–31.
24. Justo RN, Nykanen DG, Williams WG, Freedom RM, Benson LN. Transcatheter perforation of the right ventricular outflow tract as

initial therapy for pulmonary valve atresia and intact ventricular septum in the newborn. Cathet Cardiovasc Diagn 1997;40:408–13.
25. Ovaert C, Qureshi SA, Rosenthal E, Baker EJ, Tynan M. Growth of the right ventricle after successful transcatheter pulmonary valvotomy in neonates and infants with pulmonary atresia and intact ventricular septum. J Thorac Cardiovasc Surg 1998;115:1055–62.
26. Gibbs JL, Blackburn ME, Uzun D, Dickinson DF, Parsons JM, Chatrath RR. Laser valvotomy with balloon valvuloplasty for pulmonary atresia with intact ventricular septum: five years experience. Heart 1997;77:225–8.
27. Hausdorf G, Schneider M, Lange P. Catheter creation of an open outflow tract in previously atretic right ventricle outflow tract associated with ventricular septal defect. Am J Cardiol 1993;72:354–6.
28. Rosenthal E, Qureshi SA, Kakadekar AP, Anjos R, Baker EJ, Tynan M. Technique of percutaneous laser assisted valve dilation for valvar atresia in congenital heart disease. Br Heart J 1993;69:556–62.