INTRODUCTION

The management strategy of pulmonary atresia and intact ventricular septum (PA-IVS) is determined mainly by the morphologic substrates of right ventricle and coronary circulation.[1-6] It is generally agreed that establishing continuity between right ventricle and pulmonary trunk is desirable in patients with adequate right ventricular morphology. Before 1990, the results of treatment for patients with PA-IVS were discouraging. In the past few years, survival of those patients has improved because of better understanding of right ventricular morphology and use of prostaglandin.[6-8] With the advancement of transcatheter treatment, transcatheter valvotomy for patients with PA-IVS has been increasingly performed.[9-20] However, reports on outcomes of transcatheter valvotomy in PA-IVS are limited. Here the outcomes of PA-IVS patients treated with transcatheter technique are described with analysis of the predictive factors for successful outcome.

PATIENTS & METHODS

Patients: From June 1995 till January 1998, 20 neonates were diagnosed with membranous atresia of the pulmonary valve with intact ventricular septum in this institution, of whom 14 underwent an attempted pulmonary valvotomy. All 14 patients had a tripartite right ventricle and a Z-value of tricuspid valve >-3. Transcatheter valvotomy was not attempted in 6 neonates because of right ventricular dependent coronary circulation in 4 and severe hypoplasia of right ventricle in 2 (Z-value of tricuspid valve <-3).[1,8] Of the 6 patients, 4 were managed excluding right ventricular decompression by a systemic-to-pulmonary artery shunt and 2 underwent a right ventricular outflow patch and a concomitant shunt. Of the 14 patients, their ages ranged from 1 day to 28 days. Body weights ranged from 2.8 to 3.6 kg. All 14 patients required continuous infusion of prostaglandin E_1 (PGE_1) on admission because of hypoxemia. One was intubated with ventilatory support. Eight patients had symptoms of heart failure requiring anti-failure treatment.

Measurements: Each patient underwent an echocardiography using Acuson XP128 on admission. The tricuspid valve annulus was measured at end diastole on an apical four-chamber view. A Z-value of tricuspid valve was obtained according to the normogram of Hanley.[8] Right and left ventricular endocardial surface areas on apical four-chamber view were measured either on line or on play back S-VHS video tape with the same echocardiographic equipment. The ratio of right to left ventricular area was calculated to provide a rough estimation of right ventricular size.[20] Pulmonary valve annulus was measured on a right ventricular angiogram from anteroposterior and lateral projection using a known catheter diameter as reference. A Z-value of

pulmonary valve was then determined. Echocardiography was repeated for each patient within 1 week following pulmonary valvotomy. The ratio of right to left ventricular area and pressure gradient across right ventricular outflow tract was obtained on the repeated echocardiography. During follow-up, echocardiogram was performed 3 months and 6 months after discharge, and every 6 months thereafter. Pressure gradient across the pulmonary valve was measured in each follow-up echocardiography. Presence of shunt at the atrial level was also evaluated with color flow mapping.

Technique of pulmonary valvotomy: After obtaining parental consents, cardiac catheterization and angiogram were performed. One dose of cephradine arginine 25 mg/kg was given intravenously 30 minutes prior to cardiac catheterization. Local anesthesia with xylocaine and sedation using valium or ketamine were administered. Femoral venous and arterial access were obtained. Heparin (50 U/kg) was given. Patients were continuously monitored with electrocardiography and oximeter during cardiac catheterization. A complete hemodynamic study was performed in each patient. A biplane right ventriculogram was obtained with anteroposterior and lateral views to evaluate the morphology and size of right ventricular structures. The pulmonary valve annulus was measured on play-back digitalized angiogram using a known catheter diameter as reference. An aortic root angiogram was routinely performed to delineate any coronary artery anomalies and obtain images of pulmonary arteries.

Before March 1996, the soft end of a 0.038 or 0.035 superstiff guidewire (Medi-tech Watertown, MA) was used to perforate the pulmonary valve. After March 1996, perforation of pulmonary valve was performed using a radiofrequency guidewire.(PA 120, Osypka, Germany) Pulmonary valvotomy was attempted with the floppy end of a 0.038 or 0.035 superstiff guidewire in 4 patients and with a radiofrequency guidewire in 10 patients, respectively. A Berman catheter (Arrow International Inc., Reading, PA) was positioned via the femoral artery in the main pulmonary artery above the atretic pulmonary valve as a reference. A 4F or 5F right coronary catheter (Cordis, Inc, Miami, FL) or head hunter catheter (Cook, International Inc.) was advanced to right ventricular outflow tract. Hand injection of contrast media was performed several times to ensure the position of the end-hole catheter just beneath the atretic valve. The floppy end of a superstiff guidewire or a radiofrequency guidewire was advanced through the end-hole catheter to perforate the valve. The radiofrequency guidewire was connected to a radiofrequency generator (HAT 300, Osypka, Germany) with radiofrequency energy delivered at a setting of 5 watts for a few seconds. A gentle force was applied to the radiofrequency guidewire while the energy was being delivered. After perforation of the valve, the end-hole catheter was advanced to the

pulmonary trunk along the guidewire. In some occasions when an end-hole catheter failed to cross the valve, a gooseneck snare (Microvena, White Bear Lake, MN) was advanced via femoral artery crossing the ductus to the pulmonary trunk to grasp the tip of the radiofrequency guidewire. Then, the end-hole catheter was pushed across the valve along the radiofrequency guidewire, and a J-tip 0.035 guidewire was exchanged with the radiofrequency guidewire. A low profile balloon catheter (Schneider) 1.2 1.5 times of the pulmonary annulus was advanced to pulmonary valve for dilation. The balloon was inflated several times with a pressure of 7 to 10 atmospheres. Following balloon valvotomy, hemodynamic studies were repeated. Then, a second right ventriculogram was performed to demonstrate infundibular and valvular anatomy. If the residual pressure gradient across pulmonary valve was above 40 mm Hg or forward flow to pulmonary arteries was inadequate, a repeated valvuloplasty using a larger size balloon was performed. Following the procedure, patients were monitored with pulse oximeter and electrocardiography. Three doses of cephradine arginine were administered after the procedure. PGE_1 was gradually withdrawn if systemic oxygen saturation was above 80% without use of O_2. In patients who remained PGE_1 dependent for more than 2 weeks or had significant Doppler pressure gradient, cardiac catheterization was repeated to evaluate the right ventricular structure and hemodynamics. Based on the outcomes of transcatheter valvotomy, the patients were classified into 2 groups. Group Ⅰ patients are those in whom transcatheter valvotomy alone was a definitive treatment. Group Ⅱ patients are those who required surgery despite an initial successful valvotomy.

Statistics: All data were expressed as mean±standard deviation (SD). Paired t-test was used to compare the mean systolic right ventricular pressure and right to left ventricular area ratio before and after balloon valvuloplasty. Mann-Whitney rank-sum test was used to assess the significance of differences in the following parameters including tricuspid and pulmonary valve Z-value, right to left ventricular area ratio and hemodynamic characteristics between the 2 groups. A p value <0.05 was regarded as statistically significant.

RESULTS

Immediate Results: The attempt to perforate the atretic pulmonary valve was successful in 11 patients (11/14, 78%): 2 with floppy end of a superstiff guidewire and 9 with a radiofrequency guidewire; it failed in 3 patients. Of the 11 neonates with successful perforation of the atretic pulmonary valve, balloon valvuloplasty could be performed in all 11 patients. Following balloon valvuloplasty, the mean right ventricular systolic pressure decreased from 124±24 to 60±15 mm Hg. ($p<0.01$) The mean pressure gradient between right ventricle and pulmonary artery immediately following valvotomy was 19±9 mm Hg (ranging from 6 to 36 mm Hg). A repeated

right ventriculogram showed good forward flow from right ventricle to pulmonary arteries in all 11 patients (Figure 1). One week following valvotomy, the mean right to left ventricular area ratio estimated with echocardiography in these 11 patients decreased from 0.66±0.16 to 0.61±0.31. ($p<0.05$) Information regarding the 11 patients are summarized in Table Ⅰ.

Outcomes: PGE_1 could not be withdrawn in 5 patients within 14 days of whom 1 underwent a right ventricular outflow tract patch because the right ventricle was judged to be moderately hypoplastic with infundibular stenosis on echocardiography. The remaining 4 patients underwent a repeated cardiac catheterization and angiogram to evaluate the hemodynamics and morphology of right ventricular outflow tract. Judging from hemodynamic data and angiogram, 1 had significant infundibular stenosis (Figure 2), 1 had recurrent valvular pulmonary stenosis, and 2 were judged to have impaired compliance of right ventricle on the basis of a low right ventricular pressure and good forward flow from right ventricle to pulmonary arteries. A repeated balloon valvuloplasty was performed in 2 patients who had right ventricular outflow tract stenosis resulting in significant reduction in pressure gradient and right ventricular systolic pressure in 1 and no change in right ventricular pressure in another who had infundibular stenosis. Since PGE_1 could not be discontinued in the 2 patients 2 weeks following a second intervention, these 2 patients underwent a right ventricular outflow tract patch and 1 had a concomitant systemic-to-pulmonary artery shunt. Of the remaining 2 patients who were judged to have impaired right ventricular compliance, 1 was able to be weaned off PGE_1 4 weeks later and was discharged with a systemic O_2 saturation above 90%, the other underwent a modified Blalock-Taussig shunt but developed right ventricular outflow tract stenosis detected with Doppler echocardiography 1 week after the shunt. The latter then underwent a repeated balloon dilatation but without significant change in right ventricular pressure (145/3→110/2 mm Hg) and then underwent a successful right ventricular outflow patch.

Six patients had successful withdrawal of PGE_1 within 2 weeks. The sole patient who was intubated continued to have symptoms of heart failure despite antifailure treatment and discontinuation of PGE_1. He died of heart failure complicated with sepsis 10 days later. Excluding this mortality, a total of 6 patients (Group Ⅰ) achieved a two-ventricle repair by transcatheter valvotomy alone and have not required operation and 4 patients (Group Ⅱ) required a right ventricular outflow tract patch with or without a systemic-to-pulmonary artery shunt following transcatheter treatment.

Group Ⅰ vs. Group Ⅱ comparisons: Comparing the hemodynamic characteristics including right ventricular systolic pressure before and after valvotomy and pressure gradient across right ventricular outflow tract immediately after valvotomy, there were

no significant differences in hemodynamic characteristics between the 2 groups. (Table Ⅱ). Comparing the right ventricular morphologic characteristics between the 2 groups, Group Ⅰ patients had a significantly higher mean Z-value of tricuspid and pulmonary valve. (0.52±0.37 vs. -1.25±0.48, $p<0.05$ and -3.47±0.59 vs. -5.43± 0.94, $p<0.05$, respectively) The ratio of right to left ventricular area measured prior to intervention is significantly smaller in Group Ⅱ patients. (0.73±0.06 vs. 0.49±0.03, $p<0.05$). (Table II)

Complications: Perforation of right ventricular outflow tract occurred in 1 patient in whom the attempt at pulmonary valvotomy with a radiofrequency guidewire was not successful. The patient was stabilized after emergent surgical drainage of pericardial fluid. A right ventricular outflow tract patch and a shunt were performed 4 days later, but the patient did not survive the surgery. Blood transfusion was required in 2 patients. Three patients developed supraventricular tachycardia during manipulation of the catheter in the right side of the heart, which was treated with propranolol or adenosine. One had total occlusion of bilateral femoral veins found at second cardiac catheterization because a Doppler pressure gradient of 45 mm Hg was noted.

Follow-up: All of the 10 patients in both groups were available for follow-up. All had systemic oxygen saturation ＞92% during follow-up. One in Group Ⅱ had mild tachypnea and was treated with digoxin and diuretics. One patient in Group Ⅰ who had an echo pressure gradient of 45 mm Hg underwent a repeated cardiac catheterization and angiogram via internal jugular vein 6 months later with pressure gradient reduced to 30 mm Hg. After follow-up for 7 to 35 months (18±10.3 months), all the 10 patients were in good condition with a mean pulmonary valve pressure gradient of 17 ±15 mm Hg (ranging from 0 to 45 mm Hg). The modified Blalock-Taussig shunt spontaneously occluded in 1 patient. No patient had a shunt at the atrial level on the most recent echocardiogram.

DISCUSSIONS

Among PA-IVS neonates, cyanosis and heart failure are the predominant symptoms. If left untreated, many such patients may die in early infancy. Management strategies of PA-IVS are based on morphology and size of right ventricle and associated coronary artery anomalies.[1-6] For patients with major sinusoidal coronary artery communications or unfavorable right ventricular morphology, a systemic-to-pulmonary shunt is recommended. Decompression of right ventricle has been recommended for patients who have a tripartite right ventricle with a tricuspid valve Z-value greater than -2.4 or -3 and without major sinusoidal coronary artery communications.[1,7,8] Despite recent advancement in surgical treatment for congenital heart disease, surgical mortality for PA-IVS remains high.[1,7,8]

Transcatheter treatment for PA-IVS: Transcatheter pulmonary valvotomy with

laser guidewire was introduced in 1991,[9,10] since then several reports with variable success have been published.[11-13] A recent article reported a high initial success rate of pulmonary valvotomy (8/9, 89%) with a laser guidewire.[13] However, in that series 2 out of 8 patients with an initial successful valvotomy required infundibular resection and closure of atrial septal defect during follow-up. Therefore, 67% (6/9) patients were successfully treated with transcatheter valvotomy alone. Laser guidewire seems quite effective for perforation of atretic pulmonary valve, however the cost of laser generator and equipment of protective goggles were the major drawback to its use. Radiofrequency guidewire has been successfully applied in assisting pulmonary valvotomy in patients with membranous pulmonary atresia.[12,14-17] Perforation of atretic pulmonary valve using a radiofrequency guidewire is generally achieved by applying gentle force with a low energy, however perforation of right ventricular outflow tract with a radiofrequency guidewire is not infrequent, but can be managed by pericardiocentesis or surgical repair.[14,15,17] This complication occurred in our early experience here. Cautious ensuring the position of the radiofrequency guidewire by repeated hand injection of contrast media and use of a catheter in main pulmonary artery as a reference may reduce the incidence of infundibular perforation. In a recent report, the echocardiogram has been used as a guide for pulmonary valvotomy, which added more safety in pulmonary valvotomy.[21] A radiofrequency catheter has recently been used successfully in perforation of the atretic pulmonary valve. The advantages of a radiofrequency catheter are that the tip of the catheter is deflectable and it is easily manipulated to a position beneath the pulmonary valve, but the number of patients treated with this technique is limited.[18,19,22] In addition to laser guidewire or radiofrequency guidewire or catheter, the stiff end of a 0.014, 0.018 or 0.021 guidewire has been used to perforate the atretic pulmonary valve.[16,23-25] We have used the soft end of a superstiff guidewire in 4 patients, however there were 2 patients in whom puncture of the atretic valve was not successful. The success rate of perforation of the atretic valve with a guidewire seems to be lower than that with a radiofrequency or laser guidewire. Radiofrequency guidewire is considered to be a convenient, useful and less expensive tool in perforation of atretic pulmonary valve.

Impact of right ventricular structural size on ultimate success of transcatheter treatment: In the current series, transcatheter pulmonary valvotomy alone provided definitive success in 55% (6/11) PA-IVS cases. In accordance with other reports, [7,8] a larger Z-value of tricuspid valve predicted higher probability of transcatheter valvotomy alone as a definitive treatment. The Z-value of tricuspid valve and right to left ventricular area ratio were highly correlated with the size of right ventricular cavity.[8,20] The Z-value of pulmonary valve is also a predictive factor for successful outcome of pulmonary valvotomy. In this study, tricuspid valve Z-value $\geqq$-0.1,

pulmonary valve Z-value≧-4.1 and a right to left ventricular area ratio≧0.65 indicated transcatheter pulmonary valvotomy alone can be a definitive treatment. All patients with a tricuspid valve Z-value≦-0.8, pulmonary valve Z-value ≦-4.2 and right to left ventricular area ratio ≦0.54 have required a shunt and or right ventricular outflow tract patch. In the current series, a two-ventricular repair was achieved by transcatheter valvotomy and/or surgery in all patients with a tricuspid valve Z-value above -1.8 and pulmonary valve Z-value above -6.5.

Schmidt et al. has reported right ventricular stroke volume was reduced by one-third in PA-IVS patients 5 days after pulmonary valvotomy, then recovered 19 days later.[20] In the current series, reduction in right ventricular volume 1 week following valvotomy was observed, and group II patients seemed to have significantly smaller right ventricular stroke volume following valvotomy because of their relatively smaller right ventricular cavity. Development of infundibular stenosis following valvotomy has been documented in several articles.[7,13,15] In this series, infundibular stenosis was documented with echocardiography and/or angiography in 3 patients of group II 7 to 28 days following pulmonary valvotomy, of whom all had a tricuspid valve Z-value≦-0.9 and pulmonary valve Z-value≦-5.5. Inderal was administered in the 3 patients, but without any effect. Resection of infundibular muscle or placement of outflow tract patch, or both may be required.[7,26] In retrospect, a right ventricular outflow tract patch may not be necessary but a shunt was required in the remaining patient of Group II who did not develop infundibular stenosis.

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