肺動脈閉鎖病人接受瓣膜擴張術擴右心室出口整形治療發育不良之右心室成長之研究

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NSC 90-2314-B-002-392
執行期限：89 年 8 月 1 日至 90 年 7 月 31 日

一、中文摘要

目的:

本研究之目的在於探討肺動脈閉鎖且無心室中隔缺損之病人在瓣膜整型後其原先發育不良的右心室是否會生長。

方法與結果：

從 1995 年 6 月至 2001 年 8 月，在本院有 44 位新生兒患有肺動脈瓣閉鎖且心室中隔無缺損，接受心導管攝影及檢查，其中 7 例由於右心室嚴重發育不全(三尖瓣 Z 值< -4)或有冠狀動脈 ◦管無法做肺動脈瓣整形，成為對照組，其餘 37 例接受右心室減壓，包括原先使用心導管 31 例，右心室出口整形手術 6 例, 31 例初期成功 28 例(4 例使用引導線，24 例使用高周波導線)，失敗 3 例, 最終成功 20 例, (6 例需開刀，2 例死亡), 因此手術共有 3 例死亡，34 例包括 20 例心導管治療，14 例手術，供作右心室成長之分析，34 例病人之血氧飽和度均在 75%以上。34 例，追蹤平均 1.4 年，三尖瓣平均 Z 值由原先的 –1.7 ± 1.2 變成， –0.8 ± 1.3 (P <0.01, parted -t)，而對照組 7 例的三尖瓣的 Z 值沒有改變，平均值由原先–3.2 ± 0.6 成為 -3.6 ± 0.8。(P >0.05)

結論：

肺動脈瓣閉鎖且無心室中隔缺損的病人，若在新生兒期建立右心室與肺動脈的通路，發育不良的右心室是有機會成長。

關鍵詞：

肺動脈瓣閉鎖無心室中隔缺損，經心導管治療，右心室

Abstract

Between June 1995 and August 2001, 44 neonates with PA/IVS underwent cardiac catheterization and angiography. Thirty-seven patients tricuspid valve Z-value ＞-4 (ranging from –3.2 to 0.6, mean -1.4 ± 0.8) without significant sinusoid underwent right ventricle decompression in the neonatal period. Seven patients in whom decompression of right ventricle was considered as contraindicated because of severe hypoplasia of right ventricle tricuspid valve Z < -4 or sinusoid-coronary communications were used as controls. Of the 37 patients, transcatheter pulmonary valvotomy was attempted in 31 and was successful in 28 patients: 4 with a guidewire and 24 with a radiofrequency guidewire. Mean while, 6 neonates with PA/ IVS underwent right ventricular outflow tract reconstruction with or without a shunt. Six of the 28 patients further required a right ventricular out flow tract patch, despite an initial success in pulmonary valvotomy. Of the 37 patients, there were 3 mortalities.

Therefore, a total of 34 patients who underwent decompression of right ventricle (14 RVOT patch and 20 transcatheter valvotomy) were available for follow up. A total of 34 patients who were discharged with systemic O_2 saturation above 75% were enrolled in this sutudy. After a follow-up period ranging from 2 to 62 months, the mean Z value of tricuspid value measured an echocardiography increased from –1.7 $\pm$ 1.2 to –0.8 $\pm$ 1.3 during follow-up. However, of the 7 patients managed with a modified B-T shunt, the Z value of tricuspid valve remained unchanged. (–3.2 $\pm$ 0.6 vs. –3.6 $\pm$ 0.8, P >0.05)

Conclusion：Right ventricle could grow in PA-IVS patients if patients decompression could be performed in early infancy.

Key words：pulmonary atresia and intact ventricular septum, transcatheter treatment, right ventricle, radiofrequency guidewire.

二、緣由與目的

Introduction: Hypoplasia of right ventricle is frequently present in PA-IVS patients. 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] Decompression of right ventricle is contraindicated in patients with unfavorable right ventricular morphology and presence of sinusoid. Before 1990, the results of surgical treatment for patients with PA-IVS were discouraging. However, 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] Currently, transcatheter valvotomy is recognized as an alternative treatment for those with adequate right ventricle. The changes right ventricular size following pulmonary valvotomy remains unknown. We investigate the growth potential of right ventricle in patients with PA/ IVS. The follow-up results in those patients was also reported.

Methods:

Patients. From June 1995 till August 2001, 44 neonates were diagnosed with membranous atresia of the pulmonary valve with intact ventricular septum in this institution, of whom 37 underwent attempted decompression of right ventricle (RV). All 44 patients required continuous infusion of prostaglandin E_1 (PGE_1) on admission because of hypoxemia. Four was intubated with ventilatory support. Eleven patients had symptoms of heart failure requiring anti-failure treatment. All 37 patients had a tripartite right ventricle and a Z-value of tricuspid valve >-4. Decompression of RV was not attempted in 7 neonates because of right ventricular dependent coronary circulation in 4 and severe hypoplasia of right ventricle triasspid valve (Z < -4) in 3. The 7 patients, who were managed by a systemic-to-pulmonary artery shunt, were included as controls. Of the 37 patients, their ages ranged from 1 day to 28 days. Body weights ranged from 2.8 to 3.6 kg. Of the 37 patients, 31 underwent attempted transcatheter valvotomy and 6 underwent surgical valvotomy as initial treatment. (Figure 1)

Methods. In the beginning of this study, floppy tip of a 0.035 straight guidewire was used for perforation of the pulmonary valve. Since August 1996, radiofrequency (RF) guidewire was applied for perforation of atretic pulmonary valve. The RF guidewire was connected to a generator (PA120). A 5 watt was applied to heat and perforated the atretic valve. After perforation of the valve, a choice PT coronary guidewire was advanced through the perforated hole in the atretic valve. Following retriering the RF guidewire, a VACS balloon catheter was advanced along the PT coronary guidewire to dilate the valve. After balloon valvuloplasty, repeat right rentriculram was

performed. The residual pressure gradient was measured.

三、結果

Results. The attempt to perforate the atretic pulmonary valve was successful in 28 patients out of 31 attempted (28/31, 90 %): 4 with floppy end of a superstiff guidewire and 21 with a radiofrequency guidewire; it failed in 3 patients. Of the 28 neonates with successful perforation of the atretic pulmonary valve, balloon valvuloplasty could be performed in all 28 patients. Of the 28 patients, there were 2 mortalities, 1early and 1 late mortality. Meanwhile, 9 patients including the 3 failure cases underwent right ventricular outflow tract reconstruction with or without a shunt. In the 28 patients PGE_1 could not be withdrawn in 6 patients within 28 days. Therefore, the 6 patients underwent a right ventricular outflow tract patch because the right ventricle was judged to be moderately hypoplastic with infundibular stenosis on echocardiography. A total of 15 patients underwent right ventricular outflow tract patch with 1 mortality. Twenty patients were treated with transcatheter pulmonary valvotomy. Thirty-four patients surived surgery or pulmonary valvotomy were available for follow-up. The mean Z value of TV in the 34 patients was -1.7 ± 1.2 before intervention or surgery. The mean Z value of tricuspid valve in the 7 patient managed by a shunt alone was -3.2 ± 0.6.

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. One who underwent surgery had an episode of MRSA sepsis.

Changes of right ventricular size following valvotomy.

All of the 34 patients in were available for follow-up. After a mean follow-up period 14 $\pm$ 0.7 year, the mean Z value of tricuspid valve increased from -1.7 ± 1.2 to -0.8 ± 1.3. In 7 patients without right ventricular decompression, the right ventricular size remained unchanged (Z valve of tricuspid valve -3.3 ± 0.6 v.s -3.6 ± 0.5, $P > 0.05$) during follow-up. All patients who underwent RV decompression had systemic oxygen saturation $>$75% during follow-up. After follow-up for 2 to 46 months, all except 1 were in good condition but 1 required a RVOT patch. Of the 34 patients, 20 were treated non-surgically as groupI and 14 underwent RVOT patch with or without a shunt as groupII. There was no statistical significance of mean tricuspid Z valve in the last echocardiographic measurement. (-0.7 ± 1.1 vs. -0.8 ± 1.4)

DISCUSSIONS

The therapeutic 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] The growth potential of hypoplastic right ventricle remains unknown.

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] 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] Radiofrequency guidewire is considered to be a convenient, useful and less expensive tool in perforation of atretic pulmonary valve.[22-25]

Impact of right ventricular structural size on ultimate success of transcatheter treatment: In the current series, transcatheter pulmonary valvotomy alone provided definitive success in 65% (20/31) 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 annulus could predict the development of infundibular stenosis.

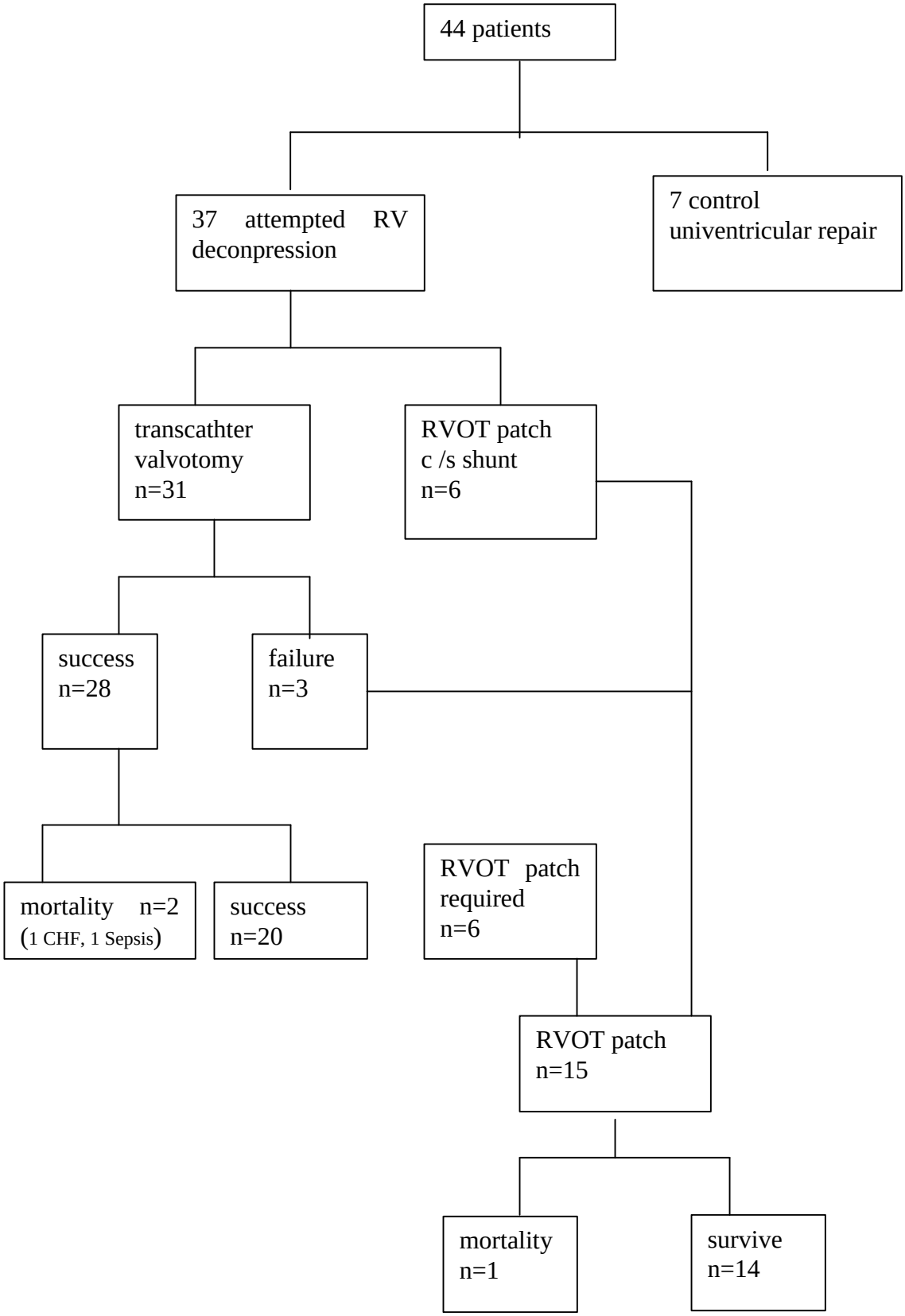
Diastolic dysfunction of right ventricle following valvotomy may be a problem which could be related to hyperplasia of endomyocardium of right ventricle. Regression of the hypertrophied muscle in right ventricle is accompanied by improvement in O_2 saturation in most patients with right ventricular outflow tract obstruction following pulmonary valvotomy. However, in some patients, the hypertrophied endocardium persisted and infundibular stenosis ensued that right ventricular outflow tract patch with or without a shunt was required. The long term outcome of infundibular stenosis required further investigation.

Growth potential of right ventricle in PA-IVS.

Growth potential of right ventricle was generally present in patients with a tricuspid value Z value > -4, who underwent a early decompression of right ventricle. There was no significant difference in the growth potential between the two subgroups who underwent right ventricular decompression with different approach. However, significant difference of growth potential was found between those who underwent decompression and those who were managed by a shunt only. In another study, no growth of the right ventricle size was documented in patients who underwent decompression with 2-ventricle circulation. More cases and centers should be involved to reach a consensus in this regard.

Acknowlegement:
This study was supported by a grant from National Science Council, Republic of China. NSC 89-2314-B-002-038.

Figure 1.


44 patients
37 attempted RV deconpression
7 control univentricular repair
transcathter valvotomy n=31
RVOT patch c /s shunt n=6
success n=28
failure n=3
mortality n=2 (1 CHF, 1 Sepsis)
success n=20
RVOT patch required n=6
RVOT patch n=15
mortality n=1
survive n=14

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