Transcatheter Laser-Assisted Balloon Valvulotomy as Primary Treatment in Newborn with Pulmonary Atresia and Intact Ventricular Septum

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Abstract

We report our initial experience and the first case in Thailand in successfully performing transcatheter laser perforation of membranous pulmonary valve and subsequent balloon dilations of the valve in a 14-day-old baby with pulmonary atresia and intact ventricular septum. After the procedure, right ventricular angiograms revealed anterograde flow across the pulmonary valve. There was no major complication. Doppler echocardiography one week later demonstrated a pressure gradient across the pulmonary valve of 30 mmHg and right ventricular systolic pressure of 60 mmHg. Her peripheral oxygen saturation improved from 70's immediately after the procedure to 92-97 per cent at one-month after the procedure.

The management of patients with pulmonary atresia and intact ventricular septum depends upon individual cardiac anatomy, especially right ventricular coronary sinusoids[1-3]. In patients with a near-normal size, tripartite right ventricle and confluent pulmonary arteries, surgical pulmonary valvulotomy alone may achieve satisfactory results [4-6]. The therapeutic aims are to increase pulmonary blood flow and promote growth of the smaller right ventricle for future palliation and/or repair. With the advent of new interventional techniques, a number of congenital heart diseases can be treated without surgery. Since 1990, some limited experiences with percutaneous valvulotomy of an imperforated pulmonary valve using the laser or radiofrequency guidewire or catheter have been reported[7-10].

We report our initial experience and the first case in Thailand of successful transcatheter laser-assisted balloon pulmonary valve dilation in a newborn with pulmonary atresia and intact ventricular septum.

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CASE REPORT

A female term neonate (birth weight of 3,360 g) developed central cyanosis at the age of 8 hours. Her peripheral oxygen saturation in room air was 63 per cent. Echocardiography revealed membranous pulmonary atresia with an intact ventricular septum. The pulmonary arteries were confluent and of normal size. The tricuspid annulus measured 10 mm in diameter (the Z-value of -1)[2]. The right ventricle was tripartite with the estimated suprasystemic systolic pressure of 110 mmHg. Color-flow mapping demonstrated a restrictive ductus arteriosus dependent pulmonary circulation. There was moderate tricuspid regurgitation. Prostaglandin E_1 (PGE_1) was administered as soon as the diagnosis was established and improved her oxygen saturation to the high 80's. It was decided to do the separated two-ventricle system.

Cardiac catheterization was performed percutaneously by the right femoral venous and arterial routes at the age of 14 days. She was orally sedated with chloral hydrate 75 mg/kg and midazolam 0.1 mg/kg intravenously twice during the procedure. Initial right and left ventricular angiograms confirmed the echocardiographic diagnosis. The right ventricular angiograms in the anteroposterior and lateral views demonstrated the atretic membranous pulmonary valve with a long infundibulum (Fig. 1). A 5 Fr right Judkins catheter was then positioned in the blind infundibulum, pointing posteriorly in the line with the pulmonary trunk. A 4 Fr right Judkins catheter was inserted through the arterial sheath and positioned in the main pulmonary artery through the ductus arteriosus. The venous and arterial catheters were positioned close to the pulmonary valve. The venous one was used to pass the laser guidewire and the other was used as a landmark. A series of angiograms, undertaken in each and both catheters in several projections to ensure that the venous catheter was in a stable and suitable position. Once this position was established, a 0.018 inch floppy laser guidewire (Prima^{TM} coronary, Spectranetics, Colorado, U.S.A.) was passed through the catheter until it came in contact with the pulmonary atretic membrane. The proximal wire was connected to the CVX-300® Excimer Laser System. The laser energy was delivered continuously at 60 mJ/mm^2, rate 25 pulses/sec. The atretic valve was perforated within 5 seconds. Then the laser wire was advanced across the valve through ductus arteriosus to the proximal descending aorta (Fig. 2). After several attempts, we could not advance the wire. We tried to advance the predilator catheter along the laser wire but it was not possible. The wire was snared and pulled down to just above the bifurcation of the common iliac artery. However, we were unable to pass the catheter or coronary balloon dilatation

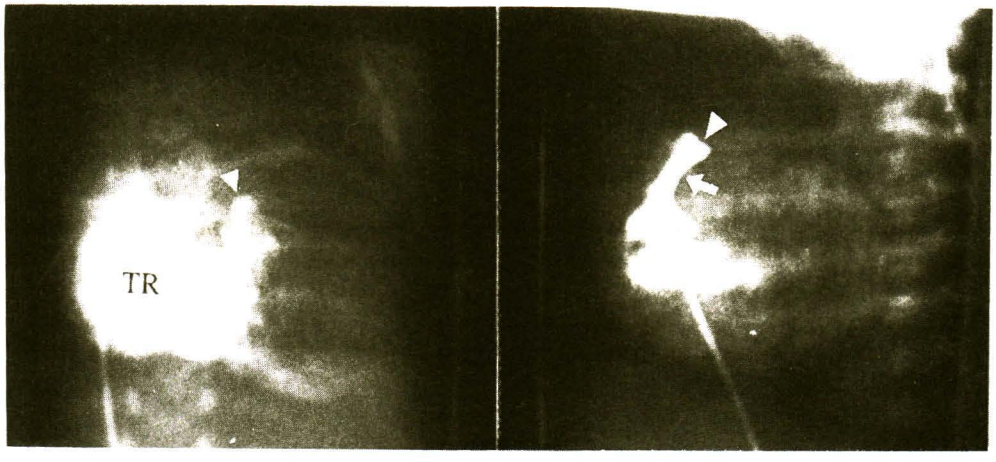

Fig. 1. The right ventricular angiograms, before the procedure in A) anteroposterior and B) lateral views demonstrated the hypertrophic and tripartite chamber, atretic pulmonary valve (arrow head) with a long infundibulum (arrow), and severe tricuspid regurgitation (TR).

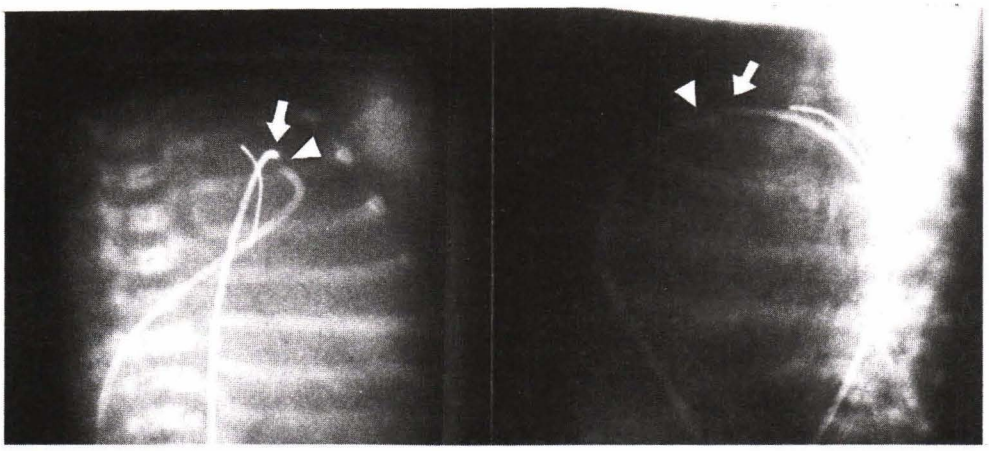

Fig. 2. The plain films in A) anteroposterior and B) lateral views demonstrated the position of the catheter in blind infundibulum from venous route (arrow head) and in main pulmonary artery through patent ductus arteriosus from arterial route (arrow). The laser wire perforated the pulmonary valve and positioned in the proximal descending aorta through the patent ductus arteriosus.

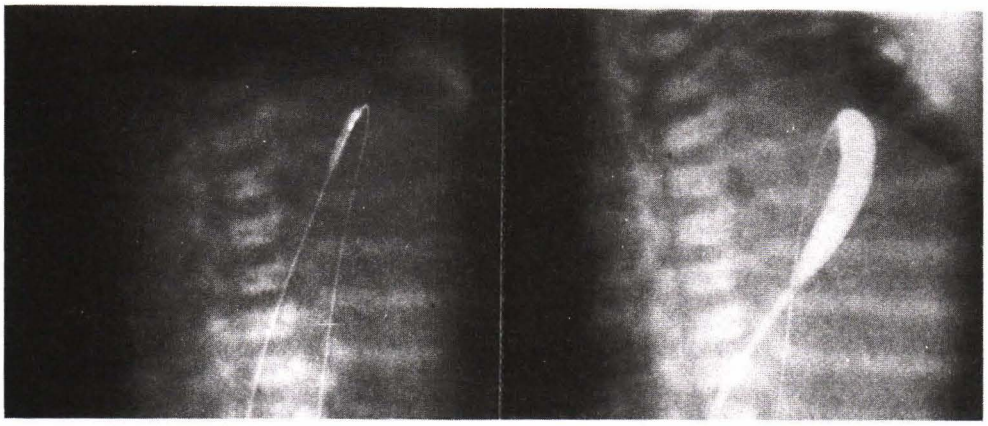

Fig. 3. The plain films in anteroposterior view of the successful balloon dilation of the pulmonary valve with A) a 2-mm coronary artery balloon catheter and B) a 6-mm Mansfield balloon catheter.

catheter over the wire. Again the wire was snared and pulled out through the arterial sheath as a rail for valvuloplasty[11]. At this time, the serial dilations of the valve with a 2-mm, 4-mm coronary artery balloon catheter and 6-mm Mansfield balloon catheter were easily performed (Fig. 3). The immediate right ventricular systolic pressure was not significantly reduced after the procedure, reflecting severe infundibular stenosis. The repeated right ventricular angiograms showed an anterograde flow into the pulmonary arteries without an extravasation of contrast outside the heart (Fig. 4). PGE_1 was discontinued. The procedure time was 3 hours. We transfused pack red cells to the baby

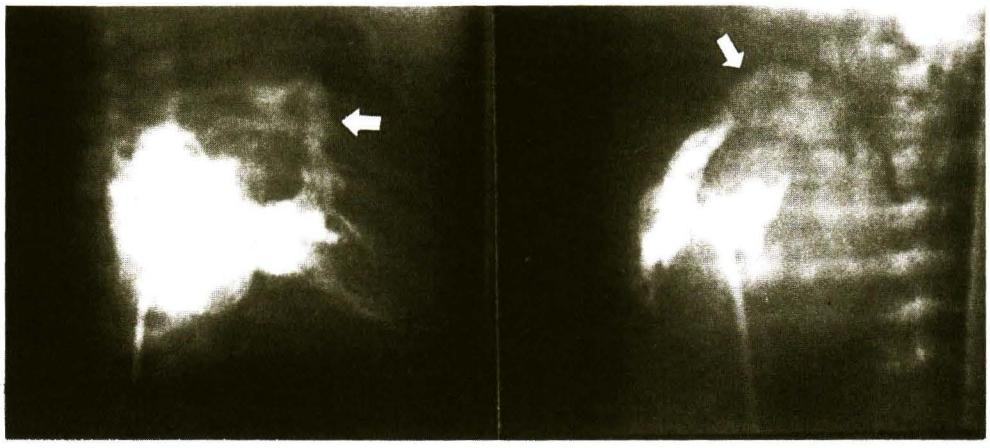

Fig. 4. The right ventricular angiograms, after the procedure in A) anteroposterior and B) lateral views demonstrated an anterograde flow into the pulmonary arteries (arrow).

because of several changes of the catheters, inspite of no significant change in hemodynamics. The episode of atrial flutter-fibrillation occurred at the end of the procedure, converted with 1 Joule energy of cardioconversion.

Echocardiography was performed at one day and one week after the procedure and revealed the anterograde flow across the pulmonary valve without a significant regurgitation. The pressure gradient across the pulmonary valve was 30 mmHg. The estimated right ventricular systolic pressure was reduced to 60 mmHg. She was discharged home 10 days after the procedure with the peripheral oxygen saturation in the high 70's. One month later, her peripheral oxygen saturation was 92-97 per cent and body weight was 4,620 g.

DISCUSSION

The prevalence for pulmonary atresia with intact ventricular septum is 0.083 per 1000 live births[12], accounting for 0.71[13]- 3.1 per cent[14] of congenital heart diseases. Approximately 50 per cent of these patients die within 2 weeks of life and about 85 per cent die by the age of 6 months[2]. Initially, the patients become cyanotic and hypoxemic, resulting from closure of the patent arterial duct[1-3]. The use of prostaglandins in the late 1970's allowed infants to be in an improved state for the procedures and overall survival increased[3]. In patients with a tripartite right ventricle and confluent pulmonary arteries, the surgical option is open pulmonary valvulotomy with or without a systemic-to-pulmonary shunt[2,15,16] with the mortality approximately 20 per cent[2]. If the blood flow through the right ventricle could be established, growth of the ventricular cavity might occur overtime[5,17,18]. It has been reported that percutaneous valvulotomy of an imperforated pulmonary valve using the laser or radiofrequency guidewire or catheter can be considered as an alternative therapy[7-10]. The good candidates for such procedures include ones who have a tripartite right ventricle and confluent pulmonary arteries, and a well developed infundibulum to obtain a satisfactory catheter position within the right ventricular outflow tract and to avoid perforation by the wire[7,10]. Although it was claimed that manipulation of a tip of radiofrequency catheter is easier than the guidewire because of controllable deflection,[7] we felt that positioning a right Judkins catheter can be performed without difficulty.

Patients whose initial procedure is a pulmonary valvulotomy or placement of a transannular patch without a concomitant systemic-to-pulmonary shunt, about one-half require a systemic-to-pulmonary artery shunt within a few weeks[2]. In our patient, she did not require prostaglandin E_1 or a shunt after the procedure, with improvement

of her oxygen saturation and cyanosis. However, long-term follow-up should be monitored because it has been cited that subsequent placement of a transannular patch is often required in patients whose initial procedure is pulmonary valvulotomy[2].

SUMMARY

We report a successful transcatheter laser guidewire perforation of an atretic membrane pulmonary valve with a subsequent balloon dilation in a newborn with pulmonary atresia and intact ventricular septum. This procedure can be considered as primary therapy in these patients. However, long-term follow-up is required.

ACKNOWLEDGEMENT

The authors wish to thank Mr. Burin Kwaosa-ard for his assistance in photographic preparation.

(Received for publication on May 7, 1998)

REFERENCES

1. Freedom RM. Pulmonary atresia and intact ventricular septum. In Emmanouilides GC, Riemenschneider TA, Allen HD, et al. (eds): Moss and Adams heart disease in infants, children, and adolescents including the fetus and young adult, 5th ed. Philadelphia, Williams & Wilkins 1995: 962-83.
2. Kirklin JW, Barratt-Boyes BG. Cardiac Surgery, Vol 2. New York, Churchill Livingstone 1993: 1035-54.
3. Riopel DA. Pulmonary valve atresia with intact ventricular septum. In: Garson A Jr, Bricker JT, McNamara DG (eds). The science and practice of pediatric cardiology, Vol II. Philadelphia, Lea & Febiger 1990: 1108-17.
4. de Leval MR, Bull C, Stark J. Pulmonary atresia and intact ventricular septum: surgical management based on a revised classification. Circulation 1982; 66: 272-80.
5. Dobell ARC, Grignon A. Early and late results in pulmonary atresia. Ann Thorac Surg 1977; 24: 264-74.
6. Cobanoglu A, Metzdorff MT, Pinson CW, et al. Valvulotomy for pulmonary atresia with intact ventricular septum. J Thorac Cardiovasc Surg 1985; 89: 482-90.
7. Qureshi SA, Rosenthal E, Tynan M, et al. Transcatheter laser assisted pulmonic valve dilation in pulmonary valve atresia. Am J Cardiol 1991; 67: 428-31.
8. Parsons JM, Rees MR, Gibbs JL. Percutaneous laser valvulotomy with balloon dilatation of the pulmonary valve as primary treatment for pulmonary atresia. Br Heart J 1991; 66: 36-8.
9. Rosenthal E, Qureshi SA, Chan KC, et al. Radiofrequency-assisted balloon dilation in patients with pulmonary valve atresia and intact ventricular septum. Br Heart J 1993; 69: 347-51.
10. Akagi T, Hashino K, Maeno Y, et al. Balloon Dilatation of the pulmonary valve in a patient with pulmonary atresia and intact ventricular septum using a commercially available radiofrequency catheter. Ped Cardiol 1997; 18: 61-3.
11. Latson L, Cheatham J, Froemmings S, et al. Transductal guidewire "rail" for balloon valvuloplasty in neonates with isolated critical pulmonary valve stenosis or atresia. Am J Cardiol 1994; 73: 713-4.
12. Forencz C, Rubin JD, McCarter RJ, et al. Congenital heart disease: prevalence at livebirth-the Baltimore-Washington infant study. Am J Epidemiol 1985; 1: 31-6.
13. Freedom RM, Perrin D. The right ventricle: morphological considerations. In Freedom RM (ed). Pulmonary atresia with intact ventricular septum. New York, Futura 1989: 53-75.
14. Fyler DC, Buckley LP, Hellenbrand WE, et al. Report of the New England regional infant cardiac program. Pediatrics 1980; 65(suppl): 376-461.
15. Hanley FL, Sade RM, Blackstone EH, et al. Outcomes in neonatal pulmonary atresia with intact ventricular septum. J Thorac Cardiovasc Surg 1993; 105: 406-27.
16. Anderson RH, McCartney FJ, Shinebourne EA, et al. Paediatric Cardiology. New York, Churchill Livingstone 1987; 711-20.
17. Trusler GA, Yamamoto N, Williams WG, et al. Surgical treatment of pulmonary atresia with intact ventricular septum. Br Heart J 1976; 38: 957-60.
18. Graham TP, Bender HW, Atwood GF, et al. Increase in right ventricular volume following valvulotomy for pulmonary atresia or stenosis with intact ventricular septum. Circulation 1974; 50(Suppl II): 69-79.

การใช้เลเซอร์ช่วยในการขยายลิ้นหัวใจพุลโมนารีย์ ในการรักษาผู้ป่วยทารกที่เป็นลิ้นหัวใจพุลโมนารีย์ตันและไม่มีรูรั่วที่ผนังกั้นหัวใจห้องล่าง

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ได้รายงานประสบการณ์แรก และเป็นรายแรกของประเทศไทย ในการรักษาทางสายสวนโดยใช้เลเซอร์ผ่านลวดเล็กๆ ในการเจาะลิ้นหัวใจพุลโมนารีย์ที่ตัน ตามด้วยการขยายลิ้นหัวใจด้วยสายสวนที่มีลูกโป่งตรงส่วนปลาย ได้เป็นผลสำเร็จในผู้ป่วยทารกอายุ 14 วัน ที่มีลิ้นหัวใจพุลโมนารีย์ตัน และไม่มีรูรั่วที่ผนังกั้นหัวใจห้องล่าง หลังการรักษา ได้ฉีดสารทึบรังสีในเวนตริเคิลขวา พบว่าสารทึบรังสีพุ่งผ่านลิ้นหัวใจพุลโมนารีย์ ไม่พบภาวะแทรกซ้อนที่สำคัญ จากการตรวจคลื่นเสียงสะท้อนความถี่สูง 1 สัปดาห์ หลังการทำพบว่า ความดันต่างที่ระดับลิ้นหัวใจพุลโมนารีย์เท่ากับ 30 มม.ปรอท และความดันเลือดในเวนตริเคิลขวาเท่ากับ 60 มม.ปรอท จากการวัดความอิ่มตัวของออกซิเจนในเลือดที่ปลายนิ้ว พบว่ามีค่าสูงขึ้นจากกว่าร้อยละ 70 หลังการรักษา เป็นร้อยละ 92 - 97 เมื่อ 1 เดือนหลังการรักษา

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