□ Case Report □

Intervention with Balloon Valvuloplasty followed by Patent Ductus Arteriosus Stent in a Patient with Pulmonary Atresia with Intact Ventricular Septum

Han Hyuk Lim, M.D., Young Deuk Kim, M.D., Jae Hwan Lee, M.D.*
Mea Young Chang, M.D. and Hong Ryang Kil, M.D.

Department of Pediatrics and Internal Medicine, College of Medicine, Chungnam National University, Daejeon, Korea*

Pulmonary atresia with intact ventricular septum (PAIVS) is rare, less than 1% of congenital heart disease. It needs a therapeutic approach according to its individual morphologic feature. Surgical treatment of valvotomy and modified Blalock-Taussig shunt or non-surgical interventional catheter balloon valvuloplasty can be used for mild to moderate hypoplasia of right ventricle. Fontan operation can be considered for less optimum morphological substrate of two ventricular repair. A 3-day-old male neonate was admitted with cyanosis and cardiac murmur. On echocardiogram, he had membranous pulmonary atresia with intact ventricular septum, normal sized tripartite right ventricle, large atrial septal defect with right-to-left shunt, small sized patent ductus arteriosus, and moderate tricuspid regurgitation. He was treated with intravenous continuous infusion of prostaglandin E_1 (PGE_1) at once. On the third day of hospitalization, Balloon valvuloplasty was performed. After insertion of patent ductus arteriosus stent on the tenth day, PGE_1 infusion was discontinued. On the fifteenth day, he was discharged. Now, he is 9 months old and has nearly normal cardiac structure and function with 97% of percutaneous oxygen saturation. **(Korean J Pediatr 2005;48:1256-1259)**

Key Words : Pulmonary Atresia, Balloon Valvuloplasty, Stent

Introduction

Pulmonary atresia with intact ventricular septum (PAIVS) needs a variable therapeutic approach according to its morphologic feature[1-3]. In mild-to-moderate hypoplasia of right ventricle, the outcome of variable therapeutic management with interventional catheterization are comparable to surgical management. If cyanosis is persistent even after transcatheter valvotomy, Blalock-Taussig shunt should be considered. Interventional stenting of patent ductus arteriosus can be an alternative to shunt procedure.

The authors report a case of PAIVS treated with interventional catheterization. The transcatheter balloon valvotomy was performed on a newborn with PAIVS and normal sized right ventricle, and then stenting the patent ductus arteriosus was performed using interventional catherization because of persistent cyanosis. He became hemodynamically nearly normal in cardiac structure and function.

접수 : 2005년 6월 27일, 승인 : 2005년 8월 9일
책임저자 : 길홍량, 충남대학교 의과대학 소아과학교실
Correspondence : Hong Ryang Kil, M.D.
Tel : 042)220-7274 Fax : 042)255-3158
E-mail : gilhong@cnu.ac.kr

Case Report

Patient : Lim○○ baby, male, 3 days old

Chief complain : Generalized cyanosis and cardiac murmur

Present illness : He was delivered uneventfully. There was no problem at birth. Cardiac murmur and generalized cyanosis were noted at third day after birth. He was transferred to our neonatal intensive care unit for further evaluation and management. Antenatal care was regularly performed without any problem, and fetal cardiac anomaly was not noted.

Birth history : Gestation age was 37^{+6} weeks, and birth weight was 3,680 g. He was born with normal vaginal spontaneous delivery as first baby. The APGAR score was 8 on 1 minute and 9 on 5 minute.

Maternal & Familial history : Mother was 28 years old and carrier of hepatitis B virus. She had no history of medication, radiation, or infection prenatally without history of abortion. The family history of hereditary or cariovascular disorders was not noted.

Physical examination : On admission, body temperature was 37.7℃, heart rate 132 beats per minute, respiratory rate 60 breaths per minute, and blood pressure 67/43 mmHg without disparity between upper and lower extremities. The body weight was 3,700 g (75-90 percentile) and decreased activity and mild cyanosis were observed. Anterior fontanel was open without bulging. Mild chest wall retraction was visible in intercostal and subcostal areas but the breathing sound was clear. The heart sound was regular and single and regurgitant murmur was audible on left lower sternal border. The continuous murmur was heard with Grade of 3/6 along left upper sternal border. Abdominal wall was soft and liver was palpable with 2 finger-breath. Moro reflex was normal, but the grasp and sucking reflex were decreased.

Laboratory findings : On admission, laboratory findings were as follows : 9,700/mm^3 of white blood cell, 19.0 g/dL of hemoglobin, 53.9% of hematocrit, 282,000/mm^3 of platelet, 20 IU/L of AST, 9 IU/L of ALT, 5.0 g/dL of total protein, 3.1 g/dL of albumin, 6.2/0.51 mg/dL of BUN/creatine, 47 mg/dL of glucose and $Na^+/K^+/Cl^-$ 142.1/3.8/110.7 mEq/L. On gas analysis of arterial blood, pH was 7.532, pCO_2 30.1 mmHg, pO_2 36.9 mmHg, base excess 4.4 mmol/L, SaO_2 80.5%, and HCO_3^- 25.4 mmol/L.

Radiologic findings : On chest X-ray, there was mild cardiomegaly without increase of pulmonary vascular markings. Sonographic findings of brain and kidney were not specific. The echocardiographic findings were as follows : membranous pulmonary atresia with intact ventricular septum, normal sized tripartite right ventricle (-0.3 of Z value of tricuspid valve[4] (Fig. 1), a large atrial septal defect with right-to-left shunt (Fig. 2), small sized patent ductus arteriosus (Fig. 3), and moderate regurgitation of tricuspid valve (ΔP=116 mmHg).

Treatment and Hospital course : On the first day of hospitalization, prostaglandin E_1 (Eglandgin®, Dong-A pharmaceutical Co, Republic of Korea) was intravenously infused in 10 ng/kg/min to maintain patency of ductus arteriosus. On the 5th day of hospitalization, the percutaneous transcatheter pulmonary valvotomy was performed with 0.14 Shinomi-stiff wire and gradual increased size of balloons (2.5 mm×20 mm, 4.0 mm×20 mm, 8.0 mm×20 mm, peripheral balloon, Boston Scientific, Ireland) (Fig. 4). The pressure gradient between right ventricle and pulmonary artery was decreased from 100 to 20 mmHg, and the pressure ratio of right and left ventricle from 1.83 to 0.92 after

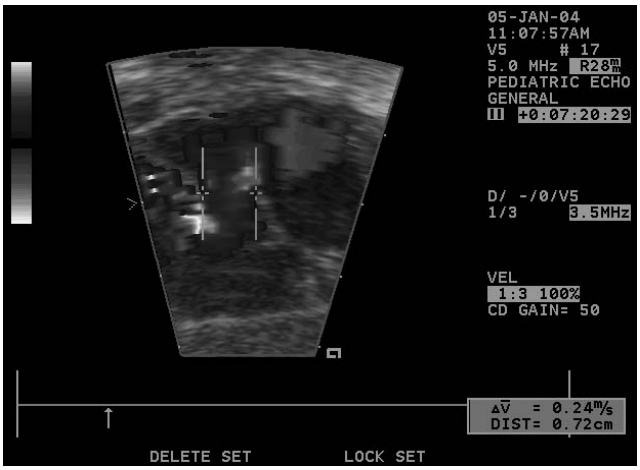

Fig. 2. On the subcostral four chamber view, the atrial septal defect is shown with right to left shunt without restriction.

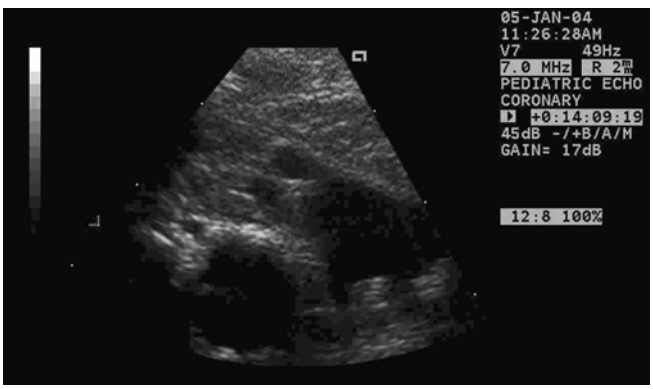

Fig. 1. On the parasternal short axis view, no pulmonary valve opening is shown.

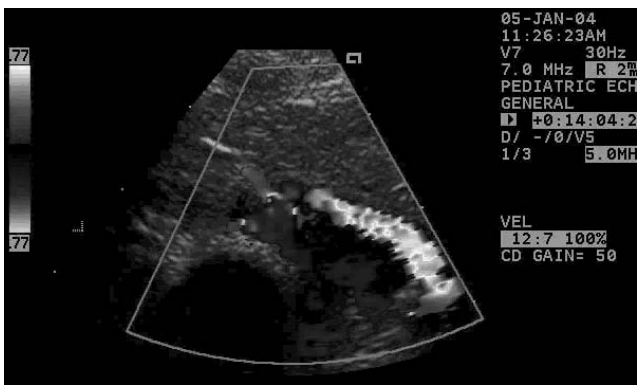

Fig. 3. On the parasternal short axis view, blood flow through opened patent ductus arteriosus is shown.

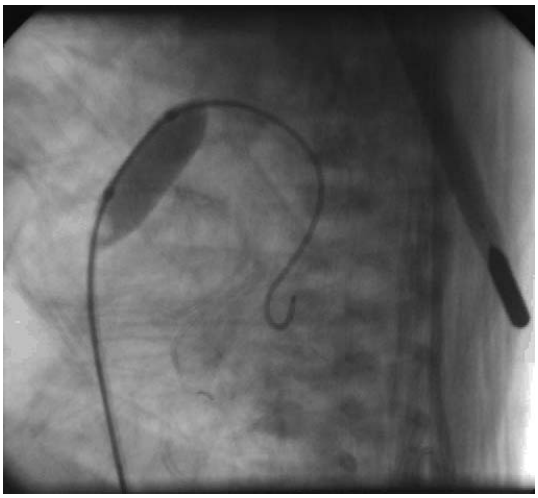

Fig. 4. On the catherization-angiogram, balloon catheter is located through center of pulmonary valve, and balloon valvotomy is performed.

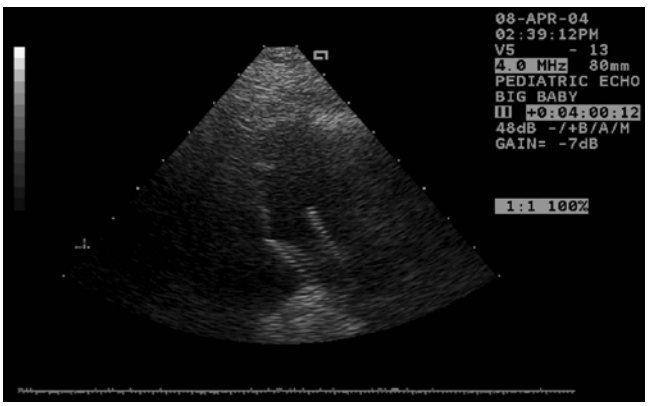

Fig. 5. On the echocardiogram, the stent is implanted at the site of patent ductus arteriosus. But 3 months later, the stent was clogged.

transcatheter valvotomy. The PGE_1 infusion could not be stopped because of persistent hypoxemia (70-80% of percutaneous oxygen saturation). On the 10th day, a stent (3.0 mm×8 mm, Express®, Boston Scientific Scimed, Inc., Maple Grove, Minnesota) was implanted using cardiac catheterization for patency of ductus arteriosus (Fig. 5). Thereafter PGE_1 infusion was discontinued. On the 15th day, he was discharged in good general condition. Two months later, the shunt flow of ductus arteriosus was not visible in spite of low dose aspirin after 2 months of stent implantation.

Now, he is 9 months old with 10.6 kg of body weight (50-75 percentile). The percutaneous oxygen saturation is 97%. On echocardiogram, there are small atrial septal defect with left-to-right shunt, mild tricuspid regurgitation (Vmax=2.7 m/sec), mild pulmonary valve stenosis (Vmax=2 m/sec), mild pulmonary regurgitation, normal ventricular function, and no cardiomegaly. He is hemodynamically as well as morphologically, nearly normal.

Discussion

PAIVS is an uncommon disorder, fewer than 1% of all congenital heart defects. Abnormalities of development of right ventricle, atresia of pulmonary valve, and shunt between right ventricle and coronary artery are morphologic features. Kutsche and Van Mierop et al. suggested that PAIVS occurs later in cardiac morphogenesis[5].

The tricuspid valve is rarely normal in patient with PAIVS. The most severely stenotic and obstructive tricuspid valve is observed in patients with the most underdeveloped right ventricle. Conversely, patients with the largest right ventricle usually have very severe tricuspid regurgitation, with a valve exhibiting features of Ebstein's and dysplasia[6]. The patients become cyanotic and hypoxemic coincidentally with functional and anatomic closure of the patent arterial duct[7, 8]. In our case, a large secundum atrial septal defect with right-to-left shunt and moderate regurgitation of tricuspid valve with normal sized tripartite right ventricle was observed. PGE_1 was started immediately at arrival in neonatal intensive unit.

Cardiac catheterization and angiography are recommended to evaluate hypoplastic severity and pressure of right ventricle and connection between ventricle and coronary arteries. It is treated with variable surgical or non-surgical interventional managements, according to the right ventricular morphology[9]. In mild to moderate hypoplasia of right ventricle, placement of a transannular right ventricle outflow patch and a systemic-to-pulmonary artery shunt seems to be a most promising for two ventricular repair, with 2-3 weeks of PGE_1 infusion to maintain ductal patency. The 1-year survival rate is 80%. With a closed transpulmonary valvotomy without cardiopulmonary bypass and a modofied Blalock-Taussig shunt, the motality rate of these procedure is below 5%.

Since 1990, transcatheter pulmonary valvotomy has become a popular approach. Ovaert et al. reported that transcatheter valvotomy showed equal or better outcome and safety than surgical valvotomy[10]. And Park et al. reported that a baby with PAIVS, weighting 2.2 kg was treated by transcatheter valvotomy with using 2-French catheter[11].

If cyanosis is observed continuously after transcatheter

valvotomy, the Blalock-Taussig shunt may be performed instead of PGE_1 infusion to maintain the patency of ductus arteriosus. However, in neonate and small infant a surgically created Blalock-Taussig shunt is now not without significant morbidity. Pleural effusion, diaphragmatic paralysis, cardiac failure due to excessive pulmonary blood flow, and distortion of branch pulmonary arteries are known complications that may compromise the feasibility or outcome of repair[12]. Stenting of PDA was thought to be a novel approach as an alternative, but results of earlier reports have been discouraging. However, with improvement in coronary stent design and delivery system, it is perhaps reasonable to reexam the role of PDA stenting as an alternative to Blalock-Taussig shunt[13]. Alwi et al. reported that stent implantation of patent ductus arteriosus in ductal dependent pulmonary circulation is more efficacious and safer[14]. Siblini et al.[15] and Bokenkamp et al.[16] reported that an infant with PAIVS was treated successfully by transcatheter valvotomy and stent implantation. In this case, Intervention with balloon valvuloplasty with gradual increased size of balloon was preformed and then PDA stent was implanted using cardiac catheterization due to persistent cyanosis instead of Blalock-Taussig shunt. He became stable hemodynamically as well as morphologically even with closure of PDA, 2 month later.

한 글 요 약

풍선판막성형술과 동맥관 스텐트를 이용하여 치료한 심실중격결손을 동반하지 않은 폐동맥 폐쇄 1례

충남대학교 의과대학 소아과학교실, 내과학교실*

임한혁 · 김영득 · 장미영 · 이재환* · 길홍량

심실중격결손을 동반하지 않은 폐동맥 폐쇄(pulmonary atresia with intact ventricular septum)는 전체 선천적 심장병의 1%를 차지하는 드문 질환으로 형태학적 특성에 따라 다양한 치료적 접근이 필요하다. 이에 저자들은 정상 크기발달의 우심실을 가진 심실중격결손을 동반하지 않은 폐동맥 폐쇄가 있는 신생아에서 풍선판막성형술 후 지속적인 저산소증을 보여 동맥관 스텐트를 이용하여 거의 정상적인 심장구조와 기능으로 호전된 1례를 경험하였기에 보고하는 바이다.

References

1) Fyler DC. Report of the New England regional infant cardiac program. Pediatrics 1985;65(suppl):376-461.
2) Ferencz C, Rubin JD, McCarter RJ. Congenital heart disease : prevalence at live birth-the Baltmore-Washington study. Am J Epidemiol 1985;121:31-6.
3) Freedom RM, Perrin D. The right ventricle : morphological considerations. In : Freedom RM, ed. Pulmonary atresia with intact ventricular septum. Mount Kisco, New York : Futura, 1985;53-75
4) Hanley FL, Sade RM, Blackstone EH, Kirklin JW, Freedom RM, Nanda NC. Outcomes in neonatal pulmonary atresia with intact ventricular septum. A multiinstitutional study. J Thorac Cardiovasc Surg 1993;105:406-27.
5) Kutsche LM, Van Mierop LHS. Pulmonary atresia with and without ventricular septal defect : a different etiology and pathogenesis for the atresia in the 2 types. Am J Cardiol 1983;51:932-5.
6) Anderson RH, Silverman NH, Zuberbuhler JR. Congenitally unguarded tricuspid orifice : its differentiation from Ebstein's malformation in association with pulmonary atresia and intact ventricular septum. Pediatr Cardiol 1990;11:86-90.
7) Freedom RM, Burrows PE, Smallhorn JF. Pulmonary atresia with intact ventricular septum. In : Freedom RM, Benson LN, Smallhorn JF, eds. Neonatal heart disease. New York : Springer-Verlag, 1992;285-307.
8) Rowe RD, Freedom RM, Mehrizi A. The neonate with congenital heart disease. New York : WB Saunders 1981;328-49.
9) Yoshimura N, Yamaguchi M, Ohashi H, Oshima Y, Oka S, Yoshida M, et al. Pulmonary atresia with intact ventricular septum : strategy based on right ventricular morphology. J Thorac Cardiovasc Surg 2003;126:1417-26.
10) Ovaert C, Qureshi SA, Rosenthal E, Baker EJ, Tynan M. Growth of the right ventricle after successful transcatheter pulmonary valvotomy in neonate and infants with pulmonary atresia and intact ventricular septum. J Thorac Cardiovasc Surg 1998;115:1055-62.
11) Park IS, Nakanishi T, Nakazawa M, Takanashi Y, Imal Y, Momma K. Radiofrequency pulmonary valvotomy using a new 2-French catheter. Cathet Cardiovasc Diagn 1988;45:37-42.
12) Tamisier D, Vouhe PR, Vermant F, Leca F, Massooat C, Neveux J. Modified Blalock Tassug shunts : results in infants less than 3 months of age. Ann Thorac Surg 1990;40:797-802.
13) Gibbs JL, Uzun O, Blackburn MEC, Wren C, Hamilton L. Watterson KG. Fate of the stented arterial duct. Circulation 1999;99:2621-5.
14) Alwi M, Choo KK, Latiff HA, Kandavello G, Samion H, Mulyadi MD. Initial results and medium-term follow-up of stent implantation of patent ductus arteriosus in duct-dependent pulmonary circulation. J Am Coll Cardiol 2004;44:438-45.
15) Siblini G, Rao PS, Singh GK, Tinker K, Balfour IC. Transcatheter management of neonates with pulmonary atresia and intact ventricular septum. Cathet Cardiovasc Diagn 1997;42:395-402.
16) Bokenkamp R, Kaulitz R, Paul T, Hausdorf G. Stepwise interventional approach in a neonate with pulmonary atresia and intact ventricular septum. Eur J Pediatr 1988;157:885-9.