

Initial Palliation of the Pulmonary Atresia with Interventricular Communication

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<국문초록>

심실간 중격결손을 동반한 폐동맥폐쇄증의 일차고식적 수술

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본 보고는 울산의대 흉부외과 교실에서 최근 2년간 치험한 15례의 심실중격 결손동반 폐동맥폐쇄에 대한 일차고식수술의 단기 성적을 분석하였다. 15례는 단순형태의 폐동맥폐쇄증이 8례 그리고 완전방실결손증, 총폐정맥환류이상증 그외 다른 복잡심기형과 동반된 폐동맥 폐쇄가 7례였다.

수술시 환자의 연령분포는 1례를 제외한 전례가 1세 미만이었으며, 5례는 1개월 미만의 영아였다. 폐혈관의 형태는 매우 다양하였으며, 대부분의 예에서 폐동맥 협착 또는 폐동맥 발육부전의 소견이 있었다. 동맥관은 폐동맥 부착부위의 협착이 통상적 소견이었다.

고식적 수술로서 Modified Blalock-Taussing shunt가 6례, Central shunt가 5례이며 이중 4례는 체외순환하에 폐동맥확장 성형술을 동시에 시행하였다. 우심실유출로 조형술(Right ventricular outflow reconstruction)이 3례, 그리고 1례는 양측 폐동맥을 직접 연결한후 Bilateral cavopulmonary shunt(BCS)를 시행하였다.

수술사망은 3례로서, 우심실유출로조형술 2례와 BCS 1례가 사망하였다. 모두 좌폐동맥의 심한 형성부전의 소견이 있으며, 그중 2례는 총폐동맥환류이상증의 동시 수술례이었다. Modified Blalock-Taussing shunt나 Central shunt 수술례에서는 수술사망례가 없었다. 그러나 Modified Blalock-Taussing Shunt 6례중 3례는 여러가지 원인으로인해 조기에 2차 고식적 수술이 필요하였다. 추시기간중 2례는 일차 수술후 각각 13, 18개월후 완전교정술을 받고 좋은 결과를 보였다.

영유아기 심실간 중격동반 폐동맥폐쇄증 환자의 고식적 수술에서 그 수술법의 선택 그 수술이 갖는 문제점들을 본 증례와 문헌고찰을 통하여 검토하였다.

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— Abstract —

The ideal approach in the staged management of patients with pulmonary atresia has been a challenging problem and the result has not been always satisfactory.

We reviewed our early result of initial palliative surgeries in fifteen cases of pulmonary atresia with interventricular communication. Included are eight cases of simple pulmonary atresia with ventricular septal defect and seven cases of pulmonary atresia associated with other complex cardiac anomalies.

The ages of the patients were less than one year except one. The morphology of pulmonary vasculature was highly variable and showed unfavorable conditions in most cases. Pulmonary artery was nonconfluent in two. Two-thirds of all cases showed significant problems such as juxtaductal stenosis or diffuse hypoplasia. The ductus arteriosus usually narrowed at its pulmonic end.

Initial palliation was done by modified Blalock-Taussig shunt in six, central shunt with or without pulmonary angioplasty in five, right ventricular outflow tract (RVOT) reconstruction in three and direct connection of nonconfluent pulmonary arteries with bilateral cavopulmonary shunt in one patient.

There were 3 hospital deaths. Two of them underwent simultaneous repair of the associated anomaly of TAPVR. Among the six patients who received modified Blalock-Taussig shunt, three needed early second palliative procedure by central shunt, RVOT patch reconstruction and pulmonary angioplasty in each case. All patients who received central shunt showed marked clinical improvement.

Among the twelve patients who survived the palliative procedures, two patients underwent total correction 13 months and 18 months after initial palliation respectively.

We think that the choice of palliative procedure must be individualized according to the morphology of the pulmonary arteries. More experience and long term follow-up data are necessary to meet this challenging problem.

The condition of pulmonary atresia may present as a form of severe Fallot's tetralogy or as an associated feature in complex cardiac malformations. Its presence usually preclude the early correction and palliative procedure to relieve hypoxia and to promote pulmonary arterial growth is necessary as an initial management. However, the best approach to the staged management of these patients remains controversial.

The results of various surgical procedures, including any form of systemic to pulmonary artery anastomosis or right ventricular outflow reconstruction have not been always satisfactory. It is mainly due to the unfavorable anatomy of the pulmonary vasculature and the source of pulmon-

ary blood flow. Without doubt, technical factors during these operations should be considered as well.

Our experience also, though limited in the number of patients, has not been uniformly satisfactory. We have reviewed our fifteen patients having the anatomy of pulmonary atresia with interventricular communication to define the variability of this anomaly and to evaluate the result of the initial palliation on these patients.

Patients

All patients who had received initial palliative operation for pulmonary atresia with int-

er-ventricular communication at Asan Medical Center between September 1989 and October 1991 were the subject of this review.

Included are 8 cases of pulmonary atresia with ventricular septal defect, featuring the morphology of Fallot's tetralogy or double outlet right ventricle(DORV), and 7 cases of pulmonary atresia associated with other complex cardiac anomalies. The details of the major associated anatomy in the latter group are followings:complete atrioventricular septal defect(AVSD) in two, total anomalous pulmonary venous return(TAPVR) of supracardiac type in one, right isomerism having common atrioventricular valve(CAVV) with infracardiac type TAPVR in one, right isomerism with mitral atresia in one, corrected transposition of the great arteries(C-TGA) in one, and tricuspid atresia in one patient(Table 1).

The age of the patients at initial operation was less than one year except one. (ranged from 7 days to 4 years, median 4.2 months) (Table 2). Body weight ranged from 2.4kg and 15.4kg(median 5.4kg). It was less than 3kg in four neonates. All patients presented with cyanosis and/or respiratory difficulty. Five patients required mechanical ventilation preoperatively.

Preoperative hemoglobin values were between

| Table 1. Major cardiac diagnosis | N=15 |
|---|----------|
| Simple Pulmonary Atresia with · VSD | 8 |
| Complex Pulmonary Atresia with · Complete AVSD | 7 (2) |
| · VSD, TAPVR | (1) |
| · Rt isomerism, CAVV, TAPVR | (1) |
| · VSD, Corrected-TGA | (1) |
| · Rt isomerism, Situs inversus, Mitral atresia | (1) |
| · Tricuspid atresia | (1) |

VSD : Ventricular septal defect.

AVSD : Atrioventricular septal Defect.

TAPVR : Total anomalous pulmonary venous return.

CAVV : Common atrioventricular valve.

TGA : Transposition of the great arteries.

Table 2. Age distribution at the initial palliation

| | | N=15 | | | | |
|-------------|-----|------|-----|------|-----|--|
| Age(month) | 0-1 | 1-3 | 3-6 | 6-12 | ≥12 | |
| No. of case | 5 | 2 | 6 | 1 | 1 | |

12.5 and 22.8mg /dl(mean 17.8mg /dl). Arterial O₂ saturations showed wide fluctuation between 32% and 88% depending on the condition of the patients and PGE₁ infusion which was given preoperatively in 5 patients.

Anatomy of the pulmonary arteries

The basic anatomy of the intra and extracardiac anomalies were defined using two-dimensional and color Doppler echocardiography. Preoperative diagnosis was focused mainly on the morphology of the pulmonary arterial tree and the source of the pulmonary blood flow. All patients underwent various angiographic studies including aortography, selective injection into the ductus arteriosus, pulmonary vein wedge angiography or countercurrent aortography through radial or brachial artery.

The morphology of the pulmonary arteries was highly variable and showed unfavorable conditions in most cases.

In two cases, the intrapericardial pulmonary artery was absent and each right and left pulmonary artery was supplied by bilateral ducti. Thirteen cases had confluent pulmonary arteries but many variations were noticed at the level of atresia, in the branching pattern and in the size of each pulmonary artery. Main pulmonary artery was atretic in five cases. Others had the main pulmonary artery of varying size connected to the right ventricular outflow tract (Table 3). Most cases including two patients with bilateral ducti showed significant pulmonary arterial problems. Two-thirds of the cases showed ipsilateral or bilateral focal stenosis of the pulmonary arteries at the juxtaductal region. It was more common in left pul-

Table 3. Anatomy of the pulmonary arteries N=15

| | |
|---------------------|-----|
| · Nonconfluent PA's | 2 |
| · Confluent PA's | 13 |
| with MPA atresia | (5) |
| without MPA atresia | (8) |

PA : Pulmonary artery

MPA : Main pulmonary artery

monary artery. One of both pulmonary arteries were diffusely hypoplastic (less than 3mm in diameter) in five cases (Fig.1,2,3).

The pulmonary flow was all ductus dependent and the ductus narrowed significantly at the pulmonary end in more than half of all cases. Major aortopulmonary collateral arteries (MAPCA's) were not seen in our cases except one, in which a medium sized MAPCA from the descending aorta was supplying some segments of the left lower lobe.

Operation

All patients received initial palliative procedure (Table 4).

Various methods were applied according to the morphology of the pulmonary arteries and intracardiac anatomy.

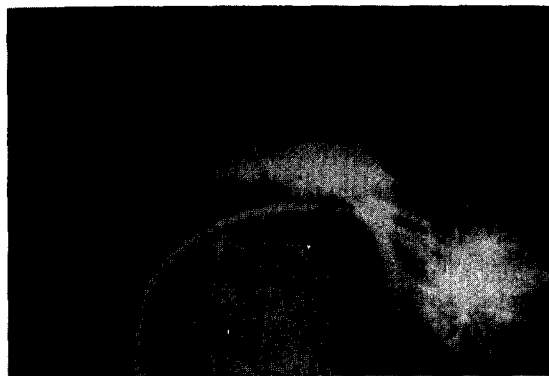


Fig. 1. Left pulmonary wedge angiography shows juxtaductal stenosis of left pulmonary artery and mild hypoplasia of right pulmonary artery.

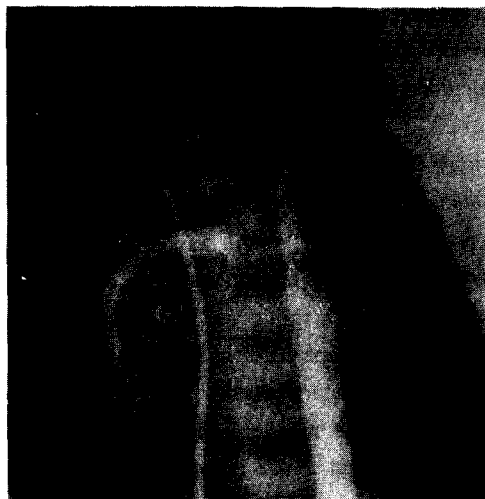


Fig. 2. Ascending aortography shows severe proximal stenosis of the diminutive left pulmonary artery.

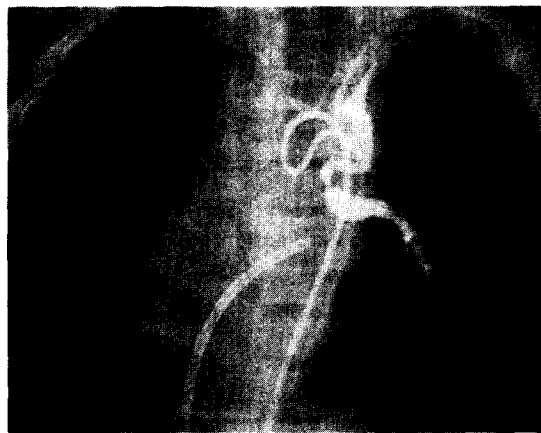


Fig. 3. Selective injection to ductus shows nonconfluent pulmonary arteries and marked hypoplasia of left pulmonary artery with narrowing of the ductus at its pulmonary end.

Six patients received modified Blalock-Taussig shunt. In remaining patients except one who had a central shunt, operations were performed under the extracorporeal circulation. The anatomic finding of pulmonary arteries and the lateralization of the ductal flow was unfavorable or considered somewhat unsafe to perform the modified

Table 4. Method of initial palliation

N=15

| Method | No | Hospital Mortality |
|-------------------------------------|-----------|--------------------|
| Modified Blalock-Taussig shunt | 6 | 0 |
| Central shunt | 1 | 0 |
| Central shunt with angioplasty | 4 | 0 |
| · Pericardial patch | (2) | |
| · Tube graft | (1) | |
| · Native artery | (1) | |
| ROVT patch reconstruction | 2 | 1* |
| RV-PA conduit & central shunt | 1 | 1 |
| SVC-to-reconstructed PA anastomosis | 1 | 1* |
| Total | 15 | 3 |

RVOT : Right ventricular outflow tract SVC : Superior vena cava

RV : Right ventricle

*Simultaneous repair of TAPVR

PA : Pulmonary artery

Blalock-Taussig shunt, so pulmonary angioplasty with central shunt or RVOT reconstruction were performed under the extracorporeal circulation in beating or fibrillating condition without aorta cross clamping. Intracardiac anomalies were not repaired during the initial operation with the exception of TAPVR which was simultaneously repaired in two patients.

Four patients had a central shunt with pulmonary angioplasty. In two of them, the stenotic pulmonary artery was widened with bovine pericardium. In another patient with severe stenosis at the proximal portion of both pulmonary arteries, a 7mm polytetrafluoroethylene (PTFE) vascular graft was connected between the two pulmonary arteries and a central shunt was made with 5mm PTFE graft. In another patient, nonconfluent pulmonary arteries were connected directly and a central shunt from innominate artery was made onto the reconstructed pulmonary artery.

In two patients, RVOT was reconstructed with autologous pericardium. The patch was extended over the stenotic area of the left pulmonary artery. In one of them, associated TAPVR of supracardiac type was repaired.

One-month old infant with marked hypoplasia of the left pulmonary artery had a pericardial conduit

connected between the right ventricle and the pulmonary artery. However due to persistent hypoxia after weaning from the cardiopulmonary bypass, a central shunt was added onto the conduit.

In one patient with right isomerism, common atrioventricular valve and TAPVR of intracardiac type, the hypoplastic left pulmonary artery was connected directly to the nonconfluent right pulmonary artery, and then right superior vena cava was anastomosed end-to-side to the right pulmonary artery. TAPVR of intracardiac type was also repaired.

Result

Three patients died within 24 hours after operation. One was eight day-old neonate who underwent RVOT patch reconstruction and TAPVR repair. Another patients died after pericardial conduit construction with an additional central shunt. Third patient, five-month old infant died after right superior vena cava-to-reconstructed pulmonary artery anastomosis and TAPVR repair. They all showed persistent hypoxia, metabolic acidosis and low cardiac output postoperatively. On angiography, they all had markedly hypoplastic left pulmonary artery. Survived twelve patients sho-

wed satisfactory palliation. One patient needed exploration for bleeding control. A 7-day-old neonate had the left diaphragm plicated 2 weeks after modified Blalock-Taussig shunt because of phrenic nerve paralysis.

Among the six patients who underwent modified Blalock-Taussig shunt, three patients needed early second palliation. One patient had patch widening of right ventricular outflow tract following thrombotic occlusion of the previous shunt. In another patient, who received plication of left diaphragm, follow-up angiography showed severe angulation and kinking of the graft at the subcalvian artery side. Repeat palliations with balloon dilation and central shunt followed by another right modified Blalock-Taussig shunt failed to solve his pulmonary vascular and ventilatory problems. This patient is considered uncorrectable by conventional surgery. Another patient who received modified Blalock-Taussig shunt on the relatively hook sized left pulmonary artery developed complete occlusion of the juxtaductal region of the right pulmonary artery associated with the recent closure of ductus arteriosus. This patient subsequently underwent pulmonary angioplasty without use of extracorporeal circulation 3 weeks after the initial operation (Table 5).

| Table 5. Result | N=15 |
|-------------------------|-------------|
| Hospital Mortality | 3 |
| Postoperative bleeding | 1 |
| Diaphragmatic paralysis | 1 |
| Second palliation | 3* |
| Total correction | 2 |

*1 : Central shunt and right modified B-T shunt

1 : RVOT patch reconstruction

1 : Pulmonary angioplasty

During the study period, total correction was done in two patients. One patient with double outlet right ventricle and pulmonary atresia underwent total correction by 'REV' procedure one year after modified Blalock-Taussig shunt. Another

patient underwent total correction by VSD closure and RVOT reconstruction with monocuspid ventricular outflow patch one and a half year after initial RVOT patch reconstruction.

Discussion

Unlike the usual Fallot's tetralogy or pulmonary atresia with intact ventricular septum, it is quite common to see severe pulmonary arterial problems in pulmonary atresia with interventricular communication^{4,23,25}. Also the source of pulmonary flow may be complex and highly variable. Our cases, though limited in number, showed rather universal finding of the pulmonary arterial problems such as diffuse hypoplasia or focal stenosis of pulmonary arteries. The ductus was commonly stenotic in its pulmonic end with juxtaductal stenosis of the related pulmonary artery.

The variations of the pulmonary arterial anatomy are known to occur at the site of discontinuity between the right ventricle and the pulmonary circulation, in the anatomy of the major pulmonary vasculature and in the pattern of arborization related to the various sources of pulmonary blood flow^{2,4,19}. In the review of 78 patients at the Mayo Clinic, approximately 60% of their cases had such problems. Common findings were isolated stenosis, diffuse hypoplasia, and non-confluence of the pulmonary arteries. Less commonly, portions of pulmonary arteries were absent associated with complex aorto-pulmonary collaterals¹⁰. These findings impose on cardiologists and surgeons many challenging problems in planning ideal management of these patients. These severe pulmonary arterial problems preclude early total correction and palliative procedures to relieve cyanosis and to promote pulmonary arterial growth are necessary.

Conventional systemic-to-pulmonary arterial shunt has been the common practice for the palliation of these patients^{3,8,16,23}. Gale et al²⁰ dem-

onstrated that the Blalock-Taussig shunt can produce effective growth in pulmonary arteries, either ipsilateral or contralateral to the shunt. But the result of systemic to peripheral pulmonary arterial shunt has not been uniformly successful in the patients with pulmonary atresia^{3,16,17,23}. This is because the pulmonary arteries are generally small and focal narrowing or nonconfluence of the pulmonary arteries are more frequent in the group of pulmonary atresia than in pulmonary stenosis^{23,25}. Moreover, it can be extremely difficult to create a perfect shunt in a very small neonate with hypoplastic pulmonary arteries²². Somerville emphasized the perfect surgical technique during shunt operation to avoid iatrogenic deforming of the small pulmonary arteries³. Most problems in the perioperative period and the late operative deaths were related to the complications from earlier shunt procedures³.

Piehler et al¹⁰, and Millikan et al¹¹, noting the frequency of anastomotic stenosis or peripheral distortion associated with the shunt procedures, reported a large experience with patch or conduit reconstruction of the right ventricular outflow tract in patients with Fallot's tetralogy and pulmonary atresia. The advantages of this approach may be the better chance of symmetrical enlargement of both pulmonary arteries, possibility of simultaneous correction of pulmonary arterial problems and the improved catheter access to the central pulmonary arteries for the subsequent diagnostic studies. Puga et al^{1,12} described a technique for establishment of the right ventricle-hypoplastic pulmonary artery continuity with a tube graft without the aid of extracorporeal circulation¹³.

Right ventricular-pulmonary arterial reconstruction has not been without its problems. Thirty-one of the 57 patients studied postoperatively in the Mayo Clinic were judged unacceptable for final repair because of inadequate pulmonary arterial growth and/of restricted peripheral arborization¹¹. Freedom et al¹³ have found the high incidence of

proximal stenosis of the left pulmonary artery and failure to achieve acceptable pulmonary arterial size for ultimate complete repair. The problem of proximal pulmonary arterial stenosis may be inherent or iatrogenic but it seems related in part to the technical factor during the operation. They suggested central shunt or implantation of the main pulmonary artery onto the side of the aorta as an alternative procedure¹³.

Others^{9,21} prefer to construct a central shunt from the aorta to the main pulmonary artery. Our main indication for central shunt was the presence of severe focal stenosis of the pulmonary arteries which needs simultaneous repair. Barbero-Marcial et al¹⁶ presented a series of simultaneous reconstruction of stenotic or nonconfluent pulmonary arteries with a Blalock-Taussig shunt.

It appears that the best approach to the staged management of patients with pulmonary atresia remains in question. A central shunt may provide more symmetrical pulmonary flow and simultaneous angioplasty can be performed easily with this procedure. However, proper palliative surgical method must be tailored to the individual patient according to the morphology of pulmonary arterial tree. Preoperatively, every effort to delineate the precise anatomy of the pulmonary arterial tree should be practised.

The important question why the pulmonary arteries do not always grow sufficiently in some patients after palliative surgeries remains unanswered. Poor surgical technique, especially in small neonates and infants may contribute to the unfavorable result³. Intrinsic factors in the wall of pulmonary arteries are also proposed^{2,4}. Frequent association of the juxtaductal stenosis or pulmonary arteries, as seen in our cases, has significant surgical implication. Momma et al¹⁶ postulated that progressive ductal constriction may cause juxtaductal stenosis of atresia of the pulmonary artery. Elzenga et al⁷ studied the relation between the pulmonary arterial stenosis and the ductal tis-

sue by histologic examination. Extension of ductal tissue to the pulmonary artery and obstructive ridge caused stenosis of the pulmonary artery similar to the mechanism in the coarctation of the aorta. Our review includes one interesting case in which the juxtaductal stenosis of right pulmonary artery progressed to complete occlusion associated with recent closure of the ductus after successful left modified Blalock-Taussig shunt, therefore, resulting no flow into the right pulmonary artery. Division of the ductus arteriosus and angioplastic widening of the pulmonary artery during the palliative procedure might prevent the untoward progression of the stenosis.

Balloon dilatation angioplasty may have a role in the management of the troublesome congenital or iatrogenic branch stenosis of the pulmonary arteries²¹. As reported by Ring et al¹⁴ and Lock et al¹⁵, it is possible that balloon angioplasty may offer an alternate method for achieving more symmetrical distribution of pulmonary arterial flow in patients following palliative procedures.

Review of our cases has shown significant morbidity and mortality. The association with more complex cardiac anomalies and presentation at early infancy in our cases might have contributed to the unfavorable result. More can be learned by continual review of our current practice and more cases accumulation and long term follow-up are necessary for the better understanding and management of this challenging problem.

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