

# Distal Type of Aortopulmonary Septal Defect with Aortic Origin of Right Pulmonary Artery and Interruption of the Aortic Arch

— A Case of Successful Surgical Report —

Yoon Seop Jeong, M.D.\*, Meong Gun Song, M.D.\*

## — Abstract —

A rare case of the association of distal aortopulmonary septal defect, aortic origin of the right pulmonary artery, intact ventricular septum, patent ductus arteriosus and interrupted aortic isthmus in a 40-day-old infant is reported. The infant was suffered from two operations with an interval of nine days. At the first operation a 10mm polytetrafluoroethylene prosthesis was inserted instead of the interrupted aortic isthmus and ductus was ligated via the left posterolateral thoracotomy. But the patient could not be weaned from the respirator because of large amount of left-to-right shunt. So the total correction was subsequently performed after an interval of nine days. At the second operation, tunneling of the right pulmonary artery to the main pulmonary artery through the aortopulmonary septal defect was performed using the Dacron patch via a longitudinal transaortic approach and a separate autologous pericardial patch was applied to the longitudinally incised margins of the anterior wall of the ascending aorta. The second postoperative course was relatively uneventful except some respiratory distress and nutritional problems. Now he is at 6 months of age and thrives well without any symptom. Because the success of the surgical repair of this complex anomalies depends upon the accurate diagnosis and meticulous design of each step of procedure prior to operation these problems are also discussed.

## INTRODUCTION

Aortopulmonary window is a rare anomaly, with an incidence of about 1.5 case per 1,000 cases of congenital heart disease<sup>1)</sup> The defect, first reported in 1830<sup>2)</sup>, results from a failure of the aortopulmonary septum to completely divide the aor-

tic into the aorta and pulmonary artery. The defects have been classified into proximal, distal and combined forms according to the location of the defect<sup>10,11)</sup>.

The defect, especially in distal type, can be either isolated or associated with other cardiovascular abnormalities, which may include patent ductus arteriosus, interrupted aortic arch and aortic origin of the right pulmonary artery. Berry et al<sup>3)</sup> recognized this association of distal type of aortopulmonary window with aortic origin of the right pulmonary artery, intact ventricular septum,

\*Department of Thoracic and Cardiovascular Surgery,  
Asan Medical Center, College of Medicine, Ulsan  
University  
1991년 7월 29일 접수

patent ductus arteriosus and interruption of the aortic isthmus as a specific syndrome on the basis of developmental and hemodynamic hypothesis during the fetal life.

Because the preoperative conditions of these patients are unstable and the surgical mortality is high as yet, accurate diagnosis and anatomic depicting of each component of this combination is very important.

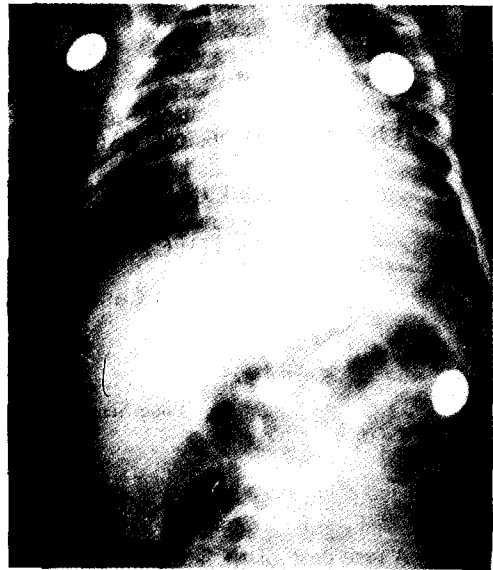
We report our experience with this rare congenital cardiac anomalies in a 40-day-old male infant and provide the review of the literature.

### CASE REPORT

A 40-day-old male infant was known to have a heart murmur since birth and transferred to our hospital for the further evaluation and treatment. He was born after 36 weeks of uneventful gestation. When he was admitted his chief complaints were respiratory distress and cyanosis on crying.

On physical examination the body weight was 3.5kg, the heart rate 156/min, and respiration rate 65/min. The blood pressure of the right and the left arms were 90/48mmHg and 100/41 mmHg, while these of the right and left legs were 64/45mmHg and 69/43mmHg respectively. There was a marked anterior prominence of the chest wall with evidence of biventricular hypertrophy on palpation. On auscultation the first sound at the left sternal border was followed by a soft, grade 3/6 late systolic murmur, which continued through the second sound into early diastole. The pulmonary component of the second sound was moderately accentuated. The chest radiograph showed cardiomegaly (CTR 70%) with a left-sided aortic arch, prominent pulmonary artery and plethoric lung fields (Fig. 1).

The electrocardiogram showed sinus rhythm with evidence of right axis deviation and biventricular hypertrophy (Fig. 2). The echocardiogram and MR imaging revealed the accurate ana-

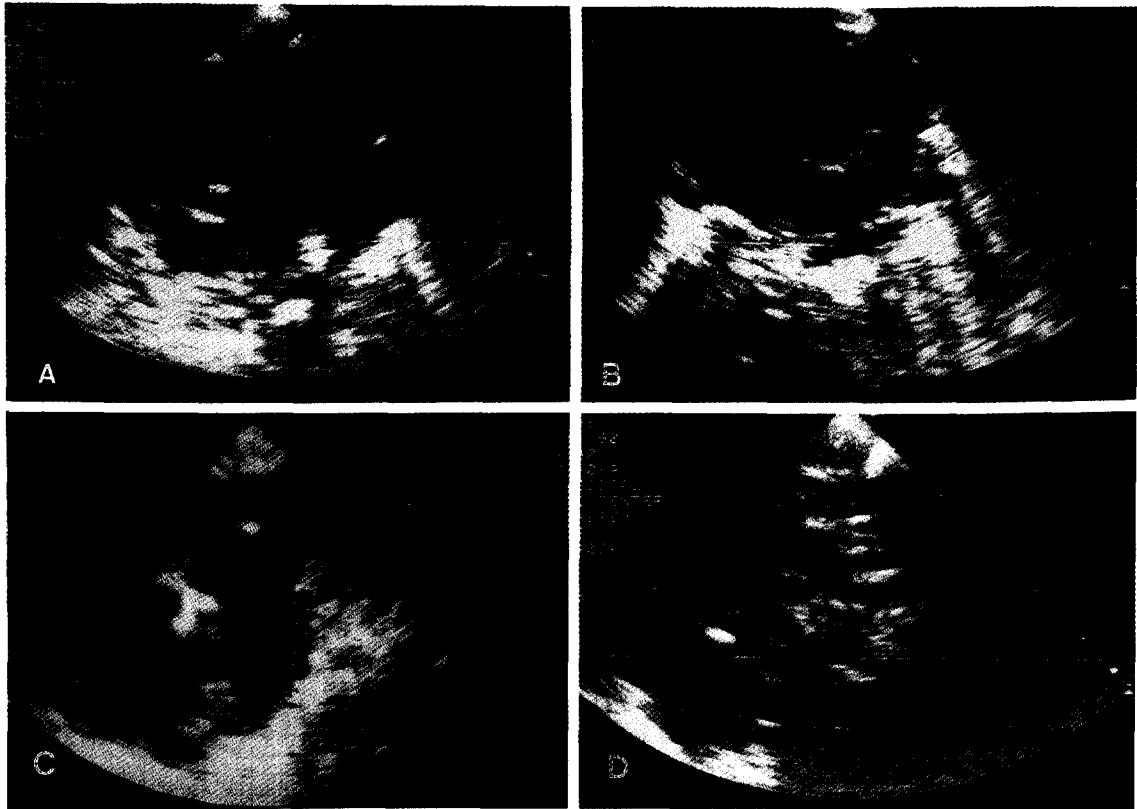


**Fig. 1.** The chest roentgenogram showed moderate cardiomegaly with increased pulmonary vascularity.



**Fig. 2.** Preoperative electrocardiogram showed normal sinus rhythm with biventricular hypertrophy.

tomous relationship of aortopulmonary window, aortic origin of the right pulmonary artery, patent ductus arteriosus and interrupted aortic isthmus. (Fig. 3,4). On angiography the contrast medium injected in the left brachial artery opacified the arch vessels and the ascending aorta, but did not opacify the descending thoracic aorta, suggesting the type A of interrupted aortic arch. In subsequent films the right pulmonary artery was noted to originate from the proximal segment of the

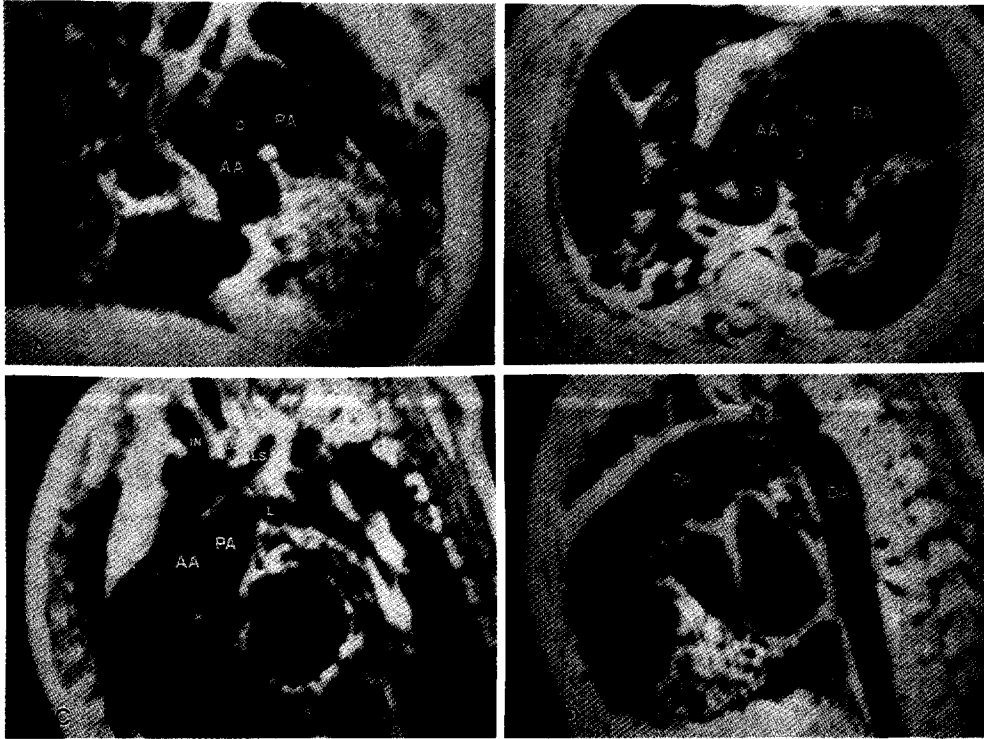


**Fig. 3.** Preoperative echocardiogram : A, The short axis view at the level of great arteries revealed that there was a large aortopulmonary septal defect (APW) between the distal ascending aorta (AAO) and the distal main pulmonary artery (MPA) and the right pulmonary artery (RPA) originated from the posterior surface of the ascending aorta. B, More proximal short axis view showed the two separate semilunar valves (AV, PV). These findings excluded the possibility of persistent truncus arteriosus or proximal type of aortopulmonary window. C, An alteration of the short axis imaging showed the distal continuation of the main pulmonary artery, as a ductus arteriosus, to the descending aorta. D, The long axis suprasternal plane image showed that the arch vessels arise from the transverse aorta but the aortic isthmus portion just distal to the left subclavian artery was interrupted or severely narrowed.

ascending aorta and was opacified preferentially. Both pulmonary arteries were tortuous and freely pulsating (Fig. 5).

A diagnosis of distal aortopulmonary septal defect, aortic origin of the right pulmonary artery, patent ductus arteriosus, intact ventricular septum and interruption of aortic isthmus was made. The patient was planned to be operated by two-stage approach. At the first stage, a left posterolateral thoracotomy was made. Initially a fibrous strand 2.5cm in length was found in the aortic isthmus site. After the arch vessels and patent

ductus arteriosus was mobilized, a 10mm-PTEE tube graft was inserted between the aortic arch just near the left subclavian artery and the descending thoracic aorta. And the ductus was ligated. Then the pericardium was opened anterior to the left phrenic nerve and banding of the pulmonary artery was tried but failed because of the distended and tortuous pulmonary artery. During the first 3 postoperative days the patient was tried to be weaned from the respirator but it could not be done due to the large amount of left-to-right shunt. So the patient was decided to be sub-



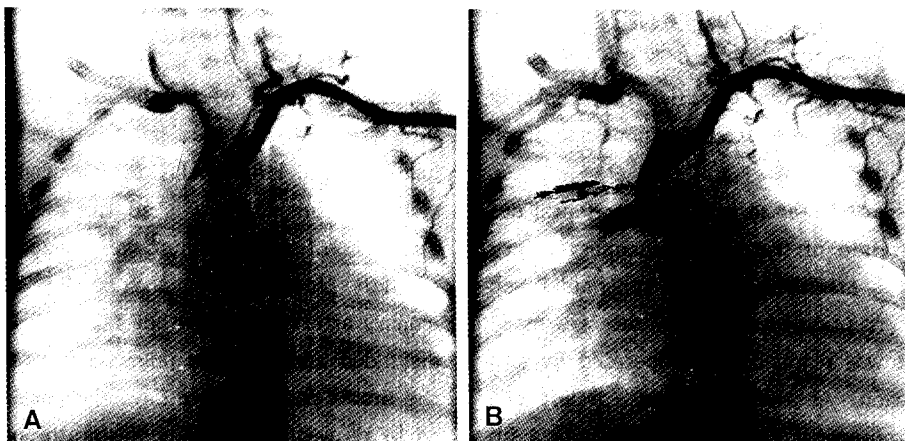
**Fig. 4.** Preoperative magnetic resonance imaging :

A. Oblique coronal MR image equivalent to angiographic right anterior oblique view showed a large aortopulmonary window (D) between distal ascending aorta (AA) and distal pulmonary artery (PA).

B. Oblique axial MR image showed an aortopulmonary window (D) and anomalous origin of right pulmonary artery (R) from posterior wall of ascending aorta. L=left pulmonary artery.

C. Oblique sagittal MR image equivalent to angiographic left anterior oblique view showed interruption of the aortic arch (arrows) distal to the origin of left subclavian artery (LS). IN=innominate artery, LC=left common carotid artery.

D. More steep oblique sagittal MR image showed a small patent ductus arteriosus (open arrow).



**Fig. 5.** Preoperative angiogram : A, Injection in the left brachial artery, anteroposterior projection. The descending aorta just distal to the left subclavian artery was not visualized. B, Subsequent film. There was preferential opacification of the right pulmonary artery from the ascending aorta.

mitted to the second stage total correction. On the 9th day after the first operation the patient was operated with the aid of cardiopulmonary bypass, deep perfusion hypothermia (18°C) and 40 minutes of circulatory arrest.

The aorta was longitudinally opened to reveal a large 12mm aortopulmonary septal defect, together with an anomalous origin of the right pulmonary artery from the posterior surface of the ascending aorta, 2cm above the sinuses of Valvulae. There was a 2mm wide interval between the orifice of the right pulmonary artery and the aortopulmonary defect, which meant that the right pulmonary artery originated as a separate branch from the aorta, not straddling the aortopulmonary septal defect. The right pulmonary artery was tunnelled into the main pulmonary artery through the aortopulmonary septal defect using the Dacron patch by means of 10 interrupted sutures of 5-0 prolene buttressed with spaghetti. Thereafter the aortotomy site of the ascending aorta was enlarged with the patient's pericardial patch by continuous sutures of 6-0 prolene to avoid the supra-valvular aortic stenosis, which might occur in the case of direct closure of the aortotomy site. The second postoperative course was relatively uneventful except the frequently changing pulmonary vascular resistance.

On the 10th day after the second stage repair the infant was weaned from the respirator but, because of persistent feeding problems, was not discharged home until the 29th postoperative day. Now, three and one-half months after operation the infant weighs 6.7Kg and thrives without any symptom.

## DISCUSSION

An aortopulmonary septal defect is a round or oval opening between the ascending aorta and the pulmonary artery as a congenital anomaly in hearts with separated aortic and pulmonary valves.

As stated by Meissner<sup>10)</sup> and Mori<sup>11)</sup> and their associates there are three distinct types of aortopulmonary septal defect according to the location. The first is a proximal, sagittally oriented window-like defect anterior to the right pulmonary arterial orifice. The second type is a distal, obliquely oriented defect involving the pulmonary bifurcation. The last type is the absent aortopulmonary septum as a combined form of proximal and distal defects.

However, aortopulmonary septal defect can be somewhat differently classified into 3 types according to the morphology<sup>6)</sup>. The first is a defect with a more or less circular border, localized approximately halfway between the arterial valves and the bifurcation of the pulmonary trunk. It is usually of moderate or small size. In the second type, the border is not continuous but describes slightly more than 1 turn of a spiral. The third type represents a defect, usually large, in which there is no posterior border.

These various appearance of aortopulmonary defects suggests a different developmental mechanism for each. The most simple explanation is that the aortopulmonary septal defect results from the failure of fusion or malalignment of the conotruncal ridges when, in the truncus arteriosus, two conotruncal ridges form proximally and fuse to create the aortopulmonary septum and more distally, the right and left sixth aortic arches fuse the main pulmonary artery to form the right and left pulmonary arteries and complete formation of the aortopulmonary septum<sup>1,5,12)</sup>. Therefore the aortopulmonary septal defect should be regarded as an error during the partition process of common arterial trunk and the association of anomalies such as the aortic origin of the right pulmonary artery and interrupted aortic isthmus with the aortopulmonary septal defect should be also understood in the viewpoint of this embryogenesis. That is, in the distal type of aortopulmonary septal defect, failure of truncal septation posteriorly

may disturb the normal flow of events and cause malattachment of the pulmonary bifurcation to this undivided truncal segment rather than to the main pulmonary arterial trunk. The right pulmonary artery thus relates to the aorta and the left pulmonary artery to the pulmonary trunk.

There may be a spectrum of the interval between the right and left pulmonary arterial orifices through the aortopulmonary septal defect because the common arterial trunk is dilated and the junction site of the right pulmonary trunk can be anywhere in the posterior wall of the common arterial trunk. Besides, the fact that these abnormal connections in the arterial trunk can bring about the expansion or obliteration of aortic arches should be consecutively considered. During the normal fetal development, the blood flow in the descending aorta is supplied through the ductus (90 percent of main pulmonary arterial flow) and through the isthmus (30 percent of the aortic flow). If the magnitude of blood flow in the ascending aorta becomes reduced by such abnormal connections as aortopulmonary septal defect and aortic origin of the right pulmonary artery, which can siphon additional highly oxygenated blood from the aorta into the pulmonary arterial circuit, the isthmus flow may become interrupted and consequently that segment of the left dorsal aorta between the left fourth and sixth arches may disappear. So the type A of interruption of aortic arch and patent ductus arteriosus may coexist with the distal type of aortopulmonary septal defect and aortic origin of the right pulmonary artery because of direct diversion of the aortic blood flow into the right pulmonary artery during the fetal life. Berry and her associates<sup>5)</sup> recognized the association of these anomalies and regarded it as a specific syndrome rather than random coincidence.

Actually, such as embryologic concepts have greatly contributed the study of pathogenesis of many complex congenital heart anomalies. For example persistent truncus arteriosus,

a hemodynamically & morphologically similar anomaly to aortopulmonary septal defect with aortic origin of the right pulmonary artery is thought to arise from the error involving the neural crest. So persistent truncus arteriosus is frequently noted to be associated with DiGeorge syndrome and the type B of interruption of aortic arch. But these anomalies (often seen in persistent truncus arteriosus) are rarely seen in aortopulmonary septal defect. Rather the type A of interruption of aortic arch is more often occurred in the aortopulmonary septal defect. These observations indicate that aortopulmonary septal defect and persistent truncus arteriosus are pathogenetically unrelated even though located in the same region of the heart<sup>6)</sup>.

Aortopulmonary septal defect with aortic isthmus obstruction causes the rapid onset of severe heart failure after birth. The newborn infant with this anomaly is dependent upon the ductus arteriosus for perfusion of the lower part of the body. As the ductus constricts and as pulmonary vascular resistance decreases, inadequate perfusion of the lower body and tremendous overperfusion of the lungs ensues. Rapid deterioration with profound metabolic acidosis develops and may lead to cardiac arrest. Especially, the infant accompanied with marked rightward displacement of the right pulmonary artery has the higher risk to survive beyond the infancy. Survival of these infants primarily depends upon early suspicion of a congenital cardiac anomaly, early transfer to an appropriate facility, prompt diagnosis with echocardiogram or magnetic resonance imaging and angiography, prevention of ductal constriction with PGE1 infusion and urgent operation. Even though the infant undergoes any operation, the operative mortality is still high. On reviewing of the literature only 3 neonates of 24 reported cases survived operation.

Surgical correction of the patients displaying this syndrome-aortopulmonary septal defect, aortic origin of the right pulmonary artery, intact ven-

tricular septum and interruption of the aortic isthmus requires both repair of hypoplastic or interrupted aortic arch and septation of aortopulmonary septal defect with exclusion of the right pulmonary arterial orifice from the aorta. In order to achieve these surgical goals, there exist many controversies in the methodology. First, one-stage or two-stage approach should be chosen. If comparable results can be achieved, one definitive corrective operation is of course superior to staged procedures. However, when comparing the results in infants with type A or B interrupted aortic arch and ventricular septal defect, the single-stage operation has not produced better results than the two-stage approach developed in an earlier era<sup>13,14</sup>. This fact is true even though techniques of deep perfusion hypothermia have improved and PGE1 is available to improve hemodynamics in these critically ill newborn infants. At this time the choice of the two-stage versus the one-stage approach for an aortopulmonary septal defect with aortic isthmus obstruction must be individualized. In severely acidotic, critically ill neonates two-stage procedure is recommended since few neonates who have been resuscitated from open operations for complex lesions in the neonatal period. For the relatively stable newborn or young infant, complete repair with deep perfusion hypothermia might actually be safer than the two-stage approach. To date there is not enough experience for us to choose between the two options.

If the two stage approach is decided, then the first stage operation has to involve both procedure to reconstruct the interrupted aortic arch and to reduce the left-to-right shunt simultaneously. Whenever considering the reconstruction of the interrupted aortic isthmus, we should also choose meticulously how to do it. If possible, end-to-end direct anastomosis has been known to be best. But in this case, which had so a relatively long interrupted segment that direct anastomosis was not

possible, the interposition of a prosthetic tube graft as wide as possible would provide palliative relief. The other methods using the left subclavian artery can be also considered to establish communication between the aortic arch and the descending aorta but these are inferior to end-to-end anastomosis or tube graft interposition because of restenosis and residual pressure gradient. To reduce the amount of left-to-right shunt, classical pulmonary artery banding is firstly remembered. However we were convinced of the technical impossibility of pulmonary artery banding in cases like this. So the plication method of aortopulmonary septal defect, proposed by Tabak et al<sup>4</sup> in 1983, is thought to be appropriate to decrease the pulmonary blood flow and protect the pulmonary vascular bed. Meanwhile, whenever the plication of aortopulmonary septal defect is difficult to do, early total correction should be carried out because of profuse heart failure, which always disturbs the smooth recovery. Timing of the second-stage operation is elective, although the procedure should be done before any permanent changes in pulmonary vascular resistance occur.

The critical aspect of the second-stage operation is the repair of the aortopulmonary septal defect complicated by aortic displacement of the right pulmonary artery. To do this completely, correct preoperative (or intraoperative) diagnosis of aortic displacement of the right pulmonary artery is important. The right pulmonary arterial orifice cannot simply be excluded from the aorta by tunneling because the simple patch will create supravalvular stenosis. In these circumstances the enlargement of the ascending aorta anteriorly with another patch may be a solution. Because the infants with these defects are often critically ill, the procedure of enlargement of ascending aorta by another patch will be unduly prolonged and may be a particular hazard. So in some cases, detaching the right pulmonary artery from the aorta and anastomosing it to the main pulmonary artery may

appear to be a more practical approach. Anyway, the various kinds of procedure described herein merely add options to the management of the patients with the unusual anomaly of aortopulmonary septal defect, aortic origin of the right pulmonary artery and aortic isthmus interruption. The success of this complicated surgical repair depends upon meticulous design of each step of procedure prior to operation, which should be individualized considering the patient's status and the surgeon's ability.

## REFERENCES

1. Kirklin and Barratt-Boyes. Cardiac Surgery 933-936 New York A Wiley Medical Publication, 1986.
2. Elliotson J : *Case of malformation of the pulmonary artery and aorta. Lancet 1 : 247, 1830.*
3. Gula G, Chew C, Smith RR, Yacoub M. : *anomalous origin of the right pulmonary artery from the ascending aorta associated with aortopulmonary window. Thorax 33 : 265, 1978.*
4. Tabak C, Moskowitz W, Wagner H, Weinberg P, Edmunds LH Jr, : *Aortic Isthmic Hypoplasia. J. Thorac. Cardiovasc. Surg. 86 : 273, 1983.*
5. Berry T.E. Bharati s, Muster AJ, Idriss FS, Santucci B, Lev M, Paul MH. : *Distal aortopulmonary septal defect, aortic origin of the right pulmonary artery, intact ventricular septum, patent ductus arteriosus and hypoplasia of the aortic isthmus : A newly recognized syndrome. Am. J. Cardiol. 49 : 108, 1982.*
6. Kutsche LM, Lodewyk HS, Mierop V. : *Anatomy and pathogenesis of aortopulmonary septal defect. Am. J. Cardiol 59 : 443, 1987.*
7. Braunlin E, Peoples WM, Freedom RM, Flyer DC, Goldblatt A, Edwards JE. : *Interruption of the aortic arch with aortopulmonary septal defect. An anatomic review Pediatr. Cardiol. 3 : 329, 1982.*
8. Mendoza DA, Ueda T, Nishioka K, et al : *Aortopulmonary window, aortic origin of right pulmonary artery, and interrupted aortic arch : Detection by two-dimensional and color Doppler echocardiography in an infant. Pediatr. Cardiol. 7 : 49, 1986.*
9. Yoo SJ, Choi YH : *Angiocardiograms in congenital heart disease. Seoul Korea Medical Publishing Company, 1989.*
10. Meisner H, Schmidt HP, Sebening F, Klinner W. : *Surgical correction of aortopulmonary septal defects : A review of the literature and report of eight cases Dis. Chest 58 : 750, 1968.*
11. Mori K, Ando M, Takao a, Ishikawa S, Imai Y. : *Distal type of aortopulmonary window. Br. Heart J. 40 : 681, 1978.*
12. Sabiston and Spencer : *Surgery of the Chest 4th Edition, 1148-1150, Philadelphia PA, W.B. Saunders Company, 1990.*
13. Fishmon NH, Bronstein MH, Berman WJr, Roe BB, Edmunds LH Jr, Robinson SJ, Rudolph AM : *Surgical management of severe aortic coarctation and interrupted aortic arch in neonates. J. Thorac. Cardiovasc. Surg. 71 : 35, 1976.*
14. Allavd JR, Williams RL, Dobell AR : *Interrupted aortic arch. Factors influencing prognosis. Ann Thoracic Surg 21 : 243, 1976.*
15. Luisi SV, Ashraf MH, Gula G, Radley-Smith R, Yacoub M : *Anomalous origin of the right coronary artery with aortopulmonary window : Functional and surgical considerations. Thorax 35 : 446, 1980.*