

부분 폐정맥 환류 이상과 심실 중격 결손을 동반한 대동맥 축착증의 완전교정

— 1예 보고 —

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Surgical Management of Coarctation of the Aorta with a Ventricular Septal Defect and Coexisting Partial Anomalous Pulmonary Venous Connection

— A case report —

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A newborn girl with a partial anomalous pulmonary venous connection, coarctation of the aorta, and ventricular and atrial septal defects underwent a complete repair successfully at 19 days of age. In this case, the left upper pulmonary vein was connected to the left innominate vein via an atypical vertical vein.

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Key words: 1. CHD, acyanotic
2. Pulmonary vein, partial anomalous return
3. Aortic arch anomalies

CASE REPORT

A newborn girl weighing 3.40 kg was referred to our hospital with a murmur at birth and tachypnea. The arm-to-leg blood pressure gradient was 30 mmHg. An echocardiogram demonstrated an unrestrictive VSD with posterior malalignment of the outlet septum, a large ductus, a juxtaductal coarctation at the level of the left subclavian artery, and good biventricular function without subaortic stenosis. The color Doppler imaging, however, showed abnormal upward blood flow around the innominate vein. The pulmonary arteriogram revealed that a left upper pulmonary vein drained

into the left innominate vein via a vertical vein (Fig. 1A, B).

Surgical correction was performed at 19 days, using a median sternotomy. The external anatomy confirmed an atypical vertical vein that was 3 mm long and connected to the left innominate vein obliquely.

Cardiopulmonary bypass was initiated and followed by double aortic and bi-caval cannulations. The right side of the distal ascending aorta was cannulated near the base of the innominate artery, and the distal descending thoracic aorta was cannulated just superior to the diaphragm through the median sternotomy.

The pH during cardiopulmonary bypass was managed using

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본 논문의 저작권 및 전자매체의 지적소유권은 대한흉부외과학회에 있다.

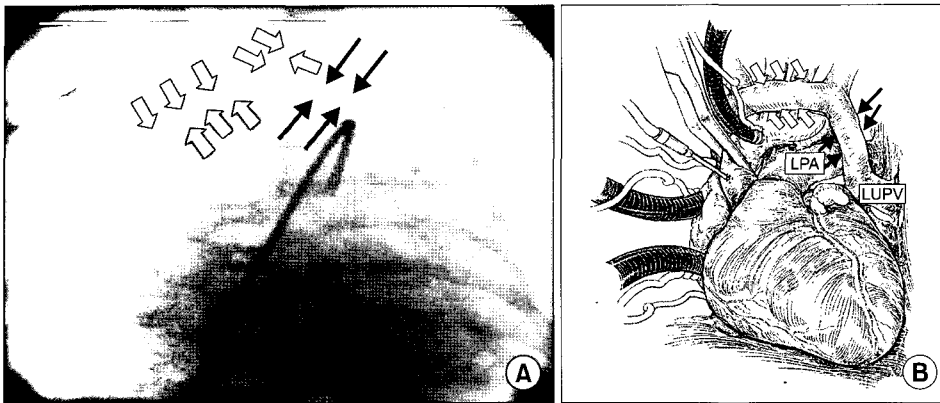


Fig. 1. (A) Preoperative pulmonary arteriography. (B) Operative scheme. Arch reconstruction was performed. Vertical vein (black arrow) is connected to the left innominate vein (white arrow). See details in the text. LPA=Left pulmonary artery; LUPV=Left upper pulmonary vein.

the alpha-stat strategy. The patient was cooled to 30.0°C while undergoing cardiopulmonary bypass. Closure of the VSD with an autologous pericardial patch was performed after cardioplegic arrest.

After cooling to deep hypothermia (18°C), the arterial cannula was advanced into the innominate artery, and held in place with a snare, while continuous low flow cerebral perfusion was maintained via the innominate artery. Subsequently, reconstruction of the aortic arch and coarctectomy were carried out; a continuous 8-0 Prolene suture using an extended end-to-end anastomosis technique was performed. Then, full flow bypass was restarted and a spontaneous heart beat with sinus rhythm resumed. The division of the vertical vein was performed, and the distal left upper pulmonary vein was anastomosed to the auricle of the left atrium; as widely as possible under full flow bypass. Continuous low flow cerebral perfusion was maintained for 50 minutes at a flow rate of about 80 ml/kg/min.

The patient was successfully weaned from cardiopulmonary bypass and the cannula in the descending aorta was removed without difficulty. In the operating room, the postoperative upper-lower extremity blood pressure gradient was estimated using the ascending and descending aortic cannula simultaneously. The pressure gradient was about 1 mmHg.

The postoperative clinical courses were uneventful. The postoperative echocardiogram demonstrated a widely patent aortic arch with good laminar flow, and drainage of the left upper pulmonary vein into the left atrium. The patient was discharged on postoperative day 19.

DISCUSSION

There are only two cases in the medical literature that have been reported to have a combination of aortic coarctation and PAPVC [1,2]. However, an additional finding of a VSD was not observed in either of these two cases. Our patient is reported to illustrate the unique anatomic features of the association of PAPVC with coarctation of aorta and a co-existing VSD in a neonate. This is the 1st report of successful surgical correction of this complex cardiac abnormality in neonate.

In our patient, a vein from the left upper lobe connected to the left innominate vein via a persistent early embryonic pathway. This is referred to a vertical vein because of its orientation. At the other end of the anatomic spectrum, a connecting vein between the left pulmonary vein and left innominate vein may be a “persistent left superior vena cava”. Infrequently, an atretic segment (ligament of Marshall) of the left superior vena cava (LSVC) is present. Therefore, to avoid inadvertent occlusion of the pulmonary venous drainage, it is important to differentiate between a vertical vein and a persistent LSVC before surgical or transcatheter interventions [3]. However, an atretic LSVC was not found in this case.

Pigula et al. [4] reported that continuous low flow cerebral perfusion and indirect innominate artery cannulation were effective and safe adjunct procedures to the surgical repair of aortic arch pathology in neonates, and that with this approach more precise anatomic repairs could be achieved without the

pressing need to minimize the duration of deep hypothermic circulatory arrest [4,5].

Coarctation of the aorta with VSD and coexisting PAPVC is a rare combination of abnormalities; total correction can be successfully carried out with continuous low flow cerebral perfusion.

However, associated anomalies coexisting with PAPVC tend to be overlooked without vigilant preoperative evaluation. Therefore, we suggest that preoperative cardiac cineangiography should be considered positively, when questionable findings are found on echocardiography, to avoid missing a minor coexisting anomaly.

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=국문 초록=

심부전으로 입원한 3.4 kg 여자 환아에게서 심실 중격 결손을 동반한 대동맥 협착증과 함께 좌측 폐정맥의 환류 이상이 의심되었다. 심혈관 영화 촬영상에서 좌측 상부 폐정맥이 무명 정맥으로 환류되는 것을 확인하였고 생후 19일째 수술적 완전 교정을 시도하여 성공적인 결과를 얻었기에 보고하는 바이다.

- 중심 단어 : 1. 비청색증 심장기형
 2. 부분 폐정맥 환류 이상
 3. 대동맥 궁 기형