

# Cor Triatriatum with Infracardiac Total Anomalous Pulmonary Venous Drainage

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## =Abstract=

We report one case of an 18-day-old female patient, weighing 3.4 kg, with severe cyanosis. The diagnosis was made with only transthoracic echocardiography, which revealed cor triatriatum with an atretic small opening of fibromuscular membrane, obstructive infracardiac total anomalous pulmonary venous drainage(TAPVD), severely restrictive interatrial communication, and scanty mitral inflow and aortic forward flow. The preoperative decision-making for biventricular repair was not easy due to collapsed left heart system caused by remarkably reduced blood flow. An emergent operation was performed due to severe cyanosis. All left heart structures were somewhat hypoplastic but thought to be adequate for systemic circulation. Biventricular repair was done without specific intraoperative problems. The postoperative course was uneventful. The patient has been doing well with no evidence of pulmonary vein stenosis or mitral regurgitation for 4 months after operation.

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**Key words:** 1. Cor triatriatum  
2. Pulmonary vein, total anomalous return

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본 논문의 저작권 및 전자매체의 지적소유권은 대한흉부외과학회에 있다.

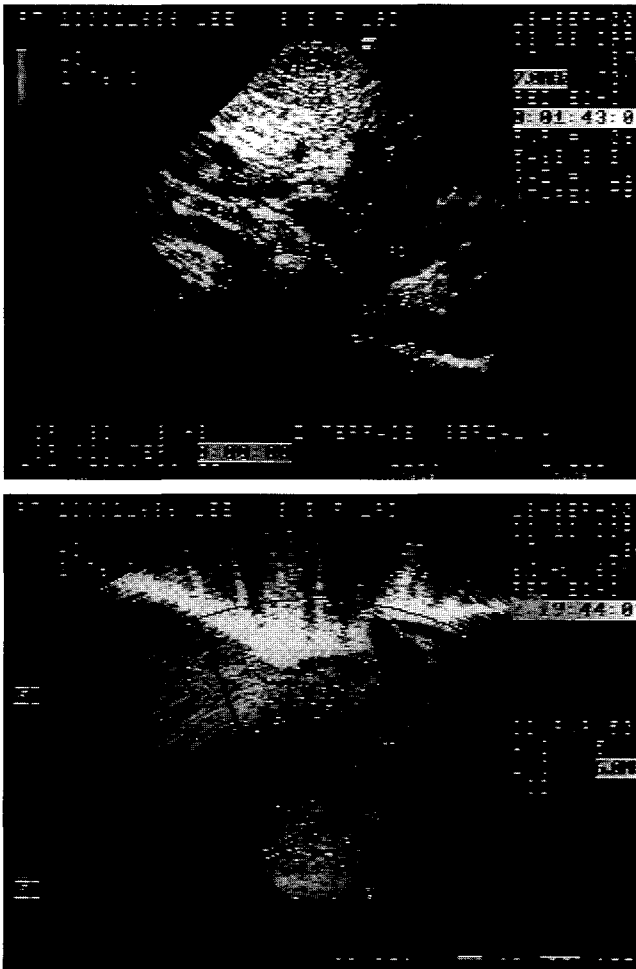


Fig. 1. Preoperative transthoracic echocardiography(TTE) shows that an anomalous vertical descending vein originated from the proximal chamber(upper) was drained into the portal venous system and drainage site(double arrow) was obstructive(lower). VV, vertical vein; PC, proximal chamber; DAO, descending aorta; PV, portal vein.

### Case

An 18-day-old female patient, weighing 3.4 kg, was referred to the our hospital because of severe cyanosis and dyspnea. Arterial blood gas analysis showed pH 7.44, PCO<sub>2</sub> 43.5 mmHg, PO<sub>2</sub> 22.3 mmHg, SaO<sub>2</sub> 40% in room air.

Echocardiographic findings were compatible with cor triatriatum associated with the infracardiac total anomalous pulmonary venous drainage(TAPVD) and ductal dependent systemic circulation. The interatrial communication between the proximal chamber and right atrium was severely restrictive. All the pulmonary veins were drained into a proximal chamber. An anomalous vertical descending vein was originated from the



Fig. 2. Suprasternal notch view shows that the ascending aortic flow was retrogradely maintained via the ductal dependent systemic circulation through a large patent ductus arteriosus. AAO, ascending aorta; DAO, descending aorta



Fig. 3. Apical four chamber view of TTE shows the thick fibromuscular membrane of cor triatriatum(arrow heads), remarkably collapsed distal chamber(DC) of left atrium, poorly visualization of mitral valve(double arrow), and squeezed left ventricle(LV). PC, proximal chamber; RA, right atrium; RV, right ventricle

proximal chamber drained into portal venous system, and its hepatic drainage site were obstructive(Fig. 1). The ascending aortic flow was retrogradely maintained via the ductal dependent systemic circulation through a large patent ductus arteriosus (PDA), 4 mm in diameter(Fig. 2). Because the forward flow through the stenotic opening of cor triatriatum was scanty and the distal chamber of left atrium was completely collapsed, mitral valve was functionally atretic and left ventricle was squeezed with 3.6 ml/m<sup>2</sup> of end diastolic volume(Fig. 3).

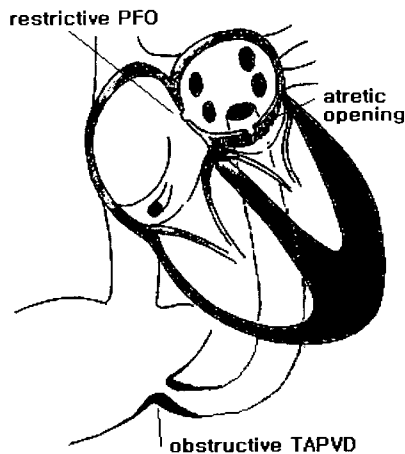


Fig. 4. Schematic diagram of intraoperative finding of nearly atretic cor triatriatum, obstructive infracardiac total anomalous pulmonary venous drainage, severely restrictive interatrial communication. ASD, atrial septal defect; TAPVD, total anomalous pulmonary venous drainage

Because the patient was critically ill, emergent surgical correction was done without invasive investigation on the next day. PDA was divided before performing cardiopulmonary bypass(CPB). After aorta cross clamp(ACC) was done and cold diluted blood cardioplegia was antegradely infused, right atrium was incised and atrial septotomy was made. Total circulatory arrest(TCA) was adopted for operative assessment. Intraoperative finding was compatible to the preoperative echocardiographic findings(Fig. 4). The left atrium was divided into a proximal and a distal chamber by the circular fibromuscular membrane and its opening was nearly atretic, 2 mm in diameter. All pulmonary veins were drained to the proximal chamber and their orifices were seemed to be grossly normal. An anomalous vertical vein originated from the posterior portion of proximal chamber was drained into portal venous system along the retrocardial course. The endocardium of proximal chamber, septal wall, and circular membrane were fibrotic and severely thickened.

The atrial septal wall and fibromuscular membrane were completely excised firstly. And then the morphology and size of mitral valve and left ventricle were evaluated and slightly hypoplastic, but the biventricular repair was thought to be possible. CPB was restarted and systemic rewarming was done. And atrial septum was reconstructed with the glutaraldehyde-treated autologous pericardium. The anomalous vertical vein was doubly ligated at the diaphragmatic level at the end of cardiac

procedures.

CPB was weaned without difficulty. Intraoperative transesophageal echocardiography showed good aortic valve, mitral valve, and ventricular function. Modified ultrafiltration was done. Total CPB, ACC, and TCA time were 84 min, 36 min, 18 min, respectively. The postoperative course was uneventful and mechanical ventilation was weaned at the postoperative day 3. The patient has been doing well with no evidence of pulmonary vein stenosis or mitral regurgitation for 4 months after operation.

## Comment

Cor triatriatum is a rare congenital anomaly characterized by an abnormal division of the left atrium into a proximal and a distal chamber. The proximal chamber receives, in most cases, all the pulmonary veins and the presence of partial anomalous pulmonary venous drainage is not uncommon. On the contrary, the combination of cor triatriatum with total anomalous pulmonary venous drainage(TAPVD) appears to be very uncommon and is a prerequisite for the maintenance of hemodynamic stability, especially in the case of atretic cor triatriatum associated with the severely restrictive interatrial communication. The embryogenesis of cor triatriatum remains controversial and is usually explained by the entrapment theory, proposed by Van Praagh and Corsini<sup>1)</sup>, or failed incorporation of the common pulmonary veins into the left atrium<sup>2)</sup>. Concomitant occurrence of cor triatriatum and TAPVD is not well explained using either theory.

The surgical correction for cor triatriatum combined with total anomalous pulmonary venous connection(TAPVC) had been reported in the literature<sup>3,4)</sup>. But the case of cor triatriatum with TAPVD was firstly reported by Al-Fadley et al<sup>5)</sup> in 1992. Their patient had mitral atresia, a good-sized communication between the proximal and distal chambers of the left atrium, and no obstruction of supracardiac TAPVD. Because there was no hemodynamic instability or severe cyanosis, the patient was discharged without operation.

However, our case differs from the previous report<sup>5)</sup> in that there was nearly atretic cor triatriatum, severely restrictive interatrial communication, infracardiac type of obstructive TAPVD and emergent operation was needed due to severe cyanosis. Though all pulmonary veins were connected to the proximal chamber, an anomalous descending vein provided an

alternative pathway from the left atrium to the portal venous system and its drainage site was obstructive. The mitral inflow through the very small opening of the fibromuscular membrane was very scanty. The distal chamber of left atrium and left ventricle were remarkably collapsed. Preoperatively we were not sure for biventricular repair due to collapsed left heart system. However, the intraoperative findings for the anatomy of mitral valve and left ventricle were grossly acceptable for systemic circulation. Biventricular repair was done without specific intraoperative problems. Herein, we report one case had atretic cor triatriatum with obstructive infracardiac TAPVD, severely restrictive interatrial communication, and functional mitral atresia but biventricular repair could be performed successfully. Our case demonstrates the difference between total anomalous pulmonary venous drainage and connection, terms that are often used synonymously.

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### =국문초록=

저자들은 심한 청색증을 주소로 내원한 체중 3.4 kg의 생후 18일된 여아를 보고한다. 환자는 경흉부 심장초음파로만 진단되었으며 직경이 2 mm 정도의 매우 작은 구멍을 가진 삼심방증과 횡격막하형의 폐쇄성 총폐정맥환류 이상, 매우 작은 심방중격결손, 그리고 승모판과 대동맥판의 전향성 혈류가 거의 없는 상태였다. 환자는 술전 검사에서 삼심방증의 매우 작은 구멍을 통한 전향성 혈류가 거의 없고 원위부 좌심방 및 좌심실의 심한 허탈로 인해 수술 전 양심실성 교정 가능성에 대한 판단이 매우 어려웠다. 심한 청색증으로 응급 수술을 시행하였으며 수술중에 관찰한 승모판 및 좌심실 구조물들의 발육부전이 있었지만 체순환을 감당할 수 있다고 판단되어 양심실성 교정을 시행하였다. 수술 후 매우 양호한 경과를 보였다. 환자는 퇴원 후 4개월째 외래추적증으로 심장초음파 검사에서 폐정맥 협착이나 승모판 기능에 이상은 없었다.

- 중심 단어: 1. 삼심방증  
2. 총폐정맥환류 이상, 횡격막하형