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Total Anomalous Pulmonary Venous Connection
— Autopsy Analysis of Seven Cases —

Jeong Ryul Lee*, Yong Jin Kim, Joon Ryang Rho,
Jeong Wook Seo, Heung Jae Lee, Ghee Young Choe,
Yong Soo Yun and Kyung Phill Suh

Department of Thoracic and Cardiovascular Surgery, Seoul National University College of Medicine, Seoul 110-744, Korea

=Abstract=**We studied 7 autopsied cases of total anomalous pulmonary venous connecton (TAPVC). Three cases were supracardiac types, showing drainage to the innominate vein through the left vertical vein. Stenosis at the beginning of the vertical vein was associated in Case 1. The left upper pulmonary vein was connected distal to the stenosis, and the left upper pulmonary lobe were severely congested after surgical ligation of the upper portion of the vertical vein and anastomosis between the common pulmonary vein and left atrium. The vertical vein in Case 2 was interposed between the left pulmonary artery and the left main bronchus, and the long segment was stenotic. The collateral channel through the paraesophageal venous plexus was present. An obstructing or stenotic segment was not found along the whole pulmonary venous pathway in Case 3. One case was a cardiac type in which both right and left pulmonary veins united to produce a common pulmonary venous channel draining into a huge coronary sinus (Case 4). Case 5 and Case 6 were infracardiac types draining into a common hepatic vein through a small opening. The vertical segment of the common pulmonary veins was short, and individual pulmonary veins were slender and long. Case 7 was a mixed form of an anomalous drainage through the portal vein and the right superior caval vein, respectively. We could find the common features of the long and slender individual pulmonary veins in these cases and short transverse common pulmonary vein segments. Unifocal narrowing of 1 pulmonary vein was seen in 1 supracardiac type case, as well as in a mixed supracardiac type and infracardiac type case, which may be present as an unexplained pulmonary infiltration before and after surgery.**

Key Words: *TAPVC, Pulmonary infiltration*

INTRODUCTION

Since 1798 when Wilson reported the first patient whose entire pulmonary venous system drained into the coronary sinus, the total anomalous pulmonary venous return, though it constitutes only 1%

* Author for correspondance
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to 2% of the congenital heart disease, still represents a difficult disease entity for diagnosis and management.

The majority of patients are symptomatic within the first few weeks or months of life, and more than 80% of infants born with TAPVC die before 1 year of age. Because of the poor survival rate, the diagnosis of TAPVC is in itself an indication for operation. The operative mortality in infants has considerably decreased since the 1960s, which shows the great improvements made in the therapy of this disease. Bove *et al.* (1975), Lincoln *et al.* (1988), and Turley *et al.* (1980) have pointed out the high pulmonary vascular resistance, high pulmonary arterial pressure, restrictiveness of interatrial communication, type of anomaly, presence of pulmonary venous obstruction, left ventricular function, and morphology as the factors adversely influencing the prognosis.

Snellen *et al.* (1968) and Delisle and associates (1976) have reported on the largest series of autopsied cases of TAPVR with a special emphasis on the classification of the patterns of pulmonary venous drainage. Here the authors have experienced 7 autopsied cases of TAPVC from 3 hospitals in Korea (Seoul National University Children's Hospital, Inha University Hospital and Sejong General Hospital) since 1983 and did detailed pathologic studies of the anatomy, giving particular effort to clarify the sites and extents of the obstruction of the pulmonary venous connection.

CASE REPORTS

Case 1

This 26-day-old female patient showed cyanosis and tachypnea after delivery by Caesarian section because of premature rupture of the membrane. The gestational age of the infant was 36 weeks, and the birth weight 2.82 kg. Her respiration rate was 70-80 breath/min, grade II/VI systolic murmur was heard at the left sternal edge, and the liver was enlarged 3cm below the costal margin. Echocardiography revealed supracardiac type TAPVC. All pulmonary veins drained into the left innominate vein via a vertical vein. Also small patent ductus arteriosus and secundum type atrial septal defect were seen. The operation was done. Anastomosis between the common pulmonary venous channel and the overlying left atrium was constructed with a ligation of the vertical vein and patent ductus. The atrial septal defect was closed primarily via

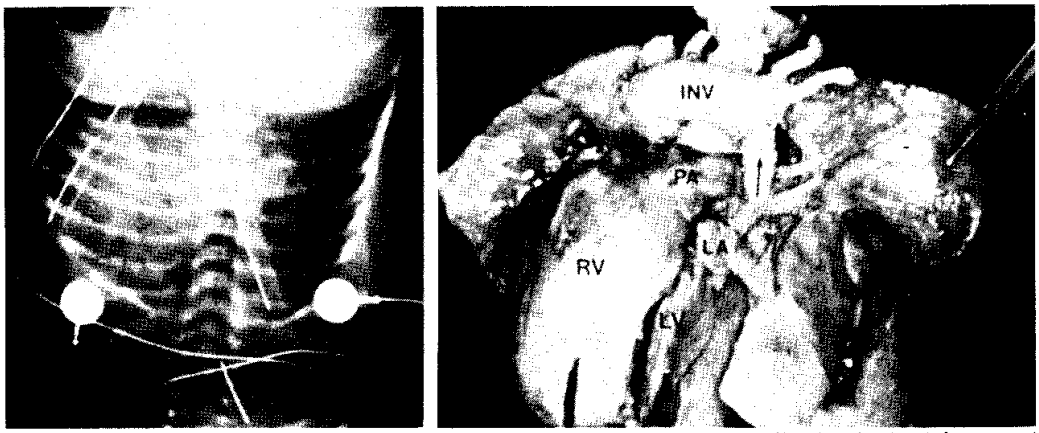

Fig. 1A. (Case 1) Postoperative chest reontgenogram demonstrates severe infiltration in the left upper lung field.

Fig. 1B. (Case 1) Thoracic organs viewed from the front. The vertical vein connects the common pulmonary vein to the innominate vein (INV). The left upper pulmonary vein draining the left upper lobe connects to the vertical vein above the stenotic segment. Surgical ligation of the vertical vein is done between the left upper pulmonary vein and the common channel (→). LA = left atrium; LV = left ventricle; PA = pulmonary artery; RV = right ventricle

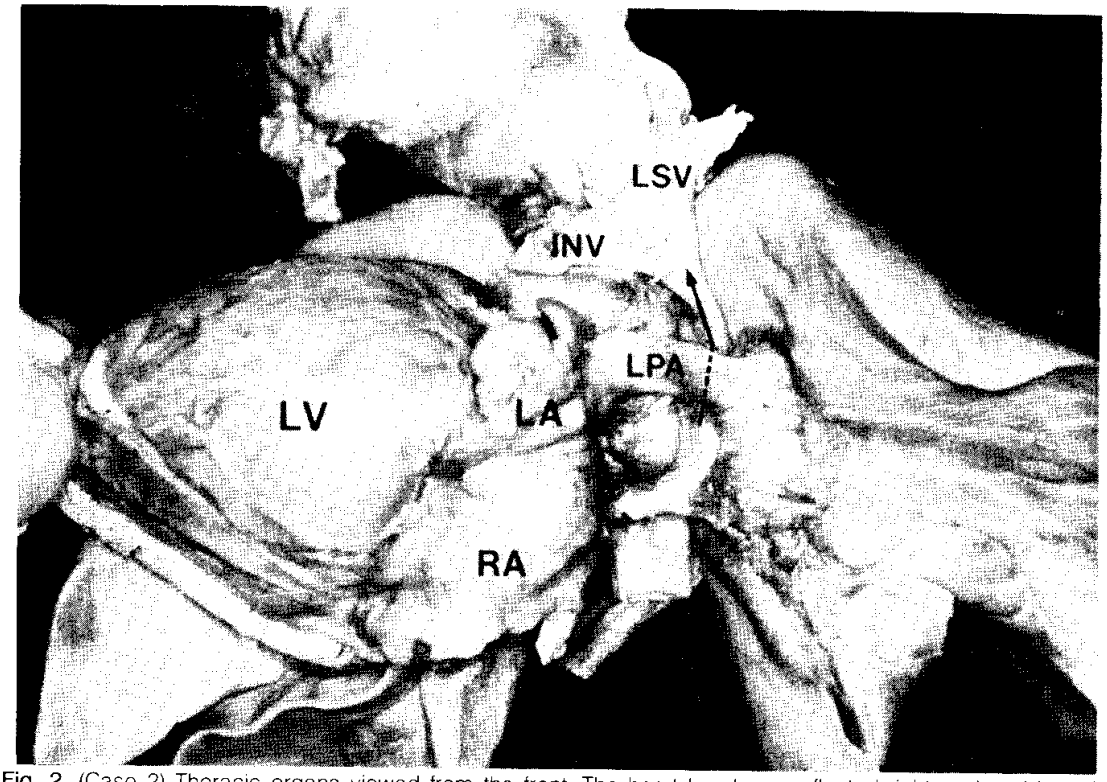

Fig. 2. (Case 2) Thoracic organs viewed from the front. The heart has been reflected rightward and laterally exposing the pulmonary veins. The vertical vein connects to the innominate vein (INV) and interposed between the left pulmonary artery (LPA) and the left main bronchus. LA = left atrium; LV = left ventricle; LSV = left superior vena cava; RA = right atrium

the right atriotomy. The patient's postoperative roentgenogram demonstrated an unexplained infiltrates in the left upper lung field (Fig. 1A). The patient died 8 hours after the operation.

Autopsy findings: The arrangement of the abdominal organs and the lobation of the lungs were normal. The heart was left-sided with its apex pointing to the left. The aortic arch was left-sided, and the aortic coarctation, 0.2 cm in diameter, was found at the isthmic area, while the ductus was ligated. The right upper and lower pulmonary veins and left pulmonary vein were slender and long, draining into a short common pulmonary venous channel. There was an abrupt narrowing at the junction between the common venous channel and the vertical vein. The left upper pulmonary vein draining the left upper lobe was connected to the vertical vein above the stenotic segment. Surgical ligation of the vertical vein was inserted between the left upper pulmonary vein and innominate vein (Fig. 1B). Postmortem examination of the lungs revealed severe congestion and hemorrhage in the left upper lobe.

Case 2

This 30-day-old male infant was diagnosed as a TAPVR supracardiac type. A cineangiocardiogram revealed that both the right and left pulmonary veins drained into the innominate vein through the vertical vein. The infant died of intractable heart failure before operation could be done.

Autopsy findings: Visceroatrial situs and ventricular loop were normal. The right upper and lower pulmonary veins and left lower pulmonary vein were long and slender. Drainage from the left upper lobe was through 4 small pulmonary veins connect-

ed separately to the vertical vein, which was connected to the innominate vein and the paraesophageal venous plexus. The vertical vein was interposed between the left pulmonary artery and left main bronchus and showed diffuse narrowing (Fig. 2). Paraesophageal venous plexus in front of the esophagus was a dilated thin-walled vessels with an uneven diameter. The foramen ovale was patent, measuring 0.4 × 1.1 cm. Ductus arteriosus was patent, but was long and narrow, measuring 1.2 cm in length and 0.2 cm in diameter. Multiple pulmonary infarcts were seen on the lower lobe of both lungs.

Case 3

This 5-month-old female infant was admitted to the hospital because of of cyanosis, tachypnea, and oliguria. On physical examination, grade II/VI systolic murmur was heard along the left sternal border. The liver was enlarged 2 cm below the right costal margin. A chest roentgenogram showed findings of cardiomegaly and pulmonary congestion. The patient died soon after admission with severe congestive heart failure before further diagnostic work-up or surgical therapy.

Autopsy findings: The patient's visceroatrial situs and ventricular loop were normal. The right ventricle was hypertrophied and dilated. Right and left upper and left lower pulmonary veins joined and formed a common pulmonary vein, 1.5 cm in length, which drained into the innominate vein through the vertical vein, while the left upper pulmonary vein drained separately into the vertical vein. There was no obstruction along the entire pulmonary venous channel. A secundum atrial septal defect, 0.4 cm in diameter, was seen. The ductus arteriosus was not patent. The kidney showed evidence of medullary congestion.

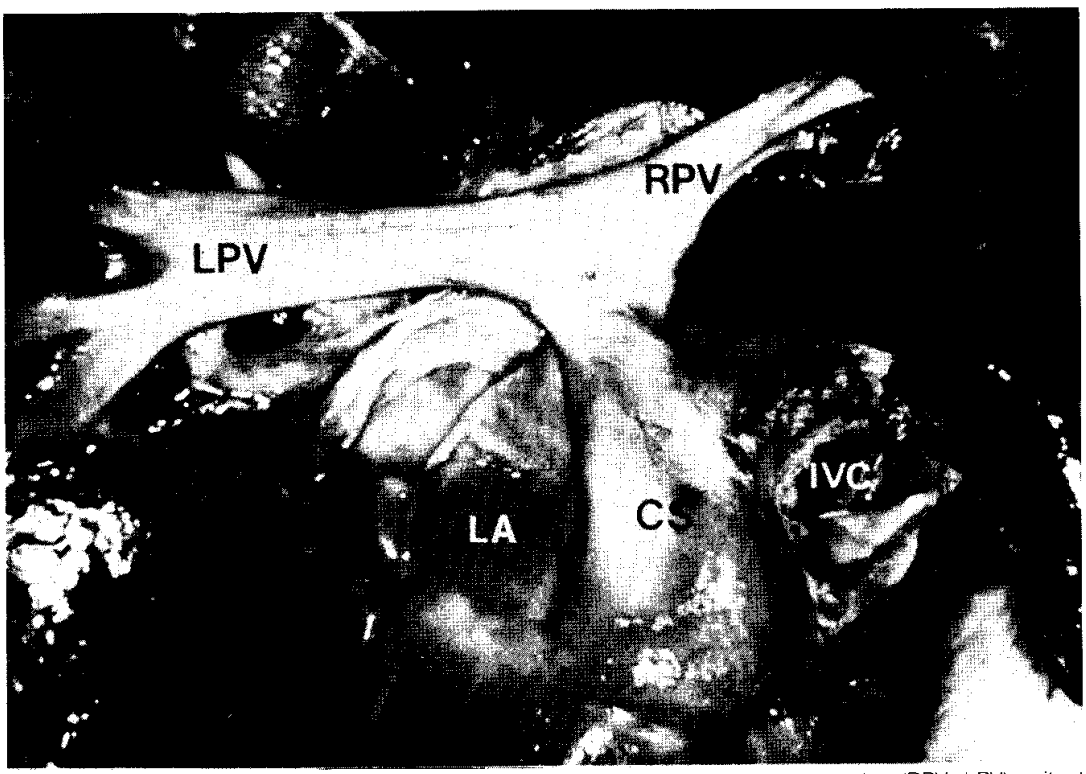

Fig. 3. (Case 4) The heart viewed from the back. Both right and left pulmonary veins (RPV, LPV) unite to produce a common pulmonary venous channel with its left portion longer than the right, which drains directly into the huge coronary sinus (CS).

Case 4

It was noted that this 5-month-old male patient had heart disease since birth. He was admitted to the nospital because of cyanosis and poor feeding. On admission his body weight was 5.2 kg, grade III/VI systolic murmur was heard along the left sternal border, and the systemic arterial oxygen tension was 42 mmHg. Cardiac catheterization was performed. A marked step-up in oxygen saturation was seen at the right atrial level. A cineangiocardiogram suggested that all the pulmonary veins drained to the coronary sinus. Surgical correction was done, but the patient expired on the postoperative 12th day due to disseminated intravascular coagulation and cerebral hemorhage after long standing ventilatory support.

Autopsy findings: The arrangement of the abdominal organs and the lobation of the lungs were normal. The heart was left-sided with its apex pointing to the left. The left atrial size was small and a secundum-type atrial septal defect, 1.2 cm in diameter, was noticed. Both right and left pulmonary veins joined to form a common pulmonary venous channel, its left portion 2.4 cm longer than the right, which drained directly into a huge coronary sinus. It had an oval-shaped orifice 1.2 cm long and 1.4 cm wide. Right upper and lower pulmonary veins opened into the coroanry sinus separately. There was no significant obstruction along the anomalous pulmonary venous pathway (Fig. 3). Multiple intracerebral and subarachnoid hemorrhage were also found.

Case 5

This 70-day-old male infant was admitted to the hospital with a history of increasing cyanosis, dyspnea, and failure to thrive. Physical examination disclosed generalized cyanosis and rapid shallow respiration. The liver edge was palpable 3cm below the right costal margin, and grade II/VI systolic murmur was heard along the left sternal border. A chest roentgenogram showed moderate cardiomegaly and pulmonary congestion. Further investigation could not be done because the patient died several hours after arriving at the emergency room.

Autopsy findings: Atrial situs and ventricular loop were normal. Two pulmonary veins on both sides joined to form a common transverse pulmonary vein which drained into a common hepatic vein ,0.5 cm in diameter, via a descending vein, 0.7 cm in diameter. The size of the common pulmonary vein was larger than respective pulmonary vein, 0.5 cm in diameter, but smaller than that of the vertical vein, 0.7 cm in diameter. The right-sided common hepatic vein was shorter than the left, 0.4 cm vs 0.7 cm in diameter. The patent foramen ovale (0.5 cm in diameter), was present with pinpoint opening. The ductus, measuring 0.5 cm in external diameter and 0.8 cm in length, was closed.

Case 6

This 3-month-old female infant was brought to the emergency room because of tachypnea, generalized cyanosis, failure to thrive, and congestive heart failure. The liver was palpable two finger breadths below the right costal margin. A chest roentgenogram showed cardiomegaly and pulmonary congestion. A two-dimensional echocardiogram revealed a flaring Doppler image at the common hepatic vein. The infant died 4 hours later.

Autopsy findings: Visceroatrial situs and ventricular loop were normal. Long and narrow pulmonary veins from both lungs united to form a short common pulmonary venous channel which drained into the common hepatic vein through a short descending vein. The oblique conjoined pulmonary vein from the left lung was slightly longer than the right one (Fig. 4A). The calibers of the vessels were similar, measuring 0.4 cm in diameter, but the descending vein became constricted at the point of opening to the common hepatic vein(Fig. 4B). The foramen ovale was patent, and the left superior caval vein drained into the coronary sinus. The ductus arteriosus was patent.

Case 7

This 28-day-old male patient was brought to the emergency room with a history of cyanosis and dyspnea since he was 10 days old. grade II/VI systolic murmur was heard along the left sternal border. Chest roentgenogram showed cardiomegaly and pulmonary congestion. He died 3 hours after arrival.

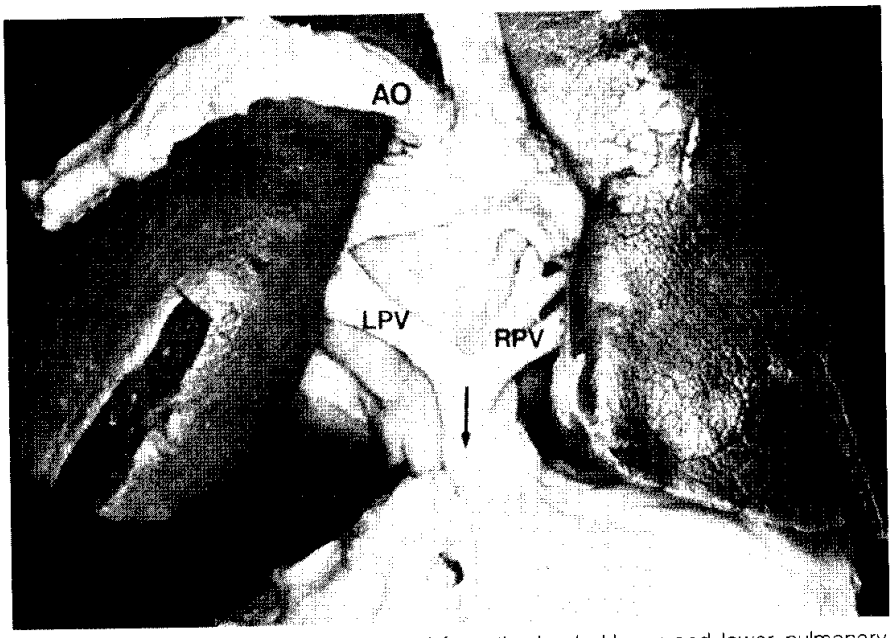

Fig. 4A. (Case 6) Thoracoabdominal organs viewed from the back. Upper and lower pulmonary veins from each lungs unite to form the left and right pulmonary veins (LPV, RPV), which in turn to form the short common pulmonary vein. It descends obliquely through the diaphragm and opens into the common hepatic vein (→). AO = aorta

Fig. 4B. (Case 6) Opening of the right atrium shows foramen ovale (FO) and the small constricted opening in the common hepatic vein to which the descending vein drains. IVS = inferior vena cava; SVC = superior vena cava; TV tricuspid valve

Autopsy findings: The cardiac position and atrioventriculoarterial connections were normal. The right upper pulmonary vein was connected directly into the right lateral aspect of the superior vena cava 0.5 cm above the veno-atrial junction. Other pulmonary veins from the right middle and lower lobes and those from the left lung joined to form a common pulmonary venous chamber and drained into the portal vein through a long descending vein. Individual pulmonary veins from each lobes of the lung were long and narrow, whereas the transverse segment of the common pulmonary vein was short. The long descending vein was slightly constricted at its junction with portal vein, and then opened into an intrahepatic ampulla formed by the confluence of the ductus venosus, umbilical vein, and portal vein draining the anomalous pulmonary vein. The ampulla perfused the liver and then drained into the inferior caval vein through the hepatic vein (Fig. 5). Atrial septal defect of secundum type was present with its opening 0.5 cm in diameter. The ductus arteriosus was closed.

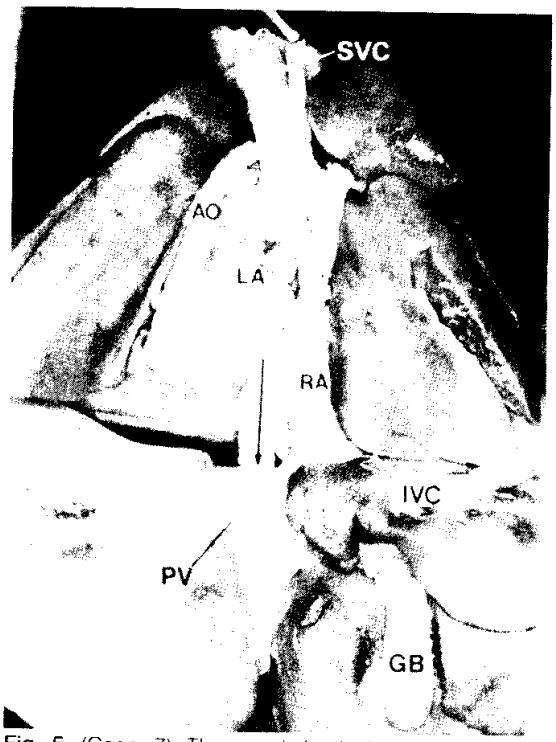

Fig. 5. (Case 7) Thoracoabdominal organs viewed from the back. Right upper pulmonary vein opens into the superior vena cava (SVC), and other pulmonary veins from the right middle and lower lobes and those from the left lung unites to form a very long channel to the portal vein (PV). The long descending vein was connected to the portal vein with mild stenosis (→) and opens into the intrahepatic ampulla. AO = aorta; LA = left atrium; GB = gall bladder; IVC = inferior vena cava; RA = right arium

DISCUSSION

Total anomalous pulmonary venous return is a relatively rare congenital heart disease which may result in death during the first year of life, and the clinical presentation of this lesion can be variable and there is a rare patient who survives until adulthood.

Several authors (Burroughs and Edwards, 1960; Snellen *et al.*, 1968; Bonham-Carter *et al.*, 1969; Gathnam *et al.*, 1970; Jensen *et al.*, 1971; Behrendt *et al.*, 1972; Delisle *et al.*, 1976; Na *et al.*, 1987) have studied and reviewed morphologic variations of this lesion with their large series of autopsied cases. Snellen *et al.* (1968) collected 52 autopsy cases and classified the patterns of abnormal venous drainage encountered so far, while Delisle *et al.* (1976) from Boston reported 93 autopsied cases. Lee *et al.* (1986) reported an autopsy case in Korea of subdiaphragmatic TAPVR in which the pulmonary veins joined the common trunk that was connected to the hepatic vein. We here have performed postmortem examination of all 4 types of autopsy cases of TAPVR (3 supracardiac, 1 cardiac, 2 infracardiac, 1 mixed), and described their morphologic characteristics with special attention given to the presence of obstructions and their locations.

To understand the morphogenesis of TAPVC, a brief review of the normal embryologic development of the pulmonary venous system is necessary. The lung develops as an outpouching from the ventral aspect of the foregut. The splanchnic plexus covering the foregut is the anlage of the venous plexus which covers the lung buds. The common pulmonary vein develops as an out-

pouching from the dorsum of the sinus venosus. The pulmonary venous plexus then develops a communication with this common pulmonary venous channel. The opening of the common pulmonary vein shifts to the left as the interatrial septum develops. The final step in this process is incorporation of the common channel into the left atrial wall so that all 4 tributaries of the pulmonary vein are connected to the left atrium separately.

Lucas *et al.* (1962) cited the mechanism of various anomalies of the pulmonary venous system as follows: Once the junction of the common pulmonary vein and the left atrium has been made, stenosis may occur between these 2 structures with the resulting formation of the cortriatriatum. In rare instances, it may cause enough obstruction to favor persistence of primitive drainage pathways. Snellen *et al.* (1968) offered another explanation on the pathogenesis of anomalies of pulmonary venous drainage: A abnormal shift of the original common pulmonary vein with regard to the atrial septum may lead to abnormal connections into the right atrium or the adjacent, embryologically related structures. Anyhow, once communication between the left atrium and common pulmonary vein is lost, systemic communications to the systemic veins such as the cardinal and umbilicovitelline systems remain persistent. And 1 or several of these collaterals enlarge (de Leval *et al.*, 1973) and provide a total anomalous pulmonary venous connection to the systemic venous system. Persisting segments of the cardinal veins eventually form the superior vena cava,the innominate vein, the coronary sinus, and the azygos vein. The umbilicovitelline system forms the inferior vena cava, the portal vein (Woodwark *et al.*, 1963), and the ductus venosus. Our cases showed a variety of draining passages of anomalous pulmonary vein: 3 supracardiac types draining into the left innominate vein via vertical vein, 1 cardiac type opening into the coronary sinus, 2 infracardiac types draining into the portal vein, and the common hepatic vein respectively, and 1 mixed type draining into the superior vena cava and the portal vein.

Burroughs and Edwards (1960) from the Mayo clinic analyzed 188 patients with total anomalous pulmonary venous connection, stating that 2 anatomic types of junction were present: 1 in which the pulmonary vein converged to form a single trunk before connecting anomalously and the other in which 2 or more pulmonary veins connected to the single structure receiving the veins. When the site of connection was more peripheral than the right atrium or the superior vena cava,the connection was always through a single trunk, and among the cases with connection to the right atrium, the superior vena cava, or the coronary sinus, either a single trunk or multiple pulmonary venous connections were found. Our Case 7 showed findings compatible with Burroughs' observation: The right upper pulmonary vein drained into the superior vena cava, and other pulmonary veins united to form a single trunk with draining into the portal vein, but in Case 4, all the pulmonary veins united to form a single trunk and drained into the right atrium via coronary sinus.

The importance of the size of the atrial communication, which was studied by Burchell (1956), is also noted in our series. The size of the atrial communication was from probe patency to 1.2 cm in diameter. In Case 5, small atrial communication was thought to be one of the major obstructing sites.

Delisle *et al.* (1976) had described various types obstruction of the pulmonary venous pathways in their 93 autopsy cases. The incidence of obstruction was remarkably high in the supracardiac type of uncomplicated TAPVC (14/28, 50%), where the commoner type of obstruction occurred when the left vertical vein passed behind the left pulmonary artery. Our Case 2 showed similar patterns of obstruction in which the vertical vein was interposed between the left pulmonary artery and the left main bronchus resulting in a hindrance to its flow. The less common sites of obstruction in the supracardiac type of TAPVC were the junction of the vertical vein and the innominate vein, the junction of the connecting vein with the right superior vena cava, the common pulmonary vein, both of intrapulmonary and extrapulmonary position, interposition of the common pulmonary vein by the carina and right bronchus posteriorly and right pulmonary artery anteriorly, and extreme hypoplasia of the common pulmonary vein between the right lower pulmonary vein and azygos vein. In our Case 1, ste-

nosis had occurred at the proximal portion of the left upper pulmonary vein between the common channel and the vertical vein where the vertical vein originated from the middle of the left upper pulmonary vein. Delisle *et al.* (1976) did not observe obstructions in the cardiac type of TAPVR as in our Case 4. In our Case 6, an infracardiac type, stenosis was present at the junction of the descending vein and the common hepatic vein.

The abnormal venous pathways in infracardiac TAPVR have been described (Woodwark *et al.*, 1963; Duff *et al.*, 1977) as commonly showing the following figures: The superior and inferior pulmonary veins on each side unite to form a common pulmonary vein, which descends to the midline as the descending vein and passes through the diaphragm. Our 2 cases (Case 5, Case 6) of infracardiac type also showed such general configurations. Our findings that deserve to be mentioned are: 1) The lengths of the individual pulmonary veins were relatively longer than those of the common pulmonary venous segment producing a tree-shaped configuration in contrast to supracardiac type; 2) The vertical veins may descend from anywhere in the common pulmonary veins, and deviate either to the right or to the left. In our Case 7, the vertical vein deviated to the right producing the left pulmonary veins longer than the right.

It is apparent that surgical treatment offers the only chance of improvement because medically treated patients with TAPVC represent a very high and early mortality (Delisle *et al.*, 1976). Since the first operation for TAPVR was done by Muller (1951), Cooley and Ochsner reported successful correction in a 6-month-old infant with the aid of a cardiopulmonary bypass. Cooley and colleagues described the principles of surgical repair: 1) the use of the pump oxygenator, 2) creation of the largest possible anastomosis between the common venous trunk and the left atrium, 3) closure of the atrial septal defect, and 4) ligation of the persistent left anterior cardinal vein,the connection to the right superior vena cava, or the connection with the portal system. Determinants of operative mortality have been stated to be age (Turley *et al.*, 1980), anatomical type of TAPVC, preoperative conditions, preoperative evaluation of pulmonary vascular resistance (Hastereiter *et al.*, 1962; Newfeld *et al.*, 1980), pulmonary venous obstruction (Haworth and Reid, 1977), small left atrial size, decreased left ventricular volume and function, and operative technique. Hawkins *et al.* (1983) stated that the most important factor in assuring long-term survival is the creation of a large,unobstructed anastomosis at the time of operation that will enlarge with the growth of the patient. Autopsy findings of our Case 1 showed narrow anastomotic site resulting in ventilatory weaning failure complicated by disseminated intravascular coagulation. Surgical treatment of TAPVC is now possible with good early and late results. Early correct diagnosis, preoperative aggressive intensive care, and prompt operative correction (Mazzucco *et al.*, 1983; Sano *et al.*, 1989) are the most important factors that provide optinum outcome (Clarke *et al.*, 1977).

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＝국 문 초 록＝

총폐정맥 환류 이상증
－7례 부검 분석－

서울대학교 의과대학 흉부외과학교실

이정렬 · 김용진 · 노준량 · 서정욱
이홍재 · 최지영 · 윤용수 · 서경필

총폐정맥 환류 이상증은 폐정맥과 좌심방 사이에 직접 연결이 없이 모든 폐정맥의 환류가 직접 또는 간접적으로 우심방으로 통하는 선천성 심장 질환으로 폐정맥 폐쇄가 심할수록 조기에 증상이 발현되며 영유아기의 사망율이 높은 질환이다.

저자 등은 7례의 총폐정맥 이상 환류증에 대한 부검을 실시하였으며,이들의 연령 분포는 26일에서 5개월이었고 남 : 녀비는 4 : 3이었다. 3례는 상부 심장형으로서, 1례(case1)는 좌상 폐정맥이 총폐정맥으로 환류되는 부위의 협착이 있어 수직정맥 결찰후 환류의 심한 장애를 초래하고 있었으며 1례(case2)는 수직 정맥이 좌기관지와 폐동맥 사이에 끼어서 환류 장애를 보였고 나머지 1례(case3)에는 협착의 소견이 관찰되지 않았다. 1례(case4)는 총폐정맥이 관상 정맥동으로 환류되는 심장형이었다. 2례(case5,case6)의 하부 심장형은 총폐정맥이 모두 수직 정맥을 통하여 간정맥으로 환류하였으며 간정맥으로 개구하는 부위에 심한 협착이 존재하였다. 1례(case7)는 우상 폐정맥이 상공정맥으로 환류되고 나머지 폐정맥들이 총폐정맥으로 합류하여 수직 정맥을 통해 간문맥으로 환류되는 혼합형이었으며 이 경우도 역시 간문맥으로 환류되는 부위의 심한 협착이 있었다.

이상 7례의 총폐정맥 이상 환류증에 대한 부검 분석을 통하여 폐정맥 환류의 협착 또는 폐쇄가 심한 환아일수록 증상이 일찍 발현된다는 사실을 관찰하였으며 특히 술후 원인이 불분명한 폐울혈의 원인이, 발견되지 않은 폐정맥 환류로의 협착인 경우가 많다는 사실을 부검을 통하여 입증하였다. 따라서 총폐정맥 환류이상증은 증상이 일찍 나타나는 환아일수록 조기의 정확한 진단과 중환자 관리, 그리고 조기에 완전 교정술을 시행하는 것이 무엇보다 중요하겠다.