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Double Outlet Right Ventricle, Taussig-Bing Type

— An autopsy case —

Jeong Wook Seo, M.D., **Je G. Chi**, M.D., **In Aee Park**, M.D., **Sang Kook Lee**, M.D.
Yong Soo Yoon*, M.D. and **Sei Woen Yang***

*Departments of Pathology and Pediatrics**, College of Medicine, Seoul National University, Seoul, Korea*

INTRODUCTION

Double outlet right ventricle (DORV) identifies a diverse group of rare cardiac malformations that share the common features that both the aorta and the pulmonary artery arise primarily from the morphologic right ventricle.[1] According to the relationship between the great vessels and ventricular septal defect, five types of morphologic variations are defined.

Taussig-Bing malformation[2] is known as a variant of DORV and shows subpulmonic ventricular septal defect. Clinical presentation of this is similar to that of transposition of great vessels, because most of the left ventricular output is directed to the pulmonary artery whereas the blood from the right ventricle flows through the aorta. In fact Taussig and Bing first described their case as "complete transposition of the aorta and a levoposition of the pulmonary artery".

We have studied a case of complex congenital malformation that could best be classified into Taussig-Bing variant of DORV.

REPORT OF CASE

J.S. Kim, a 1 year and 3 month old male baby, was admitted to Pediatric Service of the Seoul National University Hospital, due to cyanosis and growth retardation. He was the first baby and was a product of full term gestation without immediate pre- and post-natal problems. The birth weight was 3.4 kg. The mother had a brief period of unconsciousness due to family problem during the early pregnancy for which she was prescribed sedatives. Family history was not contributory and his younger sister was healthy. Cyanosis and poor sucking were noted even since he was born, for which he was brought to several hospitals. Congenital heart disease of undetermined nature was suspected, and the patient was transfered to the Seoul National University Hospital.

At the time of admission he was cyanotic and emaciated. Vital signs were; blood pressure 110/80 mmHg (left leg); pulse rate, 125/min; respiration rate, 55/min. The body weight was 5.5kg (less than 3 percentile), and the height was 66.5cm (less than 3 percentile). The head circumference was 47.5cm (75 percentile). Widened suture lines and tense fontanelles were present. Anterior chest showed bulging at the precordial area. Intercostal retraction was prominent. Thrill was palpable. Auscultation of the heart exhibited accentuation of P2 and systolic murmur at the left sternal border. The abdomen was scaphoid but the liver was palpable 1-2 finger breadths. Clubbing of fingers was present.

The hemoglobin was 21.4%, the hematocrit 65% and WBC 4700/mm³. Chest roentgenogram (Fig. 1) showed a mild cardiomegaly and increased pulmonary vascularity. Electrocardiogram (Fig. 2) showed right ventricular hypertrophy, right axis deviation and P-pulmonale. Computed tomography of head (Fig. 3) showed enlargement of head, dilatation of lateral and third ventricles, suggestive of the obstruction at the level of the aqueduct.

He was carefully digitalized and diuretics were given to combat congestive failure. The ventriculo-peritoneal shunt was performed to relieve the intracranial pressure on the 12th hospital day, but the operation wound was infected and was followed by repeated bouts of pneumonia. The patient expired despite the antibiotics and general

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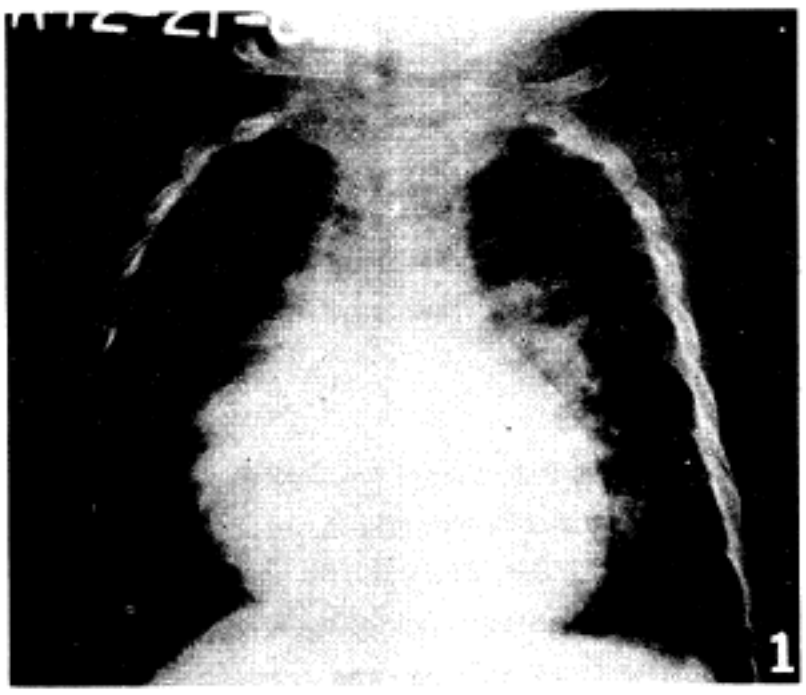

Fig. 1. Chest roentgenogram.

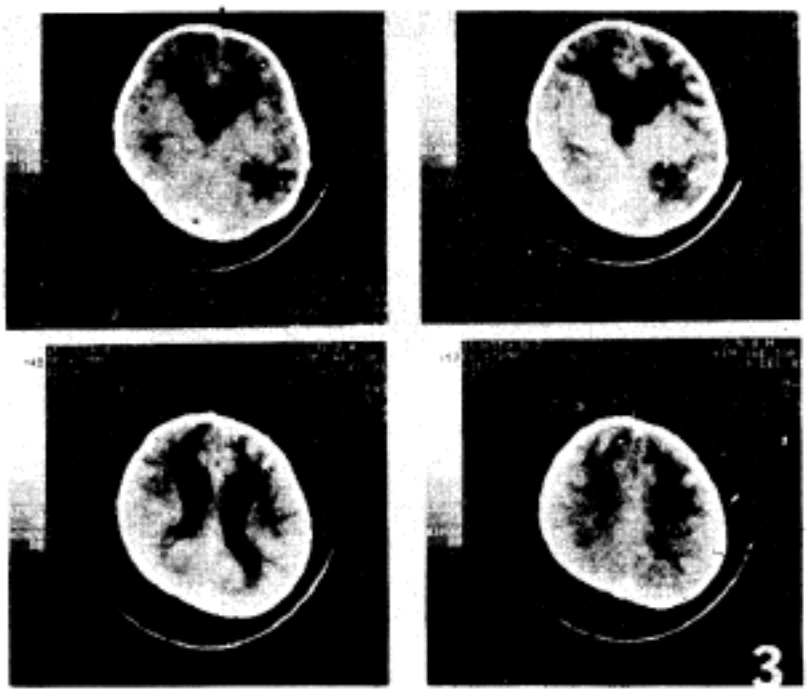

Fig. 3. Computed tomography of brain.

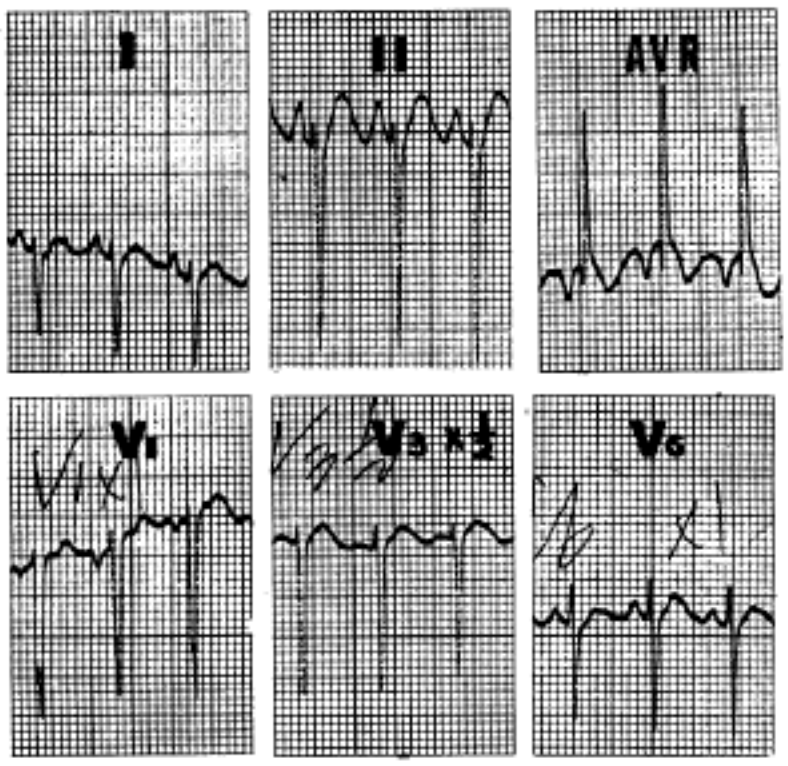

Fig. 2. Electrocardiogram.

Fig. 4. Anterior view of the great vessels in situ: The aorta arises right and slightly anterior to the pulmonary artery.

supportive measures at the age of 20 months.

AUTOPSY FINDINGS (A82-40)

The body was poorly developed and poorly nourished. Ventriculoperitoneal shunt device was implanted at the right parietal area, via subcutis to peritoneal cavity. The wound was infected with active purulent discharge.

The heart weighed 119gm. The right atrium was mildly enlarged. The superior and the inferior venae cavae opened into the right atrium in normal fashion. The interatrial septum was well developed, and foramen ovale was closed. The tricuspid valve was normal. The right ventricle was hypertrophied; its wall measured 1.0cm in

thickness. The pulmonary artery arose in its normal position. The aorta was transposed and arose from the right ventricle, right to the pulmonary artery, anterior to the anterior cusp of tricuspid valve. Subpulmonic ventricular septal defect was present. The defect was located just below the pulmonic valve and measured 0.9×0.7cm. The defect was marginated by upper free border of fused dorsal and ventral interventricular septa inferiorly and by the left cusp of pulmonic valve superiorly.

The aorta and pulmonary artery measured 1.2cm and 1.7cm in diameters, and they were separated by a thick musclar band (crista supraventricularis). The aorta and its brachiocephalic branches were normal. The ductus arteriosus was anatomically closed. Pulmonary arteries were connected to the normal lungs. Pulmonary veins drained to the left atrium in normal manner. The left ventricle was hypertrophied, 0.9cm in thickness. The left atrium and mitral valve were normal. The cardiac skeleton was carefully oriented. The tricuspid valve ring was larger than mitral valve and located right and anterior to the mitral valve. The anterior leaflet of mitral valve was in fibrous continuity with the left cusp of pulmonary valve through the ventricular septal defect. The aortic valve ring was smaller than the pulmonic ring and was right and slightly anterior to the pulmonic ring on the same plane. The aortic valve ring was separated from both mitral and pulmonic valves by thick muscular wall. The left coronary artery ran anteriorly to the pulmonic valve ring. The right coronary artery and coronary sinus were in normal position.

The brain weighed 790 gm. Cut sections of the brain showed marked, symmetrical dilatation of lateral and third ventricles. Fourth ventricle was slightly dilated and cerebral aqueduct was patent. Ventricular surface was irregular and colored dusky brown. Surrounding white matter was also dusky white. Microscopically ventricular surface lost its lining ependymal cells and astrocytes were activated and hypertrophic. Focal infarct was present at

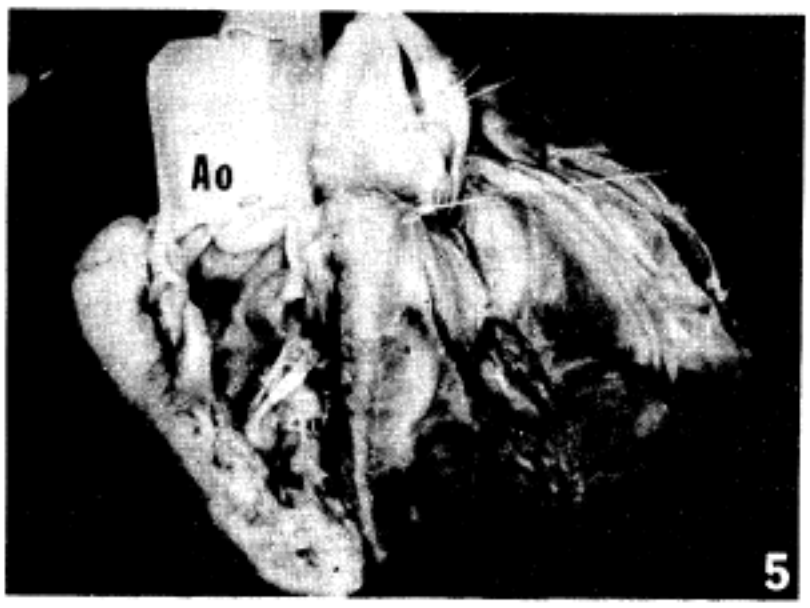

Fig. 5. The aorta arises from the right ventricle and it has subaortic muscular outflow tract, i.e. conus.

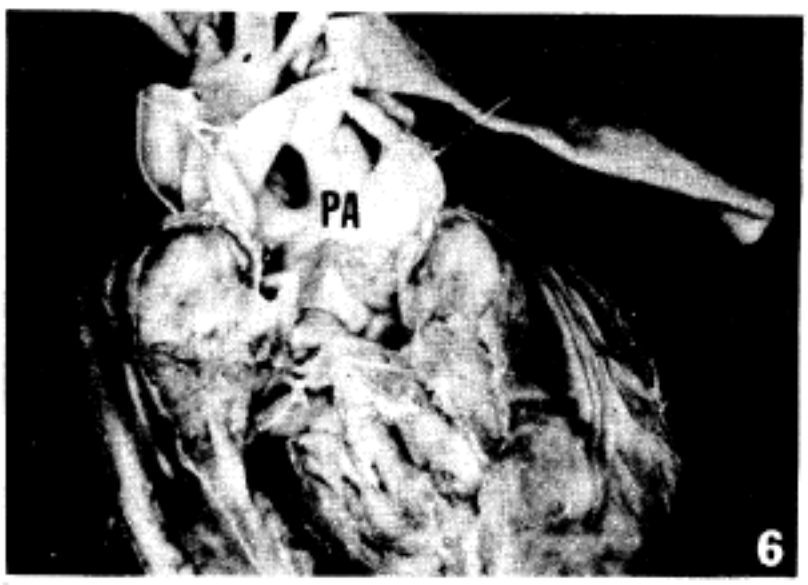

Fig. 6. The pulmonary artery arises in normal fashion. There is a subpulmonic ventricular septal defect, through which pulmonic valve and mitral valve are in fibrous continuity.

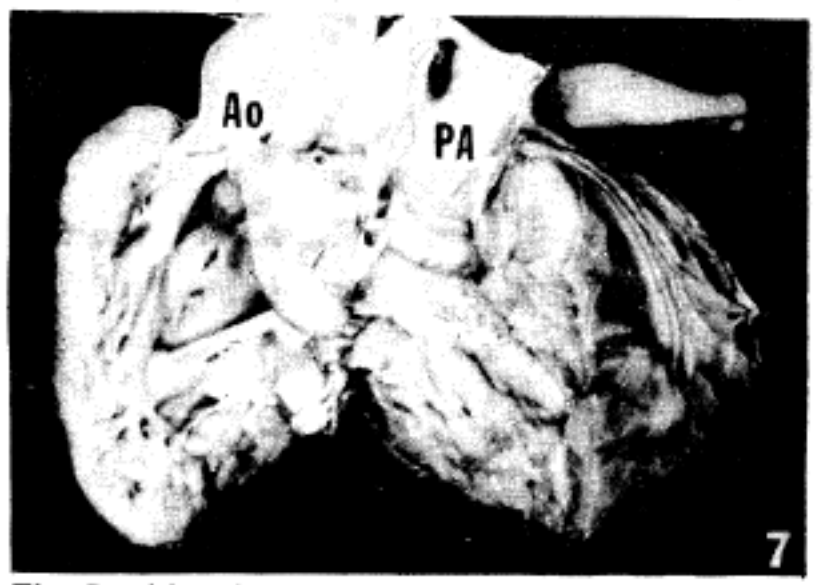

Fig. 7. After the removal of portion of right ventricular anterior wall, muscular ridge separating the aorta and the pulmonary artery is clearly visualized (bilateral conus).

lower margin of right caudate nucleus. Lung was multifocally consolidated and sections of the lung showed diffuse interstitial infiltration of acute and chronic inflammatory cells. An aberrant renal artery and an accessory spleen were also noticed.

DISCUSSION

This patient was a case with complicated anomalies of heart and brain. The brain anomaly was a communicating hydrocephalus and the brain was infected after the surgical intervention for decompression of increased intracranial pressure. The pyogenic infection was direct cause of death. The cardiac anomaly is worthy of detail discussion.

Clinical presentation of this case is cyanotic congenital heart disease with increased pulmonary vascularity, thus mimicking the transposition of great vessels. Double outlet right ventricle with subpulmonic ventricular septal defect is the next probable condition in this clinical setting. Differentiation of the two conditions is usually made by the angiocardiographic findings. However, clinical diagnosis of DORV based on angiocardiography is sometimes not correct. Because the varieties of cardiac anomalies are biologic spectrum and the secondary adaptative morphologic changes can mask the primary defect,[3] clinical diagnosis without autopsy cannot be confirmative in every case.

The description "double outlet right ventricle" was first appiied to a group of rare partial transposition complex in which both great arteries arise completely from the right ventricle.[4] Basic defect of this anomaly was considered to be unequal division of the truncus by the trunco-conal septum. Therefore either aortic or pulmonary hypoplasia was considered to be present in almost all cases. Based on that concept, DORV could be subdivided into 3 types, 1) hearts with true truncus arteriosus arising wholly from the right ventricle, 2) the "Eisenmenger type" with aortic hypoplasia and 3) the "Fallot type" with pulmonary hypoplasia, stenosis and atresia. The major differential diagnoses of DORV are Taussig-Bing malformation, tetralogy of Fallot and complete transposition of great arteries. In Taussig-Bing heart, there is neither aortic hypoplasia nor pulmonic hypoplasia.[2,5] Extreme overriding of the aorta in cases of tetralogy might be included in the spectrum of DORV, and such examples were really seen.[4,6] Meanwhile in complete transposition, there may be ventricular septal defect with overriding transposed pulmonary artery.

Taussig-Bing malformation was first described in 1949.[2] The malformation consisted of a transposed aorta, a large pulmonary artery which arised primarily from the right ventricle and partly overrode the ventricular septum. A high ventricular septal defect and the right ventricular hypertrophy were associated. In these regards our case possesses every feature to be Taussig-Bing malformation. Witham[4] considered Taussig-Bing heart as a variant of double outlet right ventricle. Although he didnot describe the type to which Taussig-Bing heart belonged, it might have been the Eisenmenger type. But the aorta Taussig-Bing heart was not hypoplastic but was only smaller than pulmonary artery.

One of the 4 components in tetralogy of Fallot is the overriding of aorta and the extreme degree in overriding may be considered the Fallot type of DORV. But as Witham himself quoted, Selzer had pointed out that the spiral course of the interventricular septum causes the aorta to "override" secondarily[3]. Great dilatation of the right ventricle can displace the septum to the left, resulting in acquired aortic transposition. Van Praagh[5] emphasized the fibrous discontinuity between the mitral valve ring and semilunar valve rings to be a Taussig-Bing heart or double outlet right ventricle. Van Praagh emphasized that the Taussig-Bing heart is a true double outlet right ventricle and confirmed that the heart had mitral-pulmonic fibrous discontinuity by interposition of subpulmonary conal free wall musculature[5]. Lev[6] proposed less strict concept saying that double outlet right ventricle is the condition in which both arterial trunks emerge almost completely or completely from the right ventricle and there may or may not be mitral-aortic or mitral-pulmonic continuity. Lev's classification of DORV is most widely used and gives clinical significance. His classification is based on the committment of ventricular septal defect. The concept is simple and physiologically correct, but some cases of tetralogy of Fallot could be confused with DORV. Anderson's description[7] is that of another difference. His view of DORV permits tricuspid-aortic fibrous continuity but not mitral-pulmonic continuity. The latter form was intermediate form between

transposition and double outlet right ventricle. It is such a difficult task to diagnose DORV correctly even with autopsy material. Therefore one should be very careful to diagnose DORV and should confirm all the necessary features of DORV with specific mention on atrioventricular-semilunar continuity in each case.

It is our feeling that DORV could best be defined as a heart in which one great artery and more than half of the other great vessel arise from the morphologic right ventricle and there is mitral-aortic fibrous discontinuity regardless of the presence of tricuspid-aortic or mitral-pulmonic fibrous continuity.

Thus the presented case should be put into category of the double outlet right ventricle. However, Van Praagh might put this into the complete transposition and extreme overriding of pulmonary artery. And Anderson might categorize this into his intermediate form.

The ventricular septal defect in this case is partly due to defect in the fusion of conal and interventricular septa. The other component of this ventricular septal defect is the resorption of bulbo-atrio-ventricular ledge. So the defect is primary interventricular foramen of embryonic heart and normal outflow tract of the left ventricle. Anterior location of the left coronary artery is another important anomaly which might have been an obstacle if this patient was operated for the surgical correction. In most patients with DORV, the origins of coronary arteries are normal,[8,10] but several cases with abnormal coronary artery crossing the right ventricular outflow tract are described.[9,10] Like the other human birth defects, the cause and mechanism of malformation in DORV are not determined. DORV is cotegorized into bulboventricular malformation which includes tetralogy of Fallot and various transposition complexes. Conal development hypothesis and straight septum hypothesis are major pathogenetic explanations of bulboventricular malformation.[7] Experimental production of DORV was successful by several authors.[11,12] Since the introduction of cardiac catheterization procedure DORV has been diagnosed not infrequently. Actual incidence in Korea is unknown but the relative incidence is reported as 0.8%.[13] Among the 1,701 cases of open heart surgery of the Seoul National University Hospital (1959-1981), 17 cases were DORV and 12 cases (70.6%) died during operation.[14] Seo[15] et al reported 18 cases of DORV (1975-1981) diagnosed by clinical findings including cardiac catheterization and angiography. Among the 18 cases, subpulmonic ventricular septal defect (VSD) type, subaortic VSD type and uncommitted VSD type were 6, 10 and 2 cases, respectively. All of the 12 cases of subaortic VSD and uncommitted VSD types were associated with pulmonary stenosis. There is also an autopsy case report of DORV in a stillborn infant who showed subaortic ventricular septal defect.[16]

SUMMARY AND CONCLUSION

An autopsy case of cardiac malformation associated with hydrocephalus is presented. He was cyanotic. Chest roentgenogram showed cardiomegaly with increased pulmonary vascularity. Electrocardiogram showed right ventricular hypertrophy, right axis deviation and P-pulmonale. At autopsy the heart weighed 119gm. The aorta arose right and anterior to the pulmonary artery and both great vessels were connected to the right ventricle. The aorta had subaortic conus. There was subpulmonic ventricular septal defect through which pulmonic-mitral fibrous continuity was present.

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=국문초록=

이중 유출로 우심실 (Taussig－Bing 형)

서울대학교 의과대학 병리학 및 소아과학교실*

서정욱·지제근·박인애·이상국·윤용수*·양세원*

이중 유출로 우심실은 1957년 Witham에 의해 명명된 기형으로 대동맥과 폐동맥이 모두 우심실에서 나가는 심장기형군이다. 이 기형은 대동맥, 폐동맥및 심실중격결손의 상호관계에서 여러가지 변형을 보이기 때문에 그에 따라 임상소견도 다양하고 유사 심장기형과의 감별이 어려우며 그 정의 자체도 학자에 따라 이견을 보이고 있다. 저자들은 생후 20개월만에 사망한 남자아기에서 Taussig-Bing형으로 생각되는 이중유출로 우심실을 부검하여 보고한다. 정상분만으로 태어난 이 아기는 출생시부터 청색증과 호흡곤란이 있었다. 머리가 점점커져 수두증을 생각하고 뇌압을 낮추기 위한 뇌실－복강 연결관을 시술하였으나 수술에 의한 합병증으로 사망하였다. 흉부 방사선 촬영상 심장종대와 폐혈관 증가가 있었고 심전도상 우심방및 우심실 비대가 있었다.

부검시 심장은 커져 있었다(119gm). 특히 우심실 비후가 현저하였다. 대동맥은 폐동맥의 우측 전방에 위치하였고 대동맥하 conus가 있어 삼첨판 및 승모판과의 연결이 없었다. 폐동맥은 정상위치에 있었으나 폐동맥하 심실중격결손이 있었고 그 결손을 통하여 폐동맥 판막과 승모판이 섬유성으로 연결되어 있었다. 좌측 관상동맥이 폐동맥 전방으로 주행하였다. 뇌의 부검소견은 연결형 수두증으로 양측 측뇌실과 제3뇌실의 심한 확장이 있었고 화농성 염증이 심하였다.

본례의 심장은 대동맥하 conus는 발달되어 있으나 폐동맥 판막과 승모판이 섬유성 연결을 보이기 때문에 이런 기형을 이중 유출로 우심실에 넣기도 하지만 학자에 따라서는 대동맥 전위증 혹은 그들의 중간형으로 분류하기도 한다.

저자들은 이중 유출로 우심실에서 대동맥과 승모판의 연결은 없어야 하지만 대동맥과 삼첨판 혹은 폐동맥과 승모판의 연결은 있어도 좋다고 생각하였고 아울러 이러한 형태학적 확인이 없이는 진단하기 어려운 기형이라고 생각하였다.