J. Tokyo Med. Coll., **49**(6) : 911～917, 1991

Case Reports

Case of An Adult Type IIb Tricuspid Atresia

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Abstract

Tricuspid atresia is a relatively rare condition among congenital heart anomalies. In particular, documented adult cases are extremely few. We hereby report on a 21-years-old woman with tricuspid atresia (type IIb) classified by the Edwards-Burchell criteria. The patient seemed to owe her long-term survival to the maintenance of appropriate pulmonary blood flow made possible by stenosis of the pulmonary valve.

Tricuspid atresia has been found to exist in 1 to 5% (average 3%), of congenital heart diseases observed postmortem. Documented cases living who reach adult hood are rare. It is said that 90% of the patients die before reaching 10 years of age. We hereby report a patient with tricuspid atresia (type IIb) according to the classification of Edwards & Burchell.

Patient

Case : Female 21 years of age

Chief Complaints : Cough and cyanosis

Past illnesses : Pneumonia at age 17

Present Ailment : At the age of 3, heart murmur was pointed out. Because of the absence of symptoms, she was not treated for the ailment. As a schoolgirl, the patient had no symptoms in everyday life, but she did experience palpitation while running around the school ground, and consequently refrained from taking part in athletic activities thereafter. At the age of 17, she caught pneumonia and heart murmur was pointed out for the second time and a detailed examination was recommended. However, again nothing was done. Upon reaching the age of 21, she began to suffer from cough and dyspnea and was admitted to a hospital for detailed examinations.

Received June 20, 1991. Accepted Sept. 4, 1991

Key words : Tricuspid asresia (三尖弁閉鎖症), Adult case (成人例), Edwards-Burchell classification (エドワード・バーセル分類)

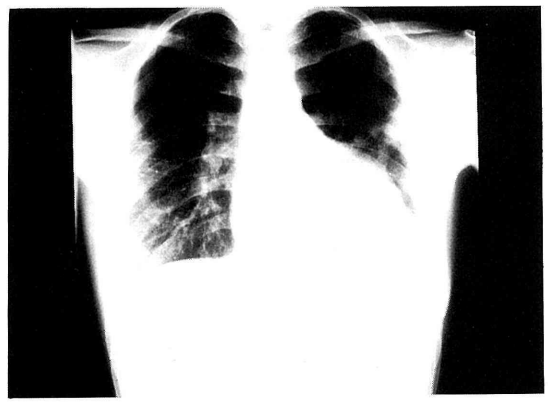

Fig. 1 Chest roentgenogram.

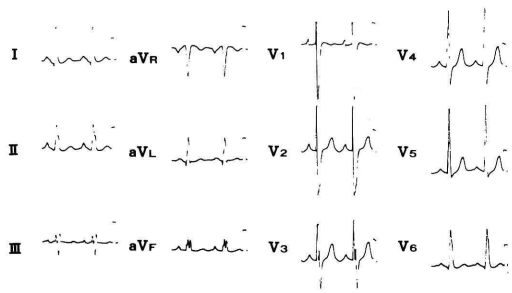

Fig. 2 Left ventricular volume load accompanied by tight atrial enlargement.

Findings on admission : Stature average, nutritional condition good, blood pressure 96/68 mmHg, pulse 96/min regular. Lips and nail beds cyanotic, clubbed fingers.

Ausculation : Fine bubbling rale over the left upper lung field, lst sound pure and 2nd sound single, Levine 4/6 pansystolic murmur was heard at left sternal border in 3rd intercostal space. Abdomen flat and soft. Liver and spleen not palpable. Jugular vein engorged. Lower extremity edema not detected. No neurological abnormalities found.

Radiologic examination : The chest X-ray finding after disappearance of respiratory symptoms are shown in Fig.1. The cardiothoracic ratio was 57%, the left 2nd, 3rd and 4th arches were protruding and the heart shadow was spherical. Pulmonary vascularity is normal. Infiltrative shadow was recognized in the left middle lung field.

ECG : The sinus rhythm was regular, and the QRS axis was +15 degrees (Fig. 2). The left ventricular volume load was accompanied by right atrial enlargement and broad QRS. The P wave was spiky in the precordial lead and biphasic in lead III.

Two dimensional echocardiogram : In the four chamber view (Fig. 3-a), a cord-shaped echo as the posterior left ventricular wall was detected where the tricuspid valve is normally located, and the right ventricular cavity was invisible. The Septum was also not detected in the ventricular cavity on other sagittal sections. Tere was a large atrial septal defect and slight prolapse the mitral valve. On the long axis view (Fig. 3-b) the interventricular septum was absent, and two great vessels continuous with the ventricular cavity were detected. On the short axis view (Fig. 3-c), the interventricular septum was also absent, and D-type malposition of the great vessels was seen, in which aorta was located to the right and

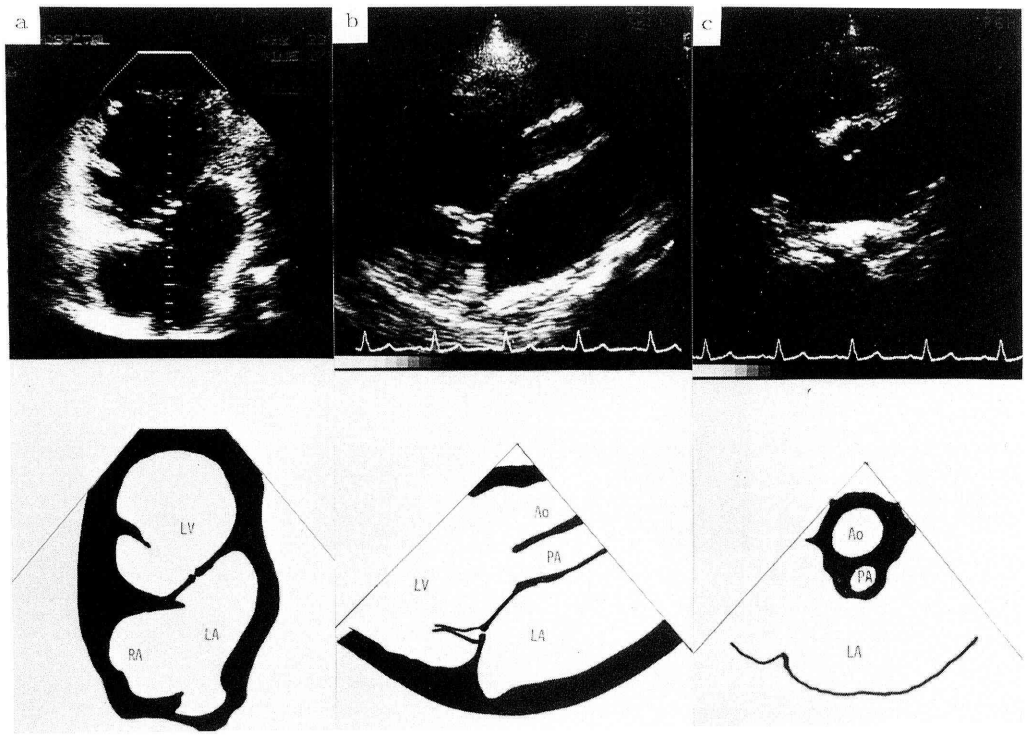

Fig. 3 The four chamber view (3 - a), revealed both tricuspid atresia and large atrioventricular defect.
In the parasternal long axis view (3-b), two great vessels continuous with venrticular cavity were detected. In the short axis view (3-c), the aorta was located to the right and anteriorly to the pulmonary artery.

anteriorly to the pulmonary artery. The ventricle had fine trabeculation and two large papillary muscles.

Cardiac catheterization : Angiocardiogram-The catheter inserted into the right femoral vein veered to the right of spine and passed through the inferior vena cava to the right atrial resion, left atrial region, left ventricle and then to the pulmonary artery. The investigation of the right ventricular cavity via the catheter failed to identify any site with right ventricular pressure curve. In the contrast study of right atrial region, the contrast medium flowed from the right atrial region to the left atrial region and then to the left ventricle. In left ventriculography (Fig. 4), two great vessels were visible through the ventricle. The anterior vessel was the aorta, and the posterior vessel was the pulmonary artery. From the above findings, the position of atrium was judged to be normal, and D-type malposition of the great vessels was diagnosed. Only one ventricle was confirmed to be present, which was identified as left ventricle from the structure of the trabeculation. No cavity considered to be the right ventricle was detected by the contrast study.

Pressure and oxygen saturation : Oxygen saturation was 85% for the left ventricle, 85% for aorta and 84% for pulmonary artery, with no difference among these structures (Table 1). The mean oxygen saturation for the right atrial region was 69%. On the other hand, difference in oxygen was detected botween pulmonary vein and left atrial region, with the value 98% for the former and 83% for the latter. In the contrast study of main pulmonary artery from inmediately below the bifurcation (Fig. 5), the

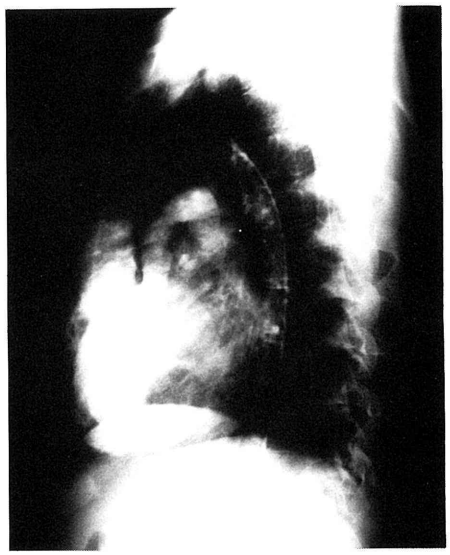

Fig. 4 Left ventriculogram. The anterior vessel was the aorta, and posterior vessel was the pulmonary artery.

Table 1 Cardiac catheterization data

	Pressures mmHg	O_2 Saturation %	O_2 content Vol %
SVC	11/8(7)	62	12.55
IVC	15/12(10)	66	13.35
RA (H)	A-17(11)	74	14.97
(M)	V-12	68	13.76
(L)		68	1315
PA (M)	16/8(12)	84	17.20
PV	11/9(7)	98	19.70
LA	11/12(9)	83	16.79
LV	98/11	85	17.20
Ao	100/62(76)	85	17.20

SVC. superior vena cava IVC. inferior vena cava RA. right atrium H. high M. middle L. low PA. pulmonary artery PV. pulmonary vein LV. left ventricle Ao. aorta

pulmonary valve showed a dome formation, suggesting valvular stenosis, and the portion above the bifurcation was dilated.

Contrast cardiac CT (Fig. 6) : D-type malposition of the great vessels in which aorta was located to the right and anteriorly to pulmonary artery was detected. Pulmonary arteries above the bifurcation were dilated.

Discussion

Tricuspid atresia is a relatively rare condition among congenital heart anomalies and in particular documented adult cases are extremely few[3]. In the present case, the problem was whether to diagnose the condition as tricuspid atresia or as atresia of the right atrioventricular orifice combined with single

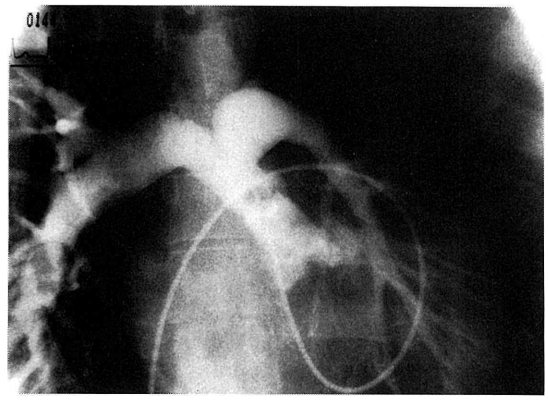

Fig. 5 The pulmonary valve showed a dome formation. The pulmonary artery showed poststenotic dilation.

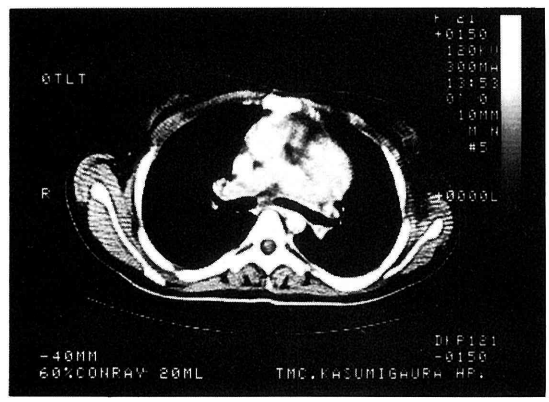

Fig. 6 D-type malposition of great vessels in which the aorta is located to the right and anteriorly to the pulmonary artery.

ventricle. The Edward & Burchell classification of tricuspid atresia specifies that the right ventricle or a residual cavity of right ventricle is present except in Type-Ia. Nonetheless, it is admitted that the interventricular septum is sometimes completely lost in cases with D-malposition, and the ventricle in such a case resembles single ventricle in form. According to Van Praagh et al., the "single ventricle" receives both the mitral and tricuspid valves or a common atrioventricular valve. It includes both cases with rudimentary outlet chamber and those without it. This difinition excludes tricuspid and mitral atresia. The present case dose not fall under the "single ventricle" as defined by Van Praagh et al., since a mitral valve was present and muscular closure was detected at the site of the tricuspid valve. Qrero et al.[4] reported on atresia of one atrioventricular orifice combined with a single ventricle. In the present study, significance is attached to the form of the ventricle beneath the valve : the homologous ventricle in atresia of the atrioventricular valve of the left atrial type is the left ventricle if the form is normal, and the right ventricle if it is L-loop type. Atresia of one atrioventricular valve combined with hypoplasia of the homologous ventricle is common in ordinary mitral or tricuspid atresia, so that such a condition excludes the case from the category "single ventricle". In the present case, the ventricle wall was slightly coarse but had two large papillary muscles, and it resembled the left ventricle in muscle structure ; the

atrioventricular valve was considered to be the mitral valve ; as for muscular closure, the ventricular cavity was absent beneath the atrium and a small depression was sometimes visible at the site of tricuspid valve. These features do not agree with the concept of single ventricle purposed by Quero, et al. From their viewpoint, this was tricuspid atresia. Cases of "single ventricle" combined with atresia of one atrioventricular valve or malposition of great vessels are now frequently reported, many of which do not fall under any category under the known systems of classification. Anderson, et al.[5] proposed a classification in which the heart having a "single ventricle" with an inlet and lacking the inlet septum is defined as a univentricle heart, and classified many cases of tricuspid atresia into this category.

According to the definition by Anderson, et al., the present case falls under the category of univentricle heart, with (1) atrial situs solitus (2) primitive or univentricle heart of left ventricular type without outlet chamber (3) absent right atrioventricular connection (4) D-transposition of great vessels (5) pulmonary stenosis. This diagnostic system is systematic and lucid.

In Japan, 6 cases of tricuspid atresia in adults have been documented[6~9] to the best of our knowledge. According to Yoshida, et al.[8] who researched the international literature, survival at ages over 20 years without operation has been confirmed in 18 cases, which account for 1.6% of the total known tricuspid atresia population. This shows that long-term survival is also rare in other countries. It has been reported that prognosis is relatively good in Type IIb, and that prognosis depends on blood flow in the pulmonary artery. The present patient seemed to owe her long-term servival to the maintenance of appropriate pulmonary blood flow made possible by the stenosis of pulmonary valve.

Conclusion

A 21-years-old woman with tricuspid atresia (Type IIb according to the Edwards-Burchell classification) is described. Her condition could be diagnosed either as "single ventricle" or tricuspid atresia. In such a case, the systematic method of representation proposed by Anederson et al seemed useful to help understand the condition.

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三尖弁閉鎖症（IIb）の成人例

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三尖弁閉鎖症は先天性心疾患剖検例において1～5%，平均3%と比較的希な心疾患であり、10歳までにその90%までが死亡すると言われ，成人までの生存例の報告は少ない．著者らは，21歳女性の三尖弁閉鎖症，Edwards-Burchell 分類における Type IIb を経験した．本例のように三尖弁閉鎖症か，右側房室弁閉鎖に単心室が合併したものか迷う症例では，Anderson 等の系統的に表現する方法が，わかりやすく良いのではないかと思われた。