IDIOPATHIC MYOCARDITIS CHARACTERIZED BY MARKED RIGHT VENTRICULAR DILATATION
REPORT OF TWO AUTOPSY CASES

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We encountered two children with chronic idiopathic myocarditis accompanied by marked right ventricular dilatation, who died of progressive right heart failure. A definitive diagnosis was made by histological examination of the myocardium at autopsy. The patients were both boys, aged 7 years and 1 year and 4 months, and a number of identical features were evident upon physical and laboratory examinations. No heart murmur was heard, and gallop rhythm was noted in distant heart sounds. Electrocardiogram revealed intraventricular block, low voltage QRS complex, and ST-T abnormality. Two-dimensional Doppler echocardiogram and right ventriculogram showed marked dilatation and decreased contractility of the right ventricle as well as tricuspid regurgitation. Thinning of the wall and marked dilatation of the right ventricle were confirmed at autopsy. Our observations showed that chronic myocarditis associated with tricuspid regurgitation may readily lead to marked right ventricular dilatation even exceeding the degree of left ventricular dilatation.

Idiopathic myocarditis associated with such unusual features is relatively rare, and may present problems in differentiation from other congenital heart diseases causing dilatation and dysfunction of the right ventricle.

MARKED right ventricular dilatation and right heart failure in two children were identified by postmortem histological examination of the myocardium as derived from idiopathic myocarditis. This condition is considered to correspond to the dilated type idiopathic myocarditis according to the classification by Okuni et al. but was especially noted for the marked right ventricular dilatation exceeding left ventricular dilatation. Differentiation of this disease from other congenital heart diseases characterized by dilatation and dysfunction of the right ventricle such as Ebstein's disease, or Uhl's anomaly and right ventricular dysplasia (RVD) may pose major clinical problems.

CASE REPORTS

Patient 1
This patient, was the third child born into his family and had a normal delivery after an uneventful gestation. The body weight at birth was 3,450 gm. The first child (female) of the family died of congestive heart failure at the age of six years, but details are unknown. The second child is healthy.

The patient had a fever of 38°C for four days when he was seven years old. A local physician observed hepatomegaly and cardiomegaly in chest roentgenograms. As swelling of the abdo-
men and edema of the eyelids were noted a month later, he was admitted to the Department of Pediatrics, Miyazaki Medical College for further examination.

On admission, the patient was 122 cm tall and weighed 20.5 kg with a pulse rate of 104/min and blood pressure of 90/58 mmHg. Cyanosis was not observed. The eyelids were slightly edematous. The lung fields appeared normal and no dyspnea was present. No heart murmur was heard, but both the first and second heart sounds were faint and presented with gallop rhythm. The liver was hard and was palpated 7 cm below the costal margin. The white blood cell count was 7,300/mm³, CRP (-), GOT 28 IU, GPT 17 IU, LDH 415 IU, and CPK 51 IU, being all normal.

The chest x-ray film (Fig. 1) showed a markedly enlarged right atrium and right ventricle, and the cardiothoracic ratio was 65%. Pulmonary vascular markings were not striking and signs of pulmonary congestion were absent. ECG (Fig. 1) indicated sinus rhythm and showed low voltage P and QRS waves, right bundle branch block, and lowering in limb leads, and inversion in precordial leads, of T wave.

In two-dimensional and M-mode echocardiograms (Fig. 2A, 2B, 2C), the right and left ventricular end-diastolic dimensions were 5 cm and 4 cm, respectively, indicating marked right ventricular dilatation and slight left ventricular dilatation. The motion of the right ventricular wall and left ventricular posterior wall was hypokinetic. The interventricular septum exhibited paradoxical movement. No downward displacement of the septal leaflet of the tricuspid valve suggestive of Ebstein's disease was noted. Pulsed Doppler echocardiogram showed flow.
signal in the right atrium indicating tricuspid regurgitation.

Cardiac catheterization disclosed no elevation of oxygen saturation in the right atrium or right ventricle. Right atrial pressure was increased, being 17 mmHg at the a wave and 30 mmHg at the v wave. Right ventricular systolic pressure was 30 mmHg, but the end-diastolic pressure was elevated to 20 mmHg, constituting a dip and plateau pattern. Right ventriculogram (Fig. 3) revealed marked dilatation and diffuse depression of contractility of the right ventricle, prolonged opacification of the right ventricle, and tricuspid regurgitation.

From the above results, the patient was suspected to have right ventricular dilated cardiomyopathy or Uhl's anomaly and was treated medically. However, he developed intractable right heart failure with progressive cardiomegaly and hepatomegaly accompanied by ascites and hypoproteinemia, and died two years and five months after the first admission.

At autopsy, the heart weighed 290 gm and showed marked dilatation of the right atrium and ventricle and slight dilatation of the left ventricle.

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The circumferences of the tricuspid ring and mitral ring were 13 and 7.8 cm, respectively, and the right ventricular wall was only 1.5 mm thick, resembling the condition of the heart with Uhl's anomaly. The position and morphology of the tricuspid valve were normal (Fig. 4). Histological examination of the left ventricle revealed degeneration and fibrosis of the myocardium. Similar changes were noted in the right ventricular myocardium and papillary muscle accompanied by moderate lymphocytic infiltration (Fig. 4).

Based on these histopathological findings, a diagnosis of idiopathic myocarditis with marked right ventricular dilatation was made.

Patient 2
This patient was born after normal gestation with a body weight of 3,400 gm. Neonatal asphyxia was absent. The family history was not contributory. Three months after birth, anemia and thrombocytopenia were noted, but these conditions were successfully treated at another hospital. At the age of 10 months, he was admitted again to the same hospital due to frequent attacks of convulsion and vomiting, and a clinical diagnosis of intracranial bleeding was made. Transient hyperglycemia was also present. Around this time, the patient presented with cardiomegaly, oliguria, hepatomegaly, and edema, and digitalis and diuretics were initiated. He was transferred to our hospital at the age of one year and four months due to cardiomegaly and persistent symptoms of right heart failure.

On admission, the patient was 80.7 cm tall and weighed 8.6 kg. The blood pressure was 110/60 mmHg. The respiratory rate was 45/min, and no cyanosis was noted. Tachycardia (220/min) and occasional arrhythmias were observed, and ECG suggested paroxysmal supraventricular tachycardia and occasional premature ventricular contractions. After intravenous injection with a digitalis preparation, a heart rate of about 120/min in atrioventricular junctional rhythm were achieved on the next day. Edema of the eyelids and horizontal nystagmus were noted bilaterally. Respiratory sounds were clear, cardiac murmur was absent, but cardiac sounds were faint with gallop rhythm. The liver was palpated for 5 cm below the costal margin.

Peripheral blood indicated no anemia, the white blood cell count was 8,000/mm$^3$, and the platelet count was $23.6 \times 10^4$/mm$^3$. CRP was negative, GOT 115 IU, GPT 78 IU, LDH 703 IU, and CPK 17 IU; GOT, GPT, and LDH levels were slightly elevated. These levels, considered to be due to congestive liver, returned to normal four days later by diuretic therapy. The condition appeared to be similar to that of Patient 1. Titration of antibodies to type A and type B Coxsakie viruses, echoviruses, type A and type
B influenza viruses, and poliovirus showed no significant elevations.

Chest roentgenogram (Fig. 5) revealed spherical dilatation of the heart, and the cardiothoracic ratio was 71%. No enhancement of pulmonary vascular markings was observed, and pulmonary congestion was absent. ECG (Fig. 5) showed regular heart rate of 100/min. No P wave preceded the QRS complex, and the rhythm was atrioventricular junctional. The QRS axis was 22°, the amplitude of the ORS wave tended to be reduced, and right ventricular conduction disturbance was noted. Depression of the ST segment and inversion of T wave were observed in leads I, aV_L, and V_2-V_7, suggesting myocardial impairment.

Two-dimensional and M-mode echocardiograms (Fig. 6A, 6B, 6C) showed dilatation of the right ventricle with an end-diastolic dimension of 3.5 cm. Paradoxical movement was noted in the upper interventricular septum (Fig. 6B). The left ventricular end-diastolic dimension was 1.7 cm, and the thickness and motion of the lower septum and the left ventricular posterior wall were normal (Fig. 6C). No findings of Ebstein's disease were noted. Pulsed Doppler echocardiogram showed flow signal in the right atrium suggestive of tricuspid regurgitation.

CT examination of the head, conducted because of the history of intracranial bleeding and the presence of bilateral horizontal nystagmus, revealed infarction of the right temporal lobe and atrophy of the entire brain.

The patient died of progressive cardiomegaly and intractable right heart failure 4 months after the admission, despite active treatment with digitalis and diuretics. He had presented with unaccountable anemia and thrombocytopenia three weeks after admission, which were resistant to treatment. Insulin therapy was also conducted for hyperglycemia that appeared 10 days before death.

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The autopsied heart weighed 95 gm and showed marked right atrial and ventricular dilatation and slight left ventricular dilatation. The circumferences of the tricuspid ring and mitral ring were 8 and 5.5 cm, respectively. The thickness of the right ventricular wall was 1.5–3 mm as compared to that of the left ventricular wall of 8 mm. No morphological abnormalities were noted in the tricuspid valve (Fig. 7). Histologically, the left ventricular myocardial tissue showed degeneration and diffuse fibrosis. The right ventricular myocardium and papillary muscle showed similar changes and massive infiltration of lymphocytes (Fig. 7).

From these histopathological observation, a diagnosis of idiopathic myocarditis with marked right ventricular dilatation was made as in Patient 1. Pathological examination revealed no pancreatitis. Although the nearly simultaneous occurrence of myocarditis and intracranial bleeding suggests a causative relationship between the two conditions, cranial autopsy was not carried out and the relationship was not clarified.

**DISCUSSION**

Patients 1 and 2 showed degeneration or necrosis and fibrosis of the myocardium as well as a number of infiltrating lymphocytes in both ventricles. These findings are considered to be sufficient grounds for a diagnosis of chronic dilated type idiopathic myocarditis. A striking characteristic in these two patients was the marked dilatation of the right ventricle compared to that of the left ventricle.

**Causes of Right Ventricular Dilatation in Chronic Myocarditis**

In Patient 1, the right ventricular systolic pressure measured by cardiac catheterization was normal, and the clinical history and chest roentgenogram suggested no pulmonary congestion.

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In Patient 2 also, the second cardiac sound was faint and pulmonary congestion was absent, excluding a consequence of left ventricular dysfunction as the major cause of the right ventricular dilatation. Histological examination showed pathologic changes in the right ventricular myocardium, and chronic myocarditis in itself was considered to result in right ventricular dilatation of varying degrees. Although what initially caused right heart failure and right ventricular enlargement is unknown, these conditions were apparently worsened by tricuspid regurgitation resulting from the above conditions. Dilated cardiomyopathy with right ventricular dilatation but without left ventricular dysfunction has been reported in adults. Therefore, in patients with chronic myocarditis or dilated cardiomyopathy, the possibility of dilatation of not only the left ventricle but also the right ventricle, which may readily attain a remarkable magnitude in the presence of tricuspid regurgitation, must be given adequate attention.

Congenital Heart Diseases Differentiated from Chronic Myocarditis Characterized by Right Ventricular Dilatation

These congenital heart diseases include Ebstein's disease, Uhl's anomaly, and RVD. As our patients had the onset in childhood, differentiation of chronic myocarditis from these diseases was particularly important in connection with the possibility of surgical treatment. An anatomical characteristic of Ebstein's disease is downward displacement of the septal and posterior leaflets of the tricuspid valve into the right ventricle resulting in division of the ventricle into atrialized and functional compartments. The possibility of this disease was readily excluded in our patients since two-dimensional echocardiogram and right ventriculogram showed no downward displacement of the tricuspid valve. Uhl's anomaly and RVD cause total or partial congenital defects of the right ventricular myocardium and is characterized by right ventricular dilatation and right heart failure or arrhythmias. In these disorders, left ventricular function is considered to be normal with rare exceptions. Patient 1 showed depressed function and slight dilatation of the left ventricle, and the presence of the left ventricular dysfunction served to differentiate this disease from Uhl's anomaly or RVD. Therefore, it is important in evaluating two-dimensional echocardiogram that dilatation or functional impairment of the left ventricle, which may appear relatively normal or even reduced in size in contrast to the markedly enlarged right ventricle, should not be overlooked. In Patient 2, dilatation or dysfunction of the left ventricle was not clear in two-dimensional and M-mode echocardiograms, but ECG showed a depression in the
ST segment and inversion of T wave in left precordial leads. Thus, changes in ECG are also important in detecting left ventricular myocardial impairment.11

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