Case Report

Two-chambered Right Ventricle
Report of Four Cases with Special Reference
to the Clinical Diagnosis

Kunitake Hashiba, Tomoyuki Katayama, Akira Takahashi, Akio Ono,
Junshi Fujita, Shuzo Matsu, Yoshimi Takahashi, Masato Yoshioka,
Kiyoshi Mori, and Shun-ichi Mochinaga

Anomalous muscle bundles sometimes divides the right ventricle into two chambers and obstructs the pathway of blood stream in the right ventricular cavity.\(^1\)\(^-\)\(^8\) This condition is usually referred to as two-chambered or double-chambered right ventricle. Although two-chambered right ventricle is mostly associated with ventricular septal defect\(^1\)\(^-\)\(^3\),\(^5\)\(^-\)\(^7\) or pulmonary stenosis,\(^4\)\(^,\)\(^8\) several cases with isolated two-chambered right ventricle have also been reported.\(^4\)\(^,\)\(^8\)

Infundibular pulmonary stenosis was associated in Cases 1 and 2 of our series, coarctation of the aorta and congenital pyroxic stenosis in Case 3, and ventricular septal defect and essential hypertension in Case 4. Case 4 was 52 years old, being the oldest case ever reported in the literature.

The angiocardiology provides the most reliable and definitive means for diagnosis of two-chambered right ventricle.\(^1\)\(^-\)\(^4\),\(^8\) Diagnostic significance of the other clinical findings have little been ever discussed. The present report concerns diagnostic significance of clinical findings in four cases, including auscultatory, roentgenologic, electrocardiographic, vectorcardiographic, cardiac catheterization and angiocardioigraphic findings.

Report of Cases
Case 1. T. M.
A 19-year old girl was admitted to the hospital because of exertional dyspnea and easy fatiguability on November 4, 1965. In her childhood she could not run around with her friends and preferred to play indoors. At the age of 4 years she was noticed to have a cardiac disease by a physician when she developed syncope while walking with her grandmother. The patient had to stop walking several times to rest on her way to school during nine years of school life. She never took part in gymnastic exercise at school. Cyanosis was not noticed. After completing school she worked for four months, but gave up her job because of easy fatiguability and exertional dyspnea.

Physical examination revealed a rather slender and alert girl, being 153 cm tall and weighing 42 kg. There were no signs of cyanosis, clubbing or dyspnea. Blood pressure was 134 mmHg systolic and 84 diastolic. Heart rate was 76 and respirations were 20 per minute. There was no chest deformity. A grade V/VI pansystolic murmur was audible at the third left intercostal space and radiated in all directions, especially upwards and toward the apex. A systolic thrill was palpable widely in the third and forth intercostal spaces. The second sound at the pulmonary area was single and slightly decreased in intensity. The diagnosis of infundibular stenosis of the right ventricle with an unusual angiocardioigraphic

(Received for Publication, October 6, 1969)
The First Department of Internal Medicine, Nagasaki University School of Medicine, Nagasaki

Japanese Circulation Journal Vol. 34, March 1970 167
features was made by right heart catetherization and selective right ventricular angiography.

The patient was operated upon on February 9, 1966. In addition to an infundibular stenosis, an anomalous muscle mass was found below the infundibulum producing severe stenosis and dividing the right ventricle into two parts. The anomalous muscle mass and infundibular stenosis were excised. A transient slight ascites developed postoperatively and disappeared with digitalis and diuretics. The patient was discharged three months later. Exertional dyspnea almost disappeared on usual activity in her daily life.

Case 2. N. K.

A 17-year old girl was admitted to the hospital because of exertional dyspnea and palpitation on June 3, 1966.

The patient was first noticed to have heart disease by a school doctor and was advised not to take part in gymnastics when she was 6 years old. However, the patient had no complaint at that time and could walk for 10 minutes to school every day without any difficulties. The patient first developed dyspnea and palpitation at the age of 10 years when she could not walk along with her classmates on a school excursion of two hours. Thereafter, the patient occasionally experienced exertional dyspnea and palpitation on her way to school, especially in winter. Exertional dyspnea and palpitation increased in intensity at the age of 15 through 17 years.

Physical examination revealed a rather obese girl being 151 cm tall and weighing 58 kg. There were no signs of clubbing and cyanosis although the cheeks were reddish-colored. Respirations were 22 and heart rate was 76 per minute. Blood pressure was 124 mmHg systolic and 80 diastolic. Although a slight rightward scoliosis of the thoracic vertebral column was observed, there was no deformity of the anterior chest wall. A grade V/VI, pansystolic murmur was audible in the third left intercostal space and radiated in all directions, especially to the left, toward the apex. A systolic thrill was palpable in the third left intercostal space and less intensively in the second space. The second pulmonic sound at the pulmonary area was single and slightly diminished in intensity. A two-chambered right ventricle associated with infundibular stenosis was diagnosed by right heart catheterization and angiography.

The patient was operated upon on September 8, 1966. Postoperative course was uneventful, and exertional dyspnea and palpitation were markedly diminished. The diagnosis was confirmed at operation.

Case 3. K. T.

A 14-year old girl was admitted to the hospital for evaluation of hypertension. Her mother had been healthy and uneventful during the period of pregnancy and delivery. The patient looked normal at birth. Vomiting developed a week after birth every time following feeding and persisted thereafter. A pyloric stenosis was diagnosed and the growth and development of the patient was retarded during childhood. She was operated upon for pyloric stenosis at the age of 13 years.

An abnormal pulsation over the right neck was noticed by her mother and a precordial murmur was first noticed by a physician when she was 6 years old. Physical activity of the patient had been limited because of easy fatiguability, palpitation and dyspnea on exertion. The patient developed pain in the legs on walking, and felt coldness of the legs in winter. Coarctation of the aorta was diagnosed by a physician who referred the patient to the hospital for further examination.

The patient was a rather small girl for her age and in no acute distress. There were no signs of cyanosis, clubbing and dyspnea. Heart rate was 82 and respirations were 20 per minute. Blood pressure was 190 mmHg systolic and 74 diastolic in the right brachial artery, 166 systolic and 70 diastolic in the left brachial, and 104 systolic in the right thigh. Both radial arteries were palpable. Neither popliteal nor dorsalis pedis arteries were palpable bilaterally. The third through seventh intercostal arteries were palpable with marked intensity.

There was no significant chest deformity. A slight systolic thrill was occasionally felt in the second right intercostal space. A grade IV/VI, systolic murmur was audible at the left second and third intercostal spaces, radiating along the left sternal border. Systolic murmurs were also heard over the neck, back and upper abdomen with intensity of grade II to III/VI. The second heart sound at the pulmonary area was slightly accentuated. Pulmonary stenosis was suspected before catheterization in addition to coarctation of the aorta. Aortography demonstrated a severe coarctation of the aorta, and right heart catheterization and angiography revealed a two-chambered right ventricle.

The patient was operated upon for coarctation

*Japanese Circulation Journal Vol. 34, March 1970*
of the aorta on October 11, 1966, using a tefron tube, for reconstruction of the aorta. Blood pressure was 112 mmHg systolic and 50 diastolic in the right brachial artery after the operation and was almost the same in the legs. The systolic murmur at the left sternal border was not changed after operation with intensity of grade IV. The patient is to be operated upon for the cardiac anomaly in future.

Case 4. T. M.

A 52-year old man was admitted to the hospital on September 16, 1966 because of pain of the left extremities, occasional anginal pain and dizziness.

His mother died of apoplexy and one of his sisters had hypertension. Two brothers died during infancy of unknown causes. Another sister died of meningitis with some heart trouble at the age of 45 years. The other four siblings of the patient are living and healthy. The patient was healthy in his infancy through adolescence and had no complaint with his heart. He developed edema of lower extremities at the age of 29 years when he was in military service and was admitted to an army hospital for six months. After discharge from the military service he worked as a cook without physical difficulties. Hypertension of 220 mmHg systolic was found when he was 42 years old and occasionally he developed slight dizziness. Three years later he suffered from left hemiparesis and gradually improved. Pain in the left extremities, especially of the leg, had persisted since the left hemiparesis developed in 1960. The patient also complained of anginal pain and dizziness since 1963.

The patient was a slightly obese man in no acute distress. His cheeks and lips looked reddish-brown although neither cyanosis nor clubbing of the nails were observed. There was no chest deformity. Heart rate was 82 and respirations were 17 per minute. Blood pressure was 178 mmHg systolic and 90 diastolic. A slight hemiparesis was observed and Déjerine-Roussy thalamic syndrome was suspected of causing sensation of pain in the left extremities. A systolic thrill was palpable in the left second and third intercostal spaces. A grade V/VI, harsh systolic murmur was audible at the third intercostal space, radiating along the left sternal border. Second heart sound was audible at the pulmonary area with a markedly decreased intensity. A two-chambered right ventricle associated with ventricular septal defect was diagnosed by cardiac catheterization and angiocardiography. The patient denied operation and was discharged.

Physical Findings

Significant retardation of the growth and development was not found in any of the four cases, although Cases 1 and 3 were rather small girls for their age. Significant deformity of the chest, cyanosis and clubbing were not found in all cases.

The systolic murmur with a diminished second pulmonic sound suggesting pulmonary stenosis was heard, and the marked systolic thrill was palpable in Cases 1, 2 and 4. In Case 3 the second heart sound was not diminished at the pulmonary area and the systolic thrill was only occasionally palpable. The maximum point of the systolic murmur and thrill was at the third intercostal space in most cases. The systolic murmur radiated along the left sternal border, and in Cases 1 and 2, also radiated toward the apex without significant decrease in intensity. It would be mentioned that the systolic murmur was more widely distributed over the precordium in these cases than in the usual cases with isolated pulmonary stenosis.

Phonocardiogram

Phonocardiograms revealed that the systolic murmur started with the first sound and ended just before the markedly diminished second sound in Cases 1, 2 and 4. In Case 3 the second sound was normal or slightly increased in intensity. The systolic murmur showed a diamond-shaped appearance with the peak of the murmur located in the middle of the systolic period in Cases 1 and 4, the same as usually observed in ventricular septal defect. The murmur in Case 2 also displayed a diamond shape although the peak of the murmur was located a little later than the middle of the systolic period. In Case 3 the murmur was of stenotic type with the peak located at the two thirds of the systolic period.

Chest Roentgenogram

Cardiomegaly was demonstrated in the chest roentgenograms of all cases, as shown in Figs. 1 through 4. Prominence of the main pulmonary artery suggesting poststenotic dilatation was not found, and pulmonary vascular markings were normal in all cases.

In Cases 1 and 2 the heart was markedly enlarged in the roentgenograms, approaching closely to the left lateral wall of the chest. The heart was of globular shape in Case 1. Lateral views showed right ventricular hypertrophy in Cases 1

*Japanese Circulation Journal Vol. 34, March 1970*
Fig. 1. Case 1. A globular, marked cardiomegaly without poststenotic dilatation of the pulmonary trunk.

Fig. 2. Case 2. A marked cardiomegaly without poststenotic dilatation of the pulmonary trunk.

Fig. 3. Case 3. A globular, moderate cardiomegaly.

Fig. 4. Case 4. A marked cardiomegaly suggesting left ventricular hypertrophy and a broadened aorta. No prominence of the pulmonary trunk.

Fig. 5. Case 1. The electrocardiogram showed right ventricular hypertrophy in the chest leads. Predominant R waves were seen in V₁ through V₆. Mean electrical axis was around +90 degrees, and a very small late r wave was found as compared to the Q wave in aVR. All leads of the electrocardiogram were taken with ½ standard. The vectorcardiogram showed that the almost entire loop of QRS was oriented anteriorly, leftwards and inferiorly.

Fig. 6. Case 2. The electrocardiogram showed right ventricular hypertrophy in the chest leads. Predominant R waves were seen in V₁ through V₆, as same as in Case 1. Mean electrical axis was around +100 degrees, but there was no r wave in aVR. All leads of the electrocardiogram were taken with ½ standard. The vectorcardiogram showed that the entire loop of QRS was orientated anteriorly, leftwards and inferiorly, as same as in Case 1.

and 2. A globular cardiomegaly was observed in the postero-anterior roentgenogram of Case 3 although right ventricular hypertrophy was equivocal in the lateral view. In Case 4 the roentgenogram showed a left ventricular hypertrophy and dilatation of the aorta, and a loss of normal prominence of the pulmonary artery.

Electrocardiogram and Vectorcardiogram

The electrocardiograms revealed right ventricular hypertrophy in Cases 1 and 2, and combined right and left ventricular hypertrophy in Case 4, as shown in Figs. 5, 6 and 8, respectively. It is the most remarkable finding in these cases that the chest lead showed qR, R, or Rs patterns in V₃R though V₃ without displaying RS or Rs patterns. Right axis deviation was very slight both in Cases 1 and 2, being +90 and +100 degrees, respectively. In Case 3 the electrocardiogram demonstrated a left ventricular hypertrophy without any signs of right ventricular hypertrophy, as shown in Fig. 7. The P waves were peaked and 0.3 mm tall both in Lead I of Case 1 and in V₁ of Case 2. No significant abnormalities of the P waves were observed in Cases 3 and 4.

The vectorcardiograms by Frank method revealed right ventricular hypertrophy in Cases 1, 2 and 4, with the entire QRS loop inscribing almost anteriorly and leftwards, as shown in Figs. 5, 6 and 8, respectively. There was little part of QRS loop orienting posteriorly or rightwards. The vectorcardiogram of Case 3 demonstrated a left ventricular hypertrophy, as shown in Figs. 7.

Right Heart Catheterization

The results of right heart catheterization are summarized in Table I. No intracardiac shunting was demonstrated in Cases 1, 2 and 3. In Case 4 neither pulmonary artery nor outflow tract of the right ventricle were entered, and the blood samples showed no significant difference in oxygen content between the right atrium and the proximal part of the right ventricle. The catheter was passed into the persistent left superior caval vein in Case 3, and into the left atrium through the persistent foramen ovale in Case 4.

Significant pressure gradients were demonstrated between the distal and proximal chambers.
### Table I Right Heart Catheterization

<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>Sex</th>
<th>Pulmonary</th>
<th>Right Ventricle</th>
<th>Cardiac output (l/min)</th>
<th>Intracardiac shunt</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td>artery</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>infundibulum</td>
<td>distal</td>
<td>proximal</td>
<td>atrium</td>
</tr>
<tr>
<td>1.</td>
<td>T.M.</td>
<td>19</td>
<td>F</td>
<td>25/6</td>
<td>52/0–3*</td>
<td>150/0–10*</td>
</tr>
<tr>
<td>2.</td>
<td>N.K.</td>
<td>17</td>
<td>F</td>
<td>30/10</td>
<td>60/6–10*</td>
<td>230/4–20*</td>
</tr>
<tr>
<td>3.</td>
<td>K.T.</td>
<td>14</td>
<td>F</td>
<td>42/10</td>
<td>46/0–5*</td>
<td>80/0–5*</td>
</tr>
<tr>
<td>4.</td>
<td>T.M.</td>
<td>52</td>
<td>M</td>
<td></td>
<td>148/0–15*</td>
<td>18-4</td>
</tr>
</tbody>
</table>

**Note:** * indicates the end-diastolic pressure

---

**Fig. 7.** Case 3. The electrocardiogram and vectorcardiogram showed left ventricular hypertrophy. The chest leads were taken with ½ standard.

**Fig. 8.** Case 4. The electrocardiogram showed right ventricular hypertrophy in the chest leads. Predominant R waves were seen in V1 through V6, as in Cases 1 and 2. Mean electrical axis was around +75 degrees, and the T wave of aVR was small. All leads of the electrocardiogram were taken with ½ standard. The vectorcardiogram showed that the almost entire loop of QRS was oriented anteriorly, leftwards and inferiorly.

---

of the right ventricle in Cases 1, 2 and 3. The other pressure gradients were recorded in Cases 1 and 2, suggesting infundibular stenosis. The higher pressure of the proximal chamber was always recorded in Cases 1, 2 and 3 when the catheter was pulled back continuously from the pulmonary artery to the right atrium. In Case 4 the pressure of the proximal part of the right ventricle was also markedly elevated. The end-diastolic pressure of the right ventricle was elevated in Cases 2 and 4. The right atrial pressure was also elevated with a prominent 'a' wave in Cases 2 and 4, and was normal in Cases 1 and 3.

**Angiocardiogram**

Selective right ventriculography was performed in all four cases. Six films were taken per second for five seconds using SHONANDER’s biplane film exchanger.

In Case 1 the early antero-posterior film showed that the right ventricle was divided by an unusual muscle band into two major chambers, larger proximal and smaller distal ones, and that a tubelike infundibular stenosis was formed between the distal chamber and the pulmonary valve (Fig. 9, Upper). The lateral view showed...
Fig. 9. Case 1. Upper: Taken 1/3 second after the beginning of the injection of contrast medium. In the anteroposterior view (left) the right ventricle was divided into two parts, larger proximal one and smaller distal one. A filling defect was observed, originating at the region just below the infundibulum and proceeding obliquely toward the apex. An infundibular stenosis was also visualized. In the lateral view (right) a diverticulum-like part of the right ventricle was observed to be bulging up along the pulmonary trunk.

Below: Taken 1/2 second after the upper films. In the antero-posterior view (left) two chambers of the right ventricle were fully filled with contrast medium and could not be identified clearly from each other. The infundibular stenosis was also demonstrated in this film. Unusually abundant trabeculation was seen in both parts of the ventricle. The lateral view (right) showed more clearly the diverticulum-like bulging of the right ventricle.
Fig. 10. Case 2. Upper: Taken 1/6 second after the beginning of injection of contrast medium. In the antero-posterior view (left) a large muscle mass was observed at the left upper part of the right ventricle. The lateral view (right) suggested hypertrophy of the septal band constricting the outflow tract of the ventricle.

Below: In diastole, 1/3 second after the upper films. More advanced filling of the right ventricle was showed. It was demonstrated that two parts of the ventricle encircled the muscle mass and approached one another at their most peripheral portions. The lateral view (right) showed that the distal chamber of the right ventricle was placed anteriorly to the proximal one, and that another small part of the ventricle was bulging up along the pulmonary trunk like a diverticulum.

*Japanese Circulation Journal, Vol. 34, March 1970*
another small part of the right ventricle bulging up along the trunk of the pulmonary artery, like a diverticulum. In the later antero-posterior film taken in diastole the distal chamber was completely filled with contrast medium, and it was impossible to identify two chambers of the right ventricle (Fig. 9, Below). The right ventricle was markedly dilated and seemed to form the left border of the heart. This film also showed an infundibular stenosis and abnormally developed trabeculation or unusual muscle bundles in the right ventricular cavity. The diverticulum-like bulging of the right ventricle was also fully filled with contrast medium in the lateral view.

In Case 2 the early antero-posterior film revealed that most of the outflow tract of the right ventricle was occupied by an unusual muscle mass (Fig. 10, Upper). The later antero-posterior film taken in diastole showed a more advanced filling of the right ventricle (Fig. 10, Below). Peripheral portions of the two chambers of the right ventricle approached one another and seemed to encircle the large unusual muscle mass. The right ventricle was dilated and formed the left border of the heart similar to Case 1. An uneven distribution of the contrast medium in the proximal chamber suggested the presence of abundant abnormal muscle bundle in the cavity. In Case 2 the lateral view also showed a diverticulum-like part of the right ventricle, bulging up along the trunk of the pulmonary artery (Fig. 10, Below).

In Case 3 the contrast medium was injected into the distal chamber of the right ventricle, and part of the contrast medium seemed to flow back into the proximal chamber. Both antero-posterior and lateral views showed an abnormal muscle mass in the cavity of the right ventricle, partitioning the right ventricle into two chambers with multiple openings. Unusually developed trabeculation was observed in the distal chamber of the right ventricle. Concentration of the contrast medium was not equal between the two chambers as shown in the three successive sets of films with time intervals of 1/6 second (Fig. 11). Aortography revealed a severe coarctation of the descending aorta.

In Case 4 the antero-posterior and lateral films showed that the right ventricle was divided into two chambers by a large muscle mass (Fig. 12, Upper). The size of the two chambers was apparently equal. Left ventriculography revealed a ventricular septal defect with a shunt from the left ventricle into the distal chamber of the right ventricle (Fig. 2, Below).

**Discussion**

**Clinical Significance**

Ventricular septal defect is the most common congenital heart disease which is associated with two-chambered right ventricle, and valvular pulmonary stenosis seems to be the second one. Patients with a two-chambered right ventricle and ventricular septal defect might simulate cyanotic or slightly cyanotic tetralogy of Fallot. Three cases of patent ductus arteriosus complicating two-chambered right ventricle were reported and two of them were associated with ventricular septal defect. Isolated two-chambered right ventricle has also been reported. Out of eleven patients with intact ventricular septum of Ward et al., anomalous muscle bundle of the right ventricle was the only cardiac anomaly in five patients, and valvular pulmonary stenosis was associated in the remainder.

In Cases 1 and 2 of our series infundibular stenosis was the additional cardiac abnormality, but neither ventricular septal defect nor valvular pulmonary stenosis were found. Case 3 had coarctation of the aorta and congenital pyriform stenosis as additional anomalies. There has not been reported such a combination of congenital anomalies as found in this case. Case 4 had ventricular septal defect and systemic hypertension, and was thought to be the oldest case of two-chambered right ventricle ever reported in the literature. Only three adult cases have been reported, with the age of 23, 26 and 30 years respectively.

Two-chambered right ventricle may be overlooked in some patients and diagnosed erroneously as an isolated ventricular septal defect, pulmonary stenosis or tetralogy of Fallot. Lucas et al. stated that failure to recognize the nature of the obstruction resulted in inadequate surgical treatment and death in two of their patients with two-chambered right ventricle. Coates et al. also stated that one of their patients associated with valvar pulmonary stenosis died postoperatively, probably because of failure to recognize and excise the anomalous muscle band at operation.

In Case 1, the first patient of our experiences, the anomalous muscle mass was first recognized at operation although unusual angiocardiographic findings had been noted before operation. In Cases 2, 3 and 4 the anomaly was diagnosed by means of cardiac catheterization and especially by angiocardiography. Two-chambered right ventricle was strongly suspected in Case 2 by auscul-
Fig. 11. Case 3. Three successive sets of films were taken with intervals of 1/6 second, respectively. In each film an unusual muscle mass was observed in the middle of the right ventricle. The right ventricle was divided into two chambers. Unusual muscle bundles were shown in the distal chamber while no marked trabeculation was observed in the proximal chamber. The first films (upper) showed almost equal concentration of the contrast medium between two chambers. The second films (middle) showed that most amount of the contrast medium was ejected out from the proximal chamber. The last films (below) suggested a significant regurgitation of the contrast medium from the distal chamber into the proximal one.

Japanese Circulation Journal Vol. 34, March 1970
Fig. 12. Case 4. Upper: Selective right ventriculography revealed that the right ventricle was divided into two chambers by an unusual muscle mass encircling the stenotic canal.

Below: Left ventriculography was performed when the tip of catheter entered into the left ventricle through a patent foramen ovale. A left-to-right shunt was visualized from the left ventricle into the distal chamber of the right ventricle. The films presented were taken in systole.

*Japanese Circulation Journal  Vol. 34, March 1970*
tatory, roentgenologic and electrocardiographic findings because of their close similarity to those of Case 1.

**Heart Murmur and Sound**

Patients with two-chambered right ventricle always have a systolic murmur which closely resembles that of ventricular septal defect or valvular pulmonary stenosis. The systolic murmur was diamond-shaped and pansystolic in Cases 1, 2 and 4 of our series, although two of these patients had no ventricular septal defect at all. In Cases 1 and 2 the systolic murmur was heard with maximum intensity in the third intercostal space and was widely distributed over the precordium without marked decrease in intensity. This was one of the clinical features which led to suspicion of two-chambered right ventricle before catheterization in Case 2. The systolic murmur of Case 4 was relatively localized at the left lower sternal border and was compatible with an usual ventricular septal defect. The systolic murmur was of stenotic type only in Case 3 of our series. The systolic murmur of patients with two-chambered right ventricle was often heard loudest at the left lower sternal border even when the ventricular septum was intact. The systolic murmur was described as being of ejection-type in three out of four cases of Coates et al. with two-chambered right ventricle as an isolated cardiac anomaly. The second heart sound was decreased in intensity at the pulmonary area in Cases 1, 2 and 4 of our series, and was incompatible with isolated ventricular septal defect because the electrocardiograms of these patients showed right ventricular hypertrophy.

**Roentgenogram**

Marked cardiomegaly of somewhat globular shape was observed, and there was no poststenotic dilatation of the pulmonary artery in the roentgenograms of Cases 1, 2 and 3 of our series. This type of marked cardiomegaly is not rare in infants or children with severe pulmonary stenosis but in adolescent or adult patients of pulmonary stenosis, cardiomegaly is usually slight or moderate even in severe cases and poststenotic dilatation of the pulmonary trunk is one of the most common roentgenographic findings. There was no significant prominence of the pulmonary trunk in Case 4 although the systolic murmur was compatible with ventricular septal defect.

Cardiomegaly in two-chambered right ventricle has been reported to be of slight to severe degree in the chest roentgenograms, probably depending on the severity of the disease and on the difference of associated cardiac anomaly. The pulmonary artery was normal in the roentgenograms in most patients in whom two-chambered right ventricle was an isolated anomaly, although slight prominence of the pulmonary artery was described in some patients. Pulmonary vascular markings were increased in patients with significant left-to-right shunt.

**Electrocardiogram and Vectorcardiogram**

The electrocardiogram showed right axis deviation and right ventricular hypertrophy in many of the cases previously reported but in others, incomplete right bundle branch block, biventricular enlargement, left ventricular overload or even normal electrocardiograms were also observed. A wide variety of the electrocardiographic findings of this anomaly could be explained by varying severity of the anomaly itself and by differences of the accompanying cardiac defects.

In Cases 1 and 2 of our series the electrocardiograms revealed right ventricular hypertrophy. The most striking electrocardiographic finding in these cases was that most of the chest leads, V3R through V7, showed R pattern. A qR pattern was observed in V1 of Case 1, and Rs pattern was observed in V2 of Case 1 and in V1 and V2 of Case 2, but either q or s waves were very small as compared with R waves in the same leads. Mean electrical axis of Case 1 was around +90 degrees and that of Case 2 was +100 degrees, and aVR displayed QS or Qr pattern with a tiny late-R in both cases, in spite of apparently severe right ventricular hypertrophy in the chest leads. In one of the cases of Coates et al. the prominent R in lead V4R, absence of a significant R in aVR, and absence of significant S in V6 were pointed out in the electrocardiogram. They stated that these findings might indicate localized hypertrophy of the inflow chamber of the right ventricle. It would, however, be an alternative and more probable explanation in Cases 1 and 2 of our series that marked hypertrophy and dilatation of the right ventricle were responsible for the peculiar electrocardiographic features. Selective right ventriculography demonstrated that the right ventricle was markedly enlarged, reaching the left border of the heart in both cases. The vectorcardiograms of these cases demonstrated that the entire loop of QRS was oriented anteriorly, and there was little part of the QRS loop inscribing poster-
iorly or rightwards. These electrocardiographic and vectorcardiographic features would have some diagnostic significance in severe cases of isolated or predominant two-chambered right ventricle.

The chest lead of Case 4 displayed tall R followed by small s in V₁ through V₆, and the mean electrical axis in the limb lead was around +60 degrees. This finding showed some similarity to those of Cases 1 and 2, although it might indicate biventricular hypertrophy due to the coexistence of two-chambered right ventricle, ventricular septal defect and systemic hypertension. The QRS loop was inscribed mostly anteriorly and leftwards, and there was no evidence of left ventricular overload in the vectorcardiogram of Case 4.

Right Heart Catheterization

Right heart catheterization is helpful in demonstrating the pressure-gradient between two chambers of the right ventricle. The pressure gradient was also recorded in our patients, except Case 4 in whom the distal chamber was not entered. Sometimes the tip of catheter might skip the proximal chamber and fail to record the high pressure. WARDEN et al. stated that in some of their patients no consistent right ventricular pressure gradients were recorded in repeated attempts, because the catheter tip immediately snapped back into the inflow portion from the pulmonary artery. Although it might be possible during catheterization to locate precisely the point of obstruction in the right ventricular cavity in some cases, it would be very difficult to differentiate between two-chambered right ventricle and infundibular stenosis only by cardiac catheterization.

In Cases 2 and 4 of our series the right atrial pressure and the end-diastolic pressure of the right ventricle were significantly elevated, indicating right ventricular dysfunction. The right atrial and right ventricular end-diastolic pressures were either normal or not listed in most of the cases previously reported, except one case.

Selective Right Ventriculography

Selective right ventriculography provides the most conclusive evidence for diagnosis of two-chambered right ventricle. Anomalous muscle mass or muscle bundles are visualized as a filling defect, usually originating in the region of the supraventricular crista and proceeding obliquely toward the apex. Biplane angiography with a rapid film exchanger is necessary for the correct diagnosis, because anomalous muscle mass is demonstrated better in systole than in diastole. The size of the two chambers of the right ventricle is almost equal in some cases. In others, the proximal chamber seemed to be larger or smaller than the distal chamber in the angiocardiograms. In Cases 1 and 2 of our series the proximal chamber was larger than the distal chamber while in Cases 3 and 4 the two chambers were of equal size. Abundant small, thin areas of filling defect were visualized within the distal chamber in Cases 1, 2 and 3 when the distal chamber was fully filled with contrast medium. This finding suggested the presence of unusual muscle bundles in the cavity, which was verified at operation in Cases 1 and 2. In Case 3 of HARTMANN et al. a coarse trabeculation was also noted in the low-pressure chamber. In none of our series was poststenotic dilatation of the pulmonary artery observed in the angiocardiograms. Absence of poststenotic dilatation of the pulmonary artery was also mentioned by HARTMANN et al. in the angiocardiograms of two patients.

Clinical Diagnosis

Clinical and laboratory findings of the two-chambered right ventricle vary depending on the severity of the disease and the difference of associated cardiac defects. However, the analyses of the clinical features of our patients reported herein would suggest that the combination of the following findings are helpful for diagnosis prior to cardiac catheterization and angiocardiography, at least in isolated or predominant two-chambered right ventricle: (1) a widely radiating, diamond-shaped systolic murmur at the left lower sternal border, (2) a diminished second heart sound at the pulmonary area, (3) a cardiomegaly of somewhat globular shape with absence of poststenotic dilatation, (4) a predominant R pattern in V₁ through V₆ with absence of significant R in aVR, and (5) a QRS loop inscribed mostly anteriorly and leftwards in the vectorcardiogram.

Summary

Four cases with two-chambered right ventricle are presented; Cases 1 and 2 were associated with infundibular pulmonary stenosis, Case 3 with coarctation of the aorta, and Case 4 with ventricular septal defect and essential hypertension. Case 4 was 52 years old, being the oldest case ever reported in the literature.
Physical, roentgenologic, electrocardiographic, vectorcardiographic, cardiac catheterization and angiocardiographic findings are discussed. Although selective right ventriculography is generally the most reliable and definitive procedure for diagnosis, the coexistence of the following clinical findings would be helpful for diagnosis of two-chambered right ventricle prior to the cardiac catheterization and angiocardioigraphy, especially in cases with isolated or predominant two-chambered right ventricle; (1) a widely distributed, diamond-shaped systolic murmur with the maximum point at the left lower sternal border, (2) a diminished second heart sound at the pulmonary area, (3) a cardiomegaly of somewhat globular shape with absence of poststenotic dilatation, (4) a predominant R pattern in V1 through V6 with absence of significant late R in aVR, and (5) a QRS loop inscribed mostly anteriorly and leftwards in the vectorcardiogram.

Acknowledgement
We would like to express our thanks to Dr. Yoshito Takaoka for his encouragement.

REFERENCES