ECHOCARDIOGRAPHIC FEATURES OF UHL'S ANOMALY
A CASE REPORT

HIRONOBU ANDO, M.D., TADAO YAMAMOTO, M.D.,
MASAHO TAMINTO, M.D., TOMOYO OGAMI, M.D.,
AND SUSUMU YORIFUJI, M.D.

M-mode echocardiography on a fifty-eight year old female with Uhl's anomaly showed several characteristic findings, which were considered to be useful in differentiating Uhl's anomaly from Ebstein's. Those findings were normal tricuspid diastolic closing velocity, the relatively early tricuspid valve opening, easy visualization of the tricuspid valve in usual position and the unusual mid-diastolic pulmonic valve full opening.

The premature opening of the pulmonic valve correlated with the right atrial and ventricular mid-diastolic pressure exceeding that of the pulmonary artery.

UHL's anomaly,
congenital hypoplasia of the right ventricular myocardium, is a rare cardiac disorder. There are only two previous echocardiographic studies on Uhl's anomaly available.

We describe the new echocardiographic findings in an adult patient of this anomaly, correlating with hemodynamic data and show the usefulness of the echocardiography in the diagnosis.

CASE REPORT

The patient was a 58 year old female. She was in good health until age 50 when she had a brief episode of bleeding from pharyngeal ulcer. At that time her family doctor pointed out cardiomegaly on a chest X-ray film at the first time in her life. One year prior to the admission, she had a spell of severe dyspnea on heavy exercise for short duration enough to squat herself associated with diaphoresis and general weakness. Two days later the swelling of her face, chest wall and lower extremities were pointed out. A systolic murmur was first noted at the apex. She was referred to the Hyogo Medical College Hospital on April 1, 1977 for further evaluation of the heart. No history of orthopnea, paroxysmal nocturnal dyspnea or hemoptyis was present. Past history showed cervical lymphadenopathy at the age of six. Family history was not contributory except for her mother's sudden death at the age of 27.

Physical examination on the admission revealed well developed, well nourished female in no distress. No cyanosis or clubbed fingers were present. Blood pressure was 125/80. Heart rate was 52/min regular. The internal jugular vein was distented up to 8.5 cm from the sternal angle with the prominent A and V waves. The cardiac action was weak despite of the enlarged heart.

The first heart sound (S₁) was distant and the second heart sound (S₂) was normal. High

Key Words:
Uhl's anomaly
Echocardiography
Ebstein's anomaly

(Received on October 31, 1978; Accepted on August 28, 1979)
The First Division, The Department of Medicine, Hyogo College of Medicine, 1-1 Mukogawa-cho, Nishinomiya-City, 663 Japan

Japanese Circulation Journal Vol. 43, December 1979 1121
pitched fourth heart sound ($S_4$) was audible. Grade I/VI systolic murmur was audible from the apex to the left lower sternal border. The murmur, however, was not accentuated with inspiration. Lungs were clear. Abdomen was intact. There were superficial varicose veins at the mid third preribial area.

Laboratory data showed normal blood counts and negative urinalysis. Blood chemistry studies were within normal limits. Chest X ray films showed a prominent cardiomegaly (cardiothoracic ratio 0.75) with the right ventricular and the right atrial dilatation. The pulmonary vascular appeared normal except for some hazy shadow at the right lower lung field. Twelve leads electrocardiogram (EKG) revealed the right atrial enlargement, incomplete right bundle branch block and diffuse inverted T waves in Leads I, II, III, aVF and $V_{1-6}$.

M-mode echocardiography of the pulmonic valve and the right ventricle was unusual. The posterior pulmonic leaflet echoes were recorded in the usual way. Assuming the posterior pulmonic valve opens where a diastolic shallow slope echo is suddenly followed by a rapid opening one, the posterior pulmonic valve opened prematurely, almost at middiastole (0.32 second after $S_2$) in inspiration and at the last quarter of the diastole (0.48 second after $S_2$) but still before atrial kick even in expiration (Fig. 1). In addition the pulmonic valve opened prematurely before the onset of the right atrial and ventricular contraction. The premature pulmonic valve opening velocity appeared more accentuated (172 mm/sec to 193 mm/sec) in inspiration than in expiration (78 mm/sec to 92 mm/sec).

The right ventricle was remarkably dilated with its dimension 5.0 cm (Fig. 2). On the contrary, the left ventricular diastolic dimension was small (2.8 cm). The interventricular septum moved paradoxically (Type A) with its thickness normal. The anterior mitral leaflet diastolic excursion (0.9 cm) was small with its early diastolic closing velocity (30 mm/sec) diminished. The tricuspid valve was easily recorded in the usual way. The anterior tricuspid leaflet showed a large diastolic excursion (D-E dimension 18 mm) with a rapid early diastolic closing velocity (EF slope, 95 mm/sec). The tricuspid valve closed 0.03 second after the mitral valve closure. It also opened 0.06 second earlier than the mitral valve (Fig. 2).
Right cardiac catheterization demonstrated a large A wave (16 mmHg) in the right atrium which was transmitted into the pulmonary artery and the right ventricle, creating an unusual pre-systolic high pressure elevation (Fig. 3, Table I).

Following it, there was only a small additional pressure rise (3 mmHg) in the pulmonary artery and in the right ventricle during the systole. The right ventricle also showed a typical dip and plateau type pressure curve in diastole, which
TABLE I  HEMODYNAMIC DATA OF THE RIGHT AND LEFT CARDIAC CATHETERIZATION

<table>
<thead>
<tr>
<th></th>
<th>SYSTOLIC (mmHg)</th>
<th>DIASTOLIC</th>
<th>MEAN</th>
<th>O₂ SATURATION</th>
</tr>
</thead>
<tbody>
<tr>
<td>SCV</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>RA</td>
<td>V = 18</td>
<td>A = 16</td>
<td>12.5</td>
<td></td>
</tr>
<tr>
<td>RV</td>
<td>19</td>
<td>dip = 6.5</td>
<td>14</td>
<td>A&amp;C = 16.5</td>
</tr>
<tr>
<td>PA</td>
<td>19</td>
<td>plateau = 10</td>
<td>13.5</td>
<td>A&amp;C = 16.5</td>
</tr>
<tr>
<td>PC</td>
<td>C = 13</td>
<td>V = 10</td>
<td>A = 12</td>
<td></td>
</tr>
<tr>
<td>LV</td>
<td>115</td>
<td>EDP = 8</td>
<td>88</td>
<td></td>
</tr>
<tr>
<td>Ao</td>
<td>115</td>
<td>75</td>
<td></td>
<td>95.1%</td>
</tr>
</tbody>
</table>

CARDIAC OUTPUT = 3.5 L/MIN.
CARDIAC INDEX = 2.5 L/MIN./M²

Abbreviation : SCV = superior caval vein, RA = right atrium, RV = right ventricle, PA = pulmonary artery, PC = pulmonary capillary, LV = left ventricle, Ao = aorta, A = A wave, C = C wave, V = V wave

---

**Fig. 4.** The right ventriculograms in 20 degree right anterior oblique view in diastole (A) and in systole (B), demonstrating a huge right ventricle with its very poor motion, and poorly developed trabeculation. The tricuspid valve is located in normal position. But a significant tricuspid regurgitation is seen.

---

was equal to or exceeded the pulmonary arterial pressure in mid-diastole, consistent with the premature mid-diastolic pulmonic valve opening and the poor right ventricular compliance. A right ventriculography in the 20° right anterior oblique view (Fig. 4A, B) demonstrated a huge right ventricular cavity with a very thin wall (1 mm) and poorly developed trabeculation with poor motion in systole, and a significant tricuspid regurgitation. Although the tricuspid valve was in normal position, there was a prolonged opacification of the right ventricle. Therefore,
the diagnosis of Uhl's anomaly was established. A left ventriculography showed a normal-sized left ventricle with a good motion. Selective coronary angiography in multiple views was normal.

**DISCUSSION**

M-mode echocardiography in Fig. 1 clearly demonstrated the mid-diastolic premature full opening of the pulmonic valve before the atrial contraction with earlier opening in inspiration than in expiration. This finding, however, was not yet reported in this anomaly to the best of our knowledge, although French et al. reported a presystolic pulmonic valve opening due to an atrial kick. The hemodynamic data of our case in Fig. 3 that the right atrial and ventricular diastolic pressure of dip and plateau type, indicating the poor compliance of the right ventricle, were equal to and/or exceeded the pulmonary diastolic pressure in the mid-diastole, verified the premature mid-diastolic full opening of the pulmonic valve. The significant tricuspid regurgitation was also present in our case. Those are the mechanism of the premature opening of the pulmonic valve in our case?

Uhl's anomaly should be differentiated from Ebstein's anomaly, because both anomalies have similar clinical pictures.

Ebstein's anomaly is known to have several characteristic echocardiographic findings such as delayed tricuspid closure, better tricuspid valve recording far to the left of the normal position, sustained abnormal anterior position of the anterior tricuspid leaflet during the entire diastole with increased amplitude of the motion of the leaflet and late tricuspid opening. The delay of the tricuspid closure, more than 0.03 second after the mitral closure is thought to be the consistent finding with Ebstein's anomaly. R. G. Williams et al. considered that a tricuspid valve closure delay by as much as 0.06 second or more after mitral closure would be probably specific for the Ebstein's anomaly. However, two previous descriptions of echocardiogram in Uhl's anomaly reported 0.07 second delay in one case with the complete right bundle branch block and 0.04 second delay in another case with normal QRS complex. Our case showed only mild tricuspid closure delay (0.03 second) with incomplete right bundle branch block (Fig. 2). Therefore the delay of the tricuspid closure may not be useful in differentiation between these two cardiac anomalies. Ebstein's anomaly cases usually show delay of the tricuspid opening from zero up to 0.06 second after the mitral opening. In our case, as in Fig. 2, the tricuspid leaflets opened 0.06 second earlier than the mitral. However, this early opening of the tricuspid valve in Uhl's anomaly was not mentioned by French et al. although his case apparently showed it. This early opening of the tricuspid leaflets can be easily understood from the hemodynamic data which revealed early equalization of the right atrial pressure to that of the right ventricle due to the severe tricuspid insufficiency. This finding would be very helpful in differentiating Uhl's anomaly from Ebstein's.

Two previous Uhl's anomaly cases and our case showed a good diastolic excursion of the anterior tricuspid leaflet followed by a normal early closing velocity irrespective of the tricuspid regurgitation (Fig. 2). On the other hand decreased early diastolic closure rate of that valve is commonly seen in Ebstein's anomaly.

Ebstein's anomaly sometimes shows leftward displacement of a prominent tricuspid valve echo which was not experienced in Uhl's anomaly cases as in our case.

**ADDENDUM**

When we finished writing this paper, two articles about echocardiographic findings of Uhl's anomaly were presented by Sugi, K. et al. and Wakamatsu, T. et al. at the 34th meeting of Japanese Association of Medical Ultrasonics. They discussed the usefulness of the cross sectional echocardiography in the diagnosis of Uhl's anomaly.

**REFERENCES**