A Case Report of Subaortic Stenosis

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As the cause of the obstruction of left ventricular outflow, supravalvular aortic, valvular aortic and subvalvular aortic have been well known. In the recent years, however, a number of reports have indicated that the fourth type of anatomical deformity might cause obstruction of left ventricular outflow.

This new lesion has been termed as "muscular subaortic stenosis" or "idiopathic hypertrophied subaortic stenosis" since the systolic contraction of hypertrophied septal muscle may narrow the ventricular outflow tract producing an obstruction within the ventricle. Since the first description by Brock, many reports appeared in foreign literatures but there has been no case report of muscular aortic stenosis in Japan up to the date.

The diagnosis of muscular subaortic stenosis is apt to be missed clinically since the physical signs of this condition have a superficial resemblance to the aortic stenosis, mitral insufficiency and ventricular septal defect without the aid of heart catheterization and selective angiocardiography. The most reliable clue for the diagnosis of this condition is the catheter-withdrawal tracing and angiocardiogram of left ventricle.

In some cases, however, the routine right heart catheterization, angiocardiogram and brachial pressure curve may possibly make an accurate preoperative diagnosis. Lately, we have experienced a case of muscular subaortic stenosis in which an accurate diagnosis was made clinically with an aid of right heart angiocardiogram and brachial pressure tracing.

Case Report

A 7-year-old boy, was referred to the Heart Center of Kurume University, Medical School for the diagnostic studies because of a heart murmur discovered at the age of 7 months together with an enlarged heart. In spite of these findings, he had been asymptomatic except for some easy fatigability and susceptibility to cold. He was delivered by cesarean section due to breech presentation and inertia uteri. There was no contributory family history for any type of heart disease. During the first 7 months of his life, no medical record concerning heart murmur was available.

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He had been followed up by a local medical doctor with a tentative diagnosis of ventricular septal defect. There was no history suggesting rheumatic fever in the past.

Physical examination on admission revealed rather poorly developed boy without any acute distress. His mental status was normal.

The height was 114 cm. and weighed 19 Kg. No cyanosis or clubbing of fingers was noticed. There was slight bulging of thorax in the left precordial region. Pulse was regular, with a rate of 90. Axillary temperature was 37.2°C. Blood pressure was 108/64 mm.Hg. on both arms. There was left ventricular lift. Tonsils were normal. A grade III to IV systolic murmur was audible over the entire precordium, especially along the left sternal border but no systolic thrill; it was loudest at the fourth intercostal space and apex, and slightly transmitted to the neck. At the apex the second heart sound was split.

![Fig. 1. Phonocardiogram at apex.](image)

- Cycle: 140 c/s
- Amplitude: 1/2

There was neither diastolic murmur nor systolic ejection click over the aortic valvular region.

Complete blood cell count revealed that RBC was $484 \times 10^4$, Hb 12.8 Gm./100 ml. and WBC $126 \times 10^8$. Circulation time was 7 sec. in arm to lung and 11 sec. in arm to tongue. Urinalysis was negative. Wassermann reaction was also negative. Kidney and liver functions were normal. Sedimentation rate was 0 mm./1 hr. and 3 mm./2 hr. and CRP gave negative result.

The roentgenogram of the chest revealed a generalized cardiac enlargement with left ventricular preponderance.

There were neither calcification of aortic valve nor poststenotic dilatation. The electrocardiogram showed a regular sinus rhythm followed by occasional WPW complex. A definite left ventricular hypertrophy with ST-depression in left precordial leads were seen with upright T waves. Inverted T waves were seen only in lead I and aVL. The presence of an increased amplitude and duration of P wave and persisted small S wave in the left precordial leads which suggest some overloading of the both left atriums and right ventricle. From these findings a
tentative diagnosis of ventricular septal defect associated with some mitral regurgitation was made.

Subsequent laboratory data demonstrated that the antistreptolysin-O titer ranged 833 Todd units, but numerous determinations of C-reactive protein, BSR and WBC were within normal limits. Because of an increased ASLO the patient was immediately placed on both penicillin and steroid hormone with a probable diagnosis of acute rheumatic fever. This high ASLO titer, however, returned to the normal range within a month. In this period his general condition, including physical and other laboratory findings, remained completely unchanged. The first right heart catheterization was performed under the local anesthesia. During the procedure, various types of arrhythmia suddenly developed when the tip of catheter entered into right atrium. A further insertion of catheter into pulmonary artery produced shock like condition and we were forced to give up the procedure. An ECG at this time showed a pattern of right bundle branch block with occasional WPW complex. About 2 hours later, an ECG changed from LBBB to bigeminy
Fig. 3. Electrocardiogram on admission. Bigeminy of WPW syndrome complex is seen only in V1.

Fig. 4. Electrocardiogram about 6 weeks after first catheterization. Continuous bigeminy of WPW syndrome complex is present.
of WPW complex which lasted for a few days and then disappeared gradually in the course of 2 months. The second right heart catheterization and angiocardiogram were carried out under the general anesthesia. No evidence of any circulatory shunts was discovered as shown in Table I. Brachial pressure curve, however,

Fig. 5. Electrocardiogram on discharge.
A single beat of WPW syndrome complex only in lead I.

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<thead>
<tr>
<th>Table I. Results of Catheterization</th>
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<td>Pressure (mm.Hg)</td>
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<td>Systolic/Diastolic</td>
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<td>Pulmonary wedge</td>
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<td>Pulmonary artery</td>
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<td>Right atrium</td>
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<td>Vena cava sup.</td>
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<td>Vena cava inf.</td>
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<td>Brachial artery</td>
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<td>Oxygen uptake</td>
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<td>Cardiac index</td>
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Fig. 6. Brachial pressure curve demonstrates the initial sharp peak in early systole and tidal wave followed by the dicrotic notch (ND).

Fig. 7. Anteroposterior projection angiocardigrams.

a. During diastolic phase
b. During mid-systolic phase
c. During end-systolic phase
demonstrated a characteristic pattern that was called the “percussion” and “tidal” wave by Brachfeld and Gorlin. The dicrotic notch was also noted.

A catheter withdrawal curve from pulmonary artery was within normal limits. A routine angiocardiogram with right atrial injection clearly demonstrated generalized thickening of the left ventricular wall and a large muscle mass protruding into the left ventricular cavity from the inferior aspect of the ventricular wall; namely a large muscle mass which appeared practically to divide the left ventricle into 2 chambers. As shown in Fig. 7c, the ventricular outflow tract appeared to be “an inverted corn”, with its base at the aortic valve during systole. At the same time there was a considerable regurgitation of contrast material into an enlarged left atrium. These characteristic findings made it possible to exclude ventricular septal defect or simple type of rheumatic mitral insufficiency.

Thus a diagnosis of idiopathic muscular subaortic stenosis was made with possible recent rheumatic lesion.

After 4 months' hospitalization, he was discharged without any surgical operation since several surgical reports have suggested that there had been no successful method for treating muscular subaortic stenosis. For the past 7 months after his discharge, he has been asymptomatic and attending a school. A regular monthly check up showed his physical condition as well as laboratory data have remained unchanged except that short run of bigeminy of anomalous atrioventricular conduction complex developed frequently on exertion.

**DISCUSSION**

The present case offered some interesting points as follows; 1) This case was suspected clinically as ventricular septal defect. 2) The right heart angiocardiogram demonstrated a characteristic configuration of the systolic muscular stenosis of the outflow tract within the left ventricle. 3) The brachial pressure curve showed a typical pattern of a case with muscular subaortic stenosis. 4) ECG revealed alternate beats of normal and anomalous atrioventricular conduction, namely the bigeminy of WPW complex.

Braunwald and coworkers reported that in most instances with this anomaly, the diagnosis of ventricular septal defect or mitral insufficiency had been suspected clinically. Many authors emphasized that a selective left angiocardiogram constitutes an important diagnostic technique in the study of the patient with this disease. This technique makes possible to confirm the anomaly of the left ventricle during phases of the cardiac cycle.

Fortunately in our case, the systolic muscular stenosis of the left outflow tract was apparently visualized by a routine angiocardiogram. This typical picture is presumably caused by a considerable stagnation of contrast material within the ventricle due to systolic obstruction of the left ventricular outflow tract and mitral regurgitation.

Braunwald and his associates have also noted an association of mitral insufficiency in 11 of the 12 cases of muscular subaortic stenosis. They also
found that the mitral valve did not show any evidence of disease and the mitral insufficiency was attributed to a distortion of the orifice secondary to myocardial hypertrophy. It has been generally accepted that one of the most reliable criteria is the presence of a zone of reduced systolic pressure in the outflow tract of the left ventricle. As the catheter is withdrawn across the narrowed outflow area, the systolic pressure drops, but the diastolic pressure remains unchanged. When the aortic valve is traversed, the diastolic pressure rises but systolic level remains unaltered. This phenomenon, however, may be observed in a case with fibrous subaortic stenosis. On the other hand, in relation to the hemodynamic changes, the arterial pressure curve presents in most cases the initial sharp peak in early phase of systole and then the secondary, or tidal wave that will not be obtained in a case with ordinary aortic or subaortic valvular stenosis.

Braunwald and other authors emphasized the fact that anomalous atrioventricular excitation or WPW syndrome was observed not infrequently in cases with this disease. In our patient ECG revealed alternate beats of normal and anomalous excitation, which had returned occasionary to the normal rhythm. As shown in the tracing of ECG, despite the presence of a typical delta wave and pattern of anomalous atrioventricular conduction, the complex have never followed one after another. Therefore, it is conceivable that the complex may be ventricular premature contraction occurring at the terminal phase of P wave. So it remained unknown whether this anomaly of ECG is caused by the hypertrophied septal muscle alone or by complication with recent rheumatic lesion.

Recently Braunwald and his coworker suggested that the disease may be classified as follows; (1) the familial, non-congenital variety seen in patients of all age groups. (2) the non-congenital variety seen in adults without evidence for any familial association. (3) the non-familial congenital variety.

In our case there was neither family history suggesting cardiac disorder nor unexplained sudden death, and a careful examination of his family, including his parents, 2 siblings and some relatives, revealed no evidence of any heart disease.

The history of his heart murmur apparently indicates that his heart disease is primarily of the non-familial congenital variety, although the higher titer of ASLO may be an indicative of complicated recent rheumatic lesion. Glycogen-storage disease of the myocardium with obstruction to left ventricular outflow reported by Ehlers and his coworkers should be etiologically differentiated from this lesion. Usually patients with glycogen-storage disease will not live more than first one or 3 years of life at the longest. In our case there was no sign of glycogen-storage disease.
Although the catheter withdrawal study is needed to identify the presence of a systolic pressure gradient within the left ventricle as was in our case but we would like to emphasize the point that even the routine right heart angiocardiogram and brachial pressure curve may give the critical point to make an accurate diagnosis in some cases with this disease.

**Summary**

A case of muscular subaortic stenosis has been reported. This case was first considered to be a ventricular septal defect because of the history and a harsh systolic murmur heard over the entire precordium with cardiac enlargement. However, the routine angiocardiogram and brachial pressure curve demonstrated a characteristic pattern of muscular subaortic stenosis. As to the frequent appearance of ECG with short P-R interval, we have suggested that anomalous conduction which may be caused by complicated rheumatic lesion rather than hypertrophied septal muscle.

**References**