

Ventricular Septal Myectomy and Mitral and Aortic Valve Replacement in the Case of Discrete Subaortic Stenosis

Tetsuo HADAMA, Hidemi TAKASAKI, Yoshiaki MORI, Keiji OKA
Osamu SHIGEMITSU, Kiminori FUJISHIMA, Tatsunori KIMURA,
Sinji MIYAMOTO, Yuzo UCHIDA, Jyoji SHIRABE

Second Department of Surgery, Medical College of Oita

Received for publication, October 7, 1989

SUMMARY: We experienced a case of discrete subaortic stenosis of fibromuscular collar type, complicated with asymmetrical thickening of the ventricular septum and abnormal attachment of the tendinous cords of the mitral leaflet accompanied by mitral incompetence and aortic incompetence due to infective endocarditis. Surgical treatment comprised resection of subaortic stenotic fibrous tissues, ventricular septal myectomy and replacement of mitral and aortic valves. Preoperative pressure gradient of 120 mmHg across the left ventricular outflow tract was improved to 22 mmHg with a favorable clinical course. This was considered a rare case where fibromuscular type of DSS was accompanied not with secondary myocardial hypertrophy but with IHSS and abnormal tendinous cords of mitral leaflet.

INTRODUCTION

Subaortic left ventricular outflow obstruction is divided into the following: I-discrete subaortic stenosis (DSS), II-idiopathic hypertrophic subaortic stenosis (IHSS), III-atrioventricular valve anomaly depending upon stenotic morphology.¹⁾

Although they are respectively understood as an independent disease, they are not clearly classified embryologically. It is well known that some type of DSS is accompanied by left ventricular myocardial hypertrophy²⁾ and in some reports it is complicated with abnormal attachment of the tendinous cords onto the ventricular wall^{2)~5)}. Although it seems that myocardial hypertrophy increases with aging, it is interesting that three categories of left ventricular outflow obstruction such as DSS, IHSS, and abnormal attachment of the ten-

dineous chords are present in this case. Hemodynamically, aortic and mitral regurgitation are seen combined with left ventricular outflow stenosis, so that the aortic and mitral valve replacement were performed together with removal of the subvalvular fibrous ridge and septal myectomy. The outcome was favorable and satisfactory.

CASE REPORT

A 59-year-old woman was referred for the purpose of surgical treatment under the diagnosis of subaortic left ventricular outflow stenosis combined with aortic and mitral valve incompetence. She had cardiac murmur since about 25 years of age and had had sometimes syncope since about 45 years of age.

Recently she had begun to complain of chest pain and dyspnea on light exercise. A grade 4/6 ejection type murmur and grade 3/6 high

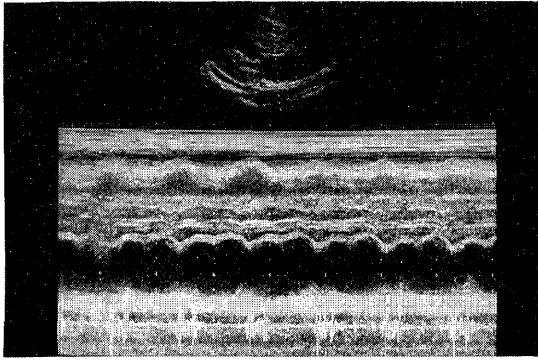


Fig. 1. Left ventricular outflow tract was severely stenotic with abnormal mass echo.

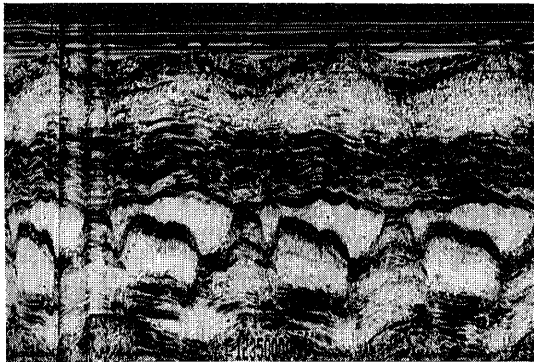


Fig. 2. M-mode echocardiogram showed ventricular septal hypertrophy and abnormal systolic anterior movement of the anterior mitral leaflet.

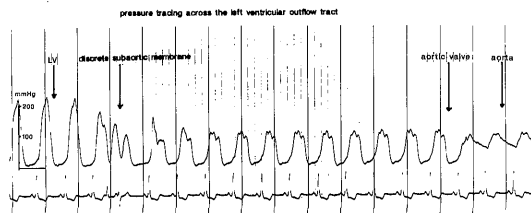


Fig. 3. Pressure tracing across the left ventricular outflow tract disclosed discrete type subaortic stenosis.

pitch diastolic murmur were heard in the 4th intercostal space at the left sternal border and a grade 3/6 holosystolic murmur in the apex. The chest X-ray revealed an enlarged left ventricle with CTR of 63% and no congestion in the lung field. Electrocardiography disclosed left axis deviation and left ventricular hypertrophy with a strain pattern but no arrhythmia.

Echocardiography revealed some characteristic findings. The aortic valve showed increased echo, systolic limitation of the opening, and diastolic separation. The left ventricular outflow was severely stenotic with abnormal mass echo. (Fig. 1) Ventricular muscle was hypertrophic, particularly the septal muscle was marked. Mitral leaflet echo was increased and abnormal tendinous cords were attached to the ventricular septum anteriorly and that in systole it was deviated forward attaching to the septum (SAM). (Fig. 2) Cardiac catheterization disclosed a left intraventricular pressure of 220/14 mmHg with a peak systolic pressure gradient of 120 mmHg across the left ventricular outflow tract. The ascending aortic pressure was 100/55 mmHg and pressure tracing showed a DSS pattern. (Fig. 3) Left ventricular cineangiography revealed a membranous substance in the outflow tract and anterior movement of the mitral leaflet toward ventricular septum in systole which exaggerated left ventricular outflow stenosis. The aortic valve and the mitral valve showed regurgitation grade 2 and 3 by Seller respectively.

SURGERY

The pericardium was opened through a median sternotomy and total cardiopulmonary bypass was established and systemic temperature was lowered to 25°C. Aortic cross-clamp was followed by a transverse incision of the aortic root. The aortic valve was thick and fragile in the three cusps and veruca was attached. These findings were considered to be due to valvular degeneration caused by abnormal blood flow over an extended period of causing DSS and also due to additional bacterial endocarditis resulting in aortic incompetence. Resection of the diseased valve revealed a stenosis with a thick ringular and fibrous ridge about 1 cm below the annulus extending toward the mitral leaflet and ventricular septum with white fibrous tissues. The stenotic outflow tract was enlarged by resection of the thickened septal myocardium according to Morrow⁶⁾ together with the fibrous ridge. The left atriotomy was performed to remove the mitral valve due to the consideration that valve replacement was necessary for

complete resolution of the left ventricular outflow stenosis and mitral incompetence, but it was impossible as the anterior leaflet and mitral annulus were strongly pulled toward the septal side so that the mitral component could not be seen. Thus, the anterior mitral leaflet and tendinous cords were removed through the aortic side. A part of the tendinous cords on the anterior leaflet was attached to the ventricular septum anteriorly. The aortic and mitral valves were replaced with 21 mmSt. Jude Medical valve and a 25 mm Medtronic-Hall valve respectively. The postoperative course was favorable with improvement of the left ventricular outflow pressure difference of 22 mmHg by the continuous wave Doppler (CWD) method. Her symptoms, such as exertional dyspnea, angina, and easy fatigability were greatly lessened.

DISCUSSION

DSS is a congenital disease which has a stenosis due to a crescent-shaped shelf or a ringular ridge of fibrous tissues in the left ventricular outflow tract apart from the aortic annulus. This form of stenosis accounts for 8-30% of congenital left ventricular outflow tract obstruction⁷⁾. DSS was first reported by Chevers⁸⁾ in 1942 and surgically repaired by Lillehei⁹⁾ and associates in 1962. Van-Praagh¹⁰⁾ explained that DSS results from maldevelopment of endocardial cushion tissue in the atrioventricular canal which forms the anterior leaflet of the mitral valve. These subaortic stenosis may occur alone or in association with other lesions. In some of DSS cases, as with our case, an abnormality that the tendinous cords of the mitral leaflet and papillary muscle attach to the anterior wall of the left ventricle or ventricular septum was reported in addition to subvalvular ring stenosis^{2)~5)11)12)}. The presence of these abnormal cases favors the developmental explanation of Van-Praagh¹⁰⁾. Pathologically and angiographically, three types have been described; the most localized type with thin membrane, the fibromuscular collar or ridge type with considerable muscular hypertrophy and the most severe tunnel variety assuming long segment narrowing²⁾¹³⁾¹⁴⁾. As

concerns for surgical treatment of DSS, membranectomy is considered sufficient for stenosis with membranous diaphragm, whereas for stenosis with fibromuscular ridge, some authors insist that the resection of the fibrous tissues alone is enough while some require myectomy according to Morrow⁶⁾. Reis²⁾ described that in the case of membranectomy alone sometimes pressure gradient persists because of retained secondary muscular hypertrophy immediately after operation, but it was always resolved with time and did not require additional surgical procedure. Ashraf¹⁵⁾ reported similar views that complete excision of the fibrous ring reduced the recurrence rate of subaortic stenosis and additional myotomy does not alter the outcome. He stated the operative technique had no relations with the late results. On the other hand, many researchers have recommended complete removal of fibrous stenotic tissues and additional myotomy or myectomy in cases with myocardial hypertrophy on the bases of the report that pressure gradient developed in a late stage even in cases of the membranous type treated by membranectomy alone or in cases where the fibromuscular ridge was considered to have been removed sufficiently^{16)~18)}. Lavee¹⁸⁾ reported that the late result was favorable in the group with myectomy in addition to membranectomy. Although many researchers agree that myectomy is necessary for the type of tunnel stenosis, in severe cases of this type in particular aortoventriculoplasty¹⁹⁾ or apico-aortic bypass²⁰⁾ are operative procedures of choice. In the case of IHSS myectomy is generally performed, Cooley²¹⁾ reported on the efficacy of mitral valve replacement in this disease. However, even in some cases of fibromuscular type of DSS we recommend mitral valve replacement combined with complete resection of fibromuscular stenotic tissues and septal myectomy. DSS is called fixed subaortic stenosis on the ground that the left ventricular outflow obstruction does not change depending upon the cardiac cycle and physiological variables including exercise loading or isoproterenol loading, whereas IHSS is understood to be dynamic obstruction²⁾. However, the fact^{17)18)22)~24)} that there are recurrence of left ventricular outflow obstruction in

cases of membranectomy alone or of insufficient resection of the stenotic site for fibromuscular stenosis and hemodynamic characteristics of tunnel obstruction prevents us from considering it appropriate that DSS is the fixed subaortic stenosis. It is of interest whether this left ventricular hypertrophy is secondary to DSS or isolated disease namely IHSS which is incidently complicated to DSS. Many researchers agree that hypertrophic stenosis developing in a postoperative course of DSS is secondary. Secondary hypertrophic stenosis differs from IHSS. In the secondary case the free wall of the left ventricle and septum is thickened asymmetrically more than the free wall²⁵⁾. Mitsuishi²⁶⁾ reported on a pediatric case of membranous type of DSS complicated with IHSS showing the presence of the case where myocardial hypertrophy is not secondary. In our case, the echocardiography disclosed asymmetrical thickening of ventricular septum and systolic anterior movement of anterior mitral leaflet which are compatible with the diagnostic criteria of IHSS. That is, the peculiarity of our case is that DSS due to a fibromuscular ridge coexists with ventricular hypertrophy which is diagnosable as IHSS and abnormality of tendinous cords of the mitral leaflet which is considered to be caused by anomalous atrioventricular canal resulting in mitral incompetence. Another complication of aortic incompetence is considered to be due to infective endocarditis. These anomalies indicated the necessity of removal of the fibromuscular ridge, ventricular septal myectomy, mitral valve replacement, and aortic valve replacement for complete correction hemodynamically. We report here that some cases of DSS are accompanied by abnormal attachment of the tendinous cords of the mitral leaflet and thickened ventricular septum requiring mitral valve replacement.

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