CASE REPORT

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Modified Konno Procedure for left ventricular outflow tract obstruction: report of two cases

Emre Oteyaka[1], Gizem Sari[2], Mehmet Turan Basunlu [2], Okan Eren Kuguoglu[1], Yılmaz Yozgat[2], Murat Ugurlucan[3], Halil Turkoglu[1]

[1] Istanbul Medipol University, Faculty of Medicine, Department of Cardiovascular Surgery, Istanbul, Türkiye
[2] Istanbul Medipol University, Faculty of Medicine, Department of Pediatric Cardiology, Istanbul, Türkiye
[3] Biruni University, Faculty of Medicine, Department of Cardiovascular Surgery, Istanbul, Türkiye

Abstract

Left ventricular outflow tract obstruction with a systolic anterior motion of the mitral valve is a challenging pathology. An intervention, either surgically or with a percutaneous technique is taken into consideration when the pressure gradient reaches critical levels. Subaortic myectomy is still the gold standard treatment modality in these particular patients. Modified Konno procedure may be added to the procedure when a significant gradient persists following subaortic resection, because of its association with increased outflow tract obstruction relief.

We herein present our experiences with two cases that underwent modified Konno procedures for the treatment of hypertrophic obstructive cardiomyopathy.

Keywords: Left Ventricular Outflow Tract Obstruction, Hypertrophic Obstructive Cardiomyopathy, Surgical Treatment, Modified Konno Procedure

INTRODUCTION

Left ventricular outflow tract (LVOT) obstructions that occur at the subvalvular level are divided into two groups. The first group consists of congenital discrete subvalvular aortic stenosis. This particular obstruction is localized by fibrosis, or a fibromuscular membrane, and may involve the entire subvalvular region in the form of a fibromuscular tunnel. The second group presents as hypertrophic obstructive cardiomyopathy consisting of a dynamic obstruction at the subvalvular level. Gold standard treatment option for both groups is still surgical muscular resection although percutaneous ablation methods are also attempted in selected cases. Unfortunately, the pathology is associated with increased incidence of recurrence and/or incomplete resection revealing significant persisting left ventricular outflow tract gradient (1).

The modified Konno procedure is an alternative option which may be added to the surgical subaortic muscle resection to provide better outflow. It is indicated in patients with diffuse subaortic stenosis consisting of a normal aortic orifice in patients with severe forms of hypertrophic obstructive cardiomyopathy and children with tunnel subaortic stenosis (2). Although

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Corresponding Author: Emre Oteyaka Istanbul Medipol University Faculty of Medicine, Department of Cardiovascular Surgery, İstanbul, Türkiye.
Email: eoteyaka@gmail.com
ORCID iD: 0000-0001-5889-2257

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the procedure is challenging and requires expertise, it allows adequate relief of diffuse subaortic obstruction, significantly decreases pressure gradient.

In this manuscript, we present our experiences with modified Konno procedure on two cases to treat hypertrophic obstructive cardiomyopathy.

Patient I

A 7-year-old male patient with chest pain, exertional dyspnea, and fatigue had been referred to our institution. In his history, there surgical treatment for subaortic stenosis at the age of 2.

The patient weighed 28,4 kg and was 142 cm tall with a heart rate of 84 beats per minute. Body surface area was calculated as 1.24 m^2. On physical examination, S1-S2 were rhythmic, 4-5/6 systolic murmur at the mesocardiac focus was heard, femoral arterial pulse was symmetrically palpable, respiratory sounds were normal and a sternotomy incision mark was present from previous procedures. Blood pressure of the patient was 102/54 mmHg. The electrocardiogram showed an increase in voltage in the chest leads, along with non-specific abnormalities in the ST segment and T-waves. Additionally, deep, narrow Q waves were observed in the lateral leads I, aVL, V5-6. The cardiothoracic index was increased on plain chest X-ray. Echocardiography indicated left ventricular outflow tract obstruction due to interventricular septal hypertrophy and systolic anterior motion of the mitral valve with an asymmetric septal hypertrophy of 29 mm and there was a pressure gradient of 100 mmHg. The aortic valve was tricuspid and there was no stenosis. The patient was scheduled for a surgical treatment following the consent of family after being informed about the risks and benefits in details.

Patient II

An 18-year-old female patient with the diagnosis of idiopathic hypertrophic obstructive cardiomyopathy was referred to our institution. Her history revealed septal myectomy and intracardiac defibrillator implantation 3 years ago. She complained of chest pain, exertional dyspnea, and fatigue.

The patient weighed 79.2 kg and was 165 cm tall with a heart rate of 68 beats per minute. Body surface area was calculated as 1.91 m2. On physical examination, S1-S2 were rhythmic, 3-4/6 systolic murmur at the mesocardiac focus was heard, femoral arteries were symmetrically palpable, respiratory sounds were normal, an implantable cardioverter-defibrillator pocket was present on the left pectoral area, and a sternotomy incision mark was present from the previous procedure. Blood pressure of the patient was 120/70 mmHg. Electrocardiography indicated a normal sinus rhythm, left bundle branch block with a QRS of 170 msn. The cardiothoracic index was increased on plain chest X-ray. Echocardiography indicated LVOT obstruction due to interventricular septal hypertrophy of 3.5 cm, moderate mitral valve insufficiency, a mild tricuspid insufficiency, and the outline of an implantable cardioverted-defibrillator lead. The aortic valve was tricuspid and there was no stenosis. A Valsalva manoeuvre provoked pressure gradient of 75 mmHg was detected. The patient with IHSS who an operative candidate was severely symptomatic was not significantly improved by optimal nonoperative treatment. Thus, the patient was operated after being informed about the risks and benefits of the procedure and following her consent.

Surgical Technique

The operations began with a redo midline sternotomy. The Aorta, both cavae, pulmonary artery, right ventricular outflow tract was prepared. Extracorporeal circulation was initiated after aortic and bicaval cannulation. Cardiac arrest was achieved at 32°C with infusion of antegrade hypothermic cold blood cardioplegia. Left ventricular outflow tract was inspected through aortotomy. The hypertrophied bands were resected. The septum was very hypertrophic and we thought the resections have not been sufficient. A transverse right infundibulotomy was also made long the right ventricular outflow tract (Figure 1). Further myectomy was performed. The septum was severely hypertrophied till the apex. A ventricular septal defect was created with linear incision till the level of the papillary muscles (Figure 2). The ventricular septal defect was closed with an oval shaped PTFE patch (Modified Konno), (Figure 3). The infundibulotomy was repaired with another piece of PTFE patch (Figure 4).

Intraoperative SAM evaluation was performed with TEE and temporary pacemaker was implanted in both patients. Operations were finalized uneventfully. Cardiopulmonary bypass and cross clamp times were 84 minutes and 62 minutes, respectively in the first patient and corresponding values were 95 minutes and 76 minutes, respectively in the second patient. Postoperative transaortic gradients were maximum of 25mmHg in the first patient and insignificant (none) in the case. Trivial to mild degree of mitral insufficiency was detected in patients after the operation. Both patients were transferred to the intensive care unit, weaned off ventilator in 6 hours, taken to the ward the next day and discharged from the hospital in 6 days. They have been followed asymptomatic for more than 6 months. The first patient was scheduled for and implantable cardiac defibrillator.

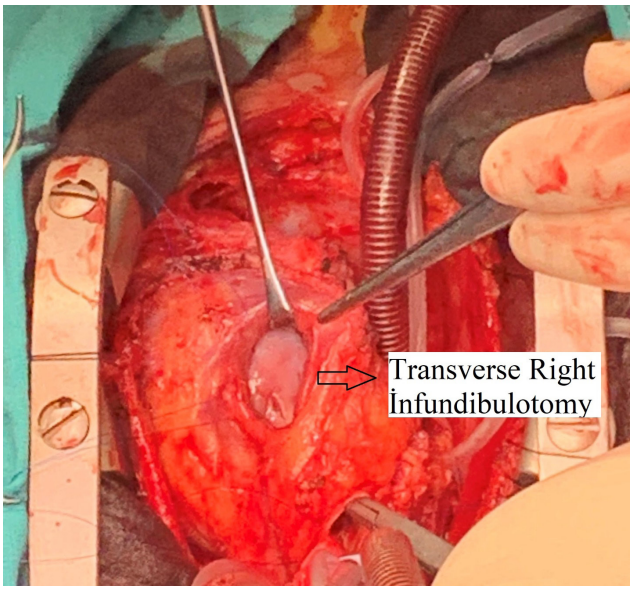

Figure 1: Transverse right infundibulotomy

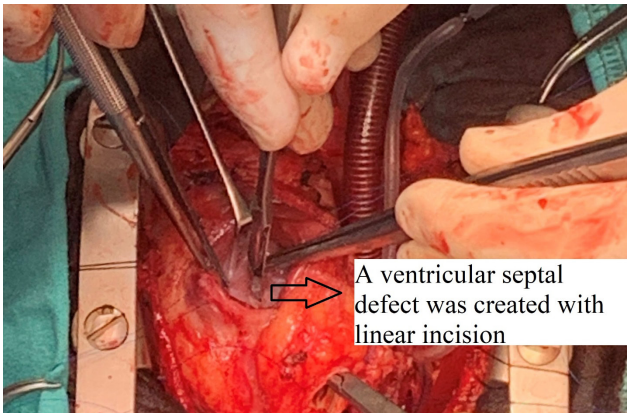

Figure 2: A ventricular septal defect was created with linear incision

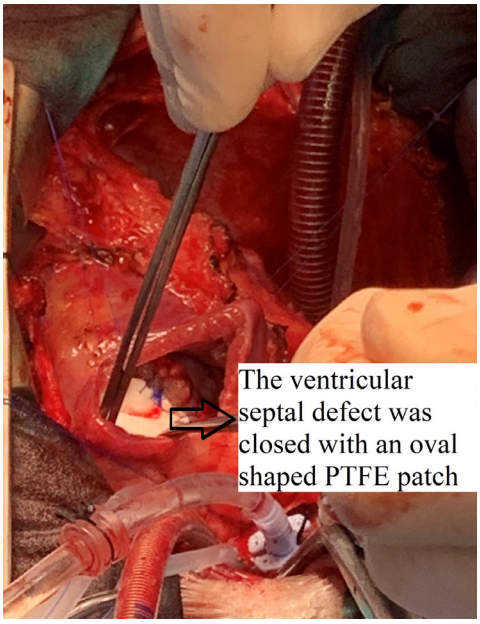

Figure 3: The ventricular septal defect was closed with an oval shaped PTFE patch

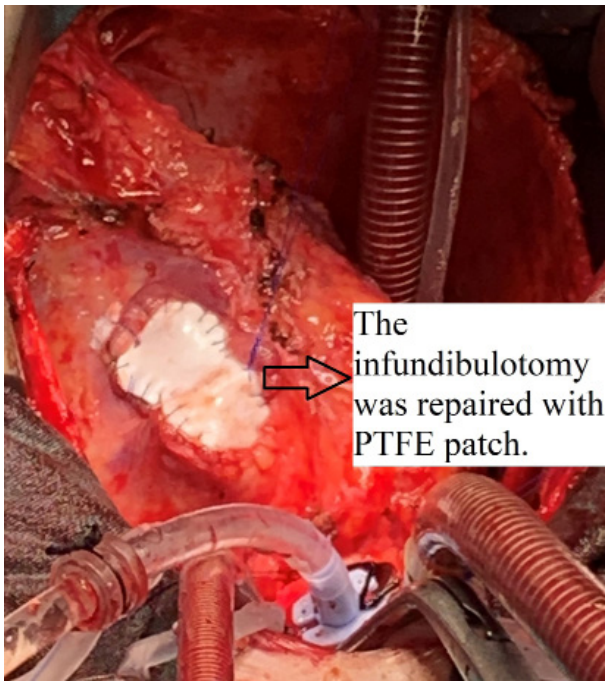

Figure 4: The infundibulotomy was repaired with PTFE patch

DISCUSSION

Idiopathic hypertrophic subaortic stenosis is an intense ventricular hypertrophy which is characterized with myocardial fibrous tissue disorders concentrated in the ventricular septum leading to dynamic obstruction in the subaortic region (3). Left ventricular outflow tract obstruction in this pathology is related with both systolic anterior motion of the anterior leaflet of the mitral valve and the hypertrophied interventricular septum.

Left ventricular outflow tract obstruction leads to acute decline of cardiac output, elevated left ventricular filling pressures, and myocardial ischemia, which can present with symptoms of chest pain, exertional dyspnea, presyncope, and syncope (4). Approximately one-third of patients with hypertrophic obstructive cardiomyopathy have left ventricular outflow tract obstruction at rest which is defined when gradients ≥30 mmHg. Another one third may present without outflow obstruction at rest; however, become symptomatic when provoked with physiologic and pharmacologic interventions that decrease left ventricular end-diastolic volume or increase left ventricular contractility such as during Valsalva maneuver or on exertion (<30 mmHg at rest and ≥30 mmHg at stress conditions). This is also known as latent left ventricular outflow tract obstruction. The third group of patients possess nonobstructive form of hypertrophic obstructive cardiomyopathy having gradients <30 mmHg at rest and stress. Marked gradients of ≥50 mmHg, either at rest or with stress represent the conventional threshold for surgical or percutaneous intervention if symptoms cannot be

controlled with medical measures (5). The treatment method of LVOT due to hypertrophic cardiomyopathy is achieved mainly through subaortic myectomy. When significant LVOT pressure gradient cannot be obtained with simple septal muscular resection, the modified Konno procedure (subaortic ventriculoplasty) which was first described by Cooley and Garrett in1986, provides excellent LVOT obstruction relief. This complex procedure is conducted to relieve tunnel or complex subaortic stenosis while preserving the aortic valve. With this technique, an extensive subaortic interventricular septum resection is performed, creating an artificial ventricular septal defect. The ventricular septal incision during modified Konno procedure should reach level of the papillary muscle level to overcome the other component of LVOT obstruction, the systolic anterior motion of the mitral valve. The artificially created ventricular septal defect is usually closed with a patch which can simply bulge away from the subaortic area, creating a spacious environment and allowing smooth flow from the left ventricle outflow tract. On the other hand, one of the disadvantages of the ventricular septal defect patch is right ventricular outflow tract (RVOT) obstruction. Hence, RVOT incision also requires special care during closure.

The modified Konno procedure is designed to address aortic and subaortic issues while minimizing the potential for aortic valve problems. Creation of an artificial ventricular septal defect poses certain risks such as injury to the mitral and the aortic valve as well as the conduction system. Therefore, the area of safe resection becomes limited. Aortotomy and right infundibulotomy is performed during the procedure to carefully inspect and consider the septum before resection. Resection of the septum adjacent to the aortic valve annulus is performed (6).

Cooley performed a simple resection and myectomy in a patient with subaortic stenosis; however, due to the recurrence of the case, decision to conduct the method known today as the modified Konno procedure was executed. However, he believed that following a ventriculotomy, a change in the morphology of the ventricle would lead to a RVOT obstruction, therefore placed another patch to reconstruct the RVOT (7). In our cases we performed the same procedure as Cooley, postoperative measurement of pressures indicated no signs of an obstruction, the operation was a success.

One other risk of modified Konno procedure is conduction disturbances related with septal myocardial resection and ventricular septal defect creation. The conduction tissue is generally located at the right side of the commissure between right and left aortic leaflets. The region of the conduction tissue is identified, located to the right of a line between the nadir of the right coronary aortic cusp and the septal attachments of the septal leaflet of the tricuspid valve. Resection at this level requires good cardiac anatomy knowledge, experience, and meticulous care (8). We did not experience conduction disturbances in our patients. However, we were more aggressive with resection in the second case whom already had an implantable cardioverter-defibrillator implanted and surgery resulted with nearly no LVOT gradient.

In IHSS, along with previously mentioned mechanisms, functional mitral insufficiency may also be seen. Whether it be an aortoventriculoplasty or a modified Konno procedure, since the left ventricular pressure decreases and the systolic anterior motion is resolved following a surgical treatment modality , functional mitral regurgitation becomes minimal or in some cases completely resolves (9). As mentioned, both of our patients who had mitral insufficiency prior to surgery, indicated no sign of mitral insufficiency following surgical treatment Whether it be an aortoventriculoplasty or a modified Konno procedure, since the left ventricular pressure decreases and the systolic anterior motion is resolved following a surgical treatment modality by intraoperative TEE. On the other hand, despite aggressive LVOT reconstruction with septal myectomy and modified Konno septal enhancement, mitral insufficiency and SAM may still persist. In such an occasion, mitral valve replacement may be an option. Another method may be simple edge to edge type repair of mitral valve which secures the anterior leaflet and prevents its systolic anterior subaortic displacement as well as subaortic obstruction (10).

In 1983, Vouhe and his colleagues, performed a similar surgery to that of a modified Konno procedure. To eliminate left ventricular outflow tract obstructions (especially IHSS and tunnel stenosis types), they performed a procedure they called the aortoseptal approach. In this method, a septal incision is made in the commissure between the aortic anulus and the right and left coronary cusps. The diffuse stenotic segment is resected, and the septum and opening of the aortic annulus is repaired. In addition, since the septum is primarily sutured without using a patch, the possibility of recurrence remains high (11). Hence, comparably, the modified Konno procedure seems more advantageous. Also, dual chamber pacing does remain a useful therapy for patients at very high risk for surgical or transcatheter therapy, or in whom these options are not available.

Following an idiopathic hypertrophic subaortic stenosis treatment with the modified Konno procedure, due to increased risk of arrhythmias and malignant fibrillation in such patients, the İmplantation of a cardioverted defibrillator is performed to prevent such complications. After postoperative rhythm evaluation, ICD insertion can be planned. One of our patients had an implanted cardioverter-defibrillator in place, however the other patient didn't, so we planned a placement of an implanted cardioverter-defibrillator.

In conclusion, the aim of treatment of hypertrophic

obstructive cardiomyopathy is to relieve subaortic occlusion. The most simple subaortic muscular resection may be associated with insignificant outflow tract relief and recurrence. In such cases modified Konno procedure seems a reliable alternative; however, technique is challenging and requires expertise and knowledge.

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The authors declare that they have no conflict of interests regarding content of this article..

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Ethical Declaration

Informed consent was obtained from the participant and Helsinki Declaration rules were followed to conduct this study.

Authorship Contributions

Concept: EO, GS, MTB, OK, YY,MU, HT Design: EO,OK Supervising: MU,HT Financing and equipment: MU,HT, Data collection and entry: EO,GS,MTB,OK,YY,MU,HT Analysis and interpretation: EO,OK, Literature search: EO,OK, Writing: EÖ, Critical review: GS,MTB.

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