Title: Video-assisted Transarterial Modified Konno Procedure with Concurrent Myectomy for Hypertrophic Obstructive Cardiomyopathy

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Conflict of Interest (COI) Statement: There is no conflict.

Funding Statement: There is no funding.

Central Message: Adequate and safe septal myectomy at the mid-LV level can be achieved with Video-assisted transarterial Modified Konno Procedure without ventriculotomy.

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Article word count: 744/750 words
Glossary of Abbreviations: cardiac magnetic resonance: CMR, hypertrophic obstructive cardiomyopathy: HOCM, left ventricle: LV, left ventricular outflow tract: LVOT, right ventricle: RV, right ventricular outflow tract: RVOT, ventricular septal defect: VSD.

Background:
Transaortic septal myectomy is the gold-standard treatment for patients with hypertrophic obstructive cardiomyopathy (HOCM). However, under particular conditions, such as severe and diffuse left ventricular outflow tract (LVOT) obstruction, the procedure is challenging and often results in inadequate muscle resection using only the transaortic approach, thus leading to residual stenosis. The modified Konno procedure is an effective alternative to relieve mid-left ventricle (LV) level obstruction in such patients. However, this procedure requires right ventriculotomy that potentially causes right ventricular outflow tract (RVOT) obstruction and arrhythmia. We describe a novel technique for the modified Konno procedure via aortotomy and pulmonary arteriotomy combined with endoscopy without incision of the LV or right ventricle (RV). (IRB/ERBCRB6180001, 23/03/2020; written patient consent was obtained for publication of this case report)
Clinical summary:

A 15-year-old girl (body weight: 45 kg, body surface area: 1.41 m²) presented with a systolic heart murmur. Echocardiography revealed a mean pressure gradient of 40 mmHg across the LVOT (Supplemental material 1). Computed tomography and cardiac magnetic resonance (CMR) imaging revealed long regional hypertrophied ventricular septum > 25 mm thick (Fig. 1). Given that the stenosis was tunnel-like and hypertrophy was severe, the modified Konno procedure was chosen. The surgery was performed via median sternotomy with cardiopulmonary bypass. After cardiac arrest, aortotomy was performed, followed by the main pulmonary arteriotomy. First, septal myectomy was performed through the aortic valve in the standard fashion. A ventricular septal defect (VSD) was cautiously created at the LVOT so as not to compromise the conduction system and, through the pulmonary valve, the VSD was enlarged. Additional myectomy was performed toward the apex at the level of the mid-portion. Abnormal tissues, including secondary chords between the papillary muscle and septum, were excised. During the procedure deeper inside the ventricle, endoscopy provided clear visualization to enable adequate release of the obstruction due to hypertrophied muscle and abnormal chordae (Video 1, Supplemental material 2). The VSD was closed using an expanded polytetrafluoroethylene patch. Postoperative echocardiography revealed a mean pressure gradient across the LVOT of 7 mmHg, with no aortic or pulmonary valve regurgitation, while CMR imaging revealed a wide open LVOT (Fig. 2). The patient has not experienced
atrioventricular block or life-threatening arrhythmia. The follow-up period is projected to be five years; currently, she has not experienced recurrence of stenosis and her symptoms are stable, with New York Heart Association functional class I.

Discussion:

HOCM is a standard criterion for transaortic septal myectomy. However, under specific conditions, such as severe and diffuse LVOT obstruction, small-size aortic annulus, the procedure often results in inadequate exposure of the LVOT, which results in inadequate resolution of the obstruction. This may lead to insufficient myectomy and unexpected injury to valves and the conduction system. In these situations, additional left ventriculotomy at the apex cannot avoid the possibility of damage to other structures as well. Cho et al. reported that inadequate muscle resection causes residual or recurrent stenosis, which accounts for 84% of reoperations. (2) In such situations, Laredo et al. reported on the efficacy of the modified Konno procedure and its favorable long-term results in high-risk patients with LVOT obstruction. (3)

However, the modified Konno procedure requires incision of the RV, and we believe this is unnecessary for patients without RVOT obstruction. With the use of endoscopy, only the transaortic and pulmonic approaches are sufficient to achieve adequate myectomy at the mid-LV level, in addition to the modified Konno procedure without right or left ventriculotomy. It potentially prevents RVOT obstruction and late arrhythmia caused by scar tissue. In addition
to the above, combining the direct field of view with endoscopy avoids valve and papillary muscle injuries.

Central Picture: The endoscopy provided clear visualization to enable adequate release of the obstruction due to hypertrophied muscle and abnormal chordae without additional incision of ventricles.

References


Figure and Tables

Figure 1: Preoperative cardiac magnetic resonance (A, B) and computed tomography (C) indicate long-regional left ventricular outflow tract obstruction. Red asterisk: left ventricle, black asterisk: aorta.

Figure 2: Postoperative cardiac magnetic resonance (A, B) shows the release of left ventricular tract obstruction. Red asterisk: left ventricle, black asterisk: aorta.

Video 1: This movie shows the video-assisted Transarterial Modified Konno Procedure.

Supplemental material 1: Echocardiography revealed that mean pressure gradient of 40 mmHg across the LVOT. A: systolic period, B: diastolic period.

Supplemental material 2: This schema depicts the video-assisted transarterial modified Konno procedure with concurrent myectomy without additional LV or RV incisions.