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## Original Article

## Hypertrophic Obstructive Cardiomyopathy Repair with Coronary Artery Bypass Surgery: A Case Report

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## ABSTRACT

**Background:** Hypertrophic obstructive cardiomyopathy (HOCM) is a cardiac condition of an abnormal interventricular septum (IVS) thickening. It is due to an autosomal dominant mutation in sarcomeric protein genes. It had many phenotypes with different distribution of myocardial hypertrophy. Septal myectomy remains the treatment of choice for hypertrophic obstructive cardiomyopathy resistant to medical therapy. However, controversy exist regarding standard treatment option(s).

**Description of the Case:** Here we described a case of 47-year-old patient admitted with diagnosis of acute coronary syndrome, with typical chest pain of angina class II, shortness of breath class-II (NYHA), smoker on regular renal hemodialysis. Echocardiography showed HOCM, IVS thickening, normal Left ventricle global systolic function and mild to moderate eccentric regurge. Coronary angiography revealed normal left main with total (LAD) proximal occlusion, proximal and mid (LCX) (50%) occlusion, first (OM) mid (90%) lesion, (RCA) dominant with non-flow limiting lesions. He was submitted to open heart surgery with four grafts, sub-valvular myomectomy, and successful myomectomy. Distal anastomosis was done first then myomectomy then proximal anastomosis. Patient extubated after (9) hours with stable hemodynamics and complete neurological condition, with accepted drains output. He gradually weaned from inotropic support and chest drains removed on 3<sup>rd</sup> postoperative day. All lines and pacemakers were removed, the wound was dry with stable sternum and improvement of all symptoms. Then discharged home after 9 days of hospital stay on medical treatment. Postoperative echocardiography showed significant improvement.

**Conclusion:** Surgical treatment is the standard option in treatment of HOCM to improve symptoms.

Keywords: Hypertrophic Obstructive Cardiomyopathy; Coronary Artery Bypass Graft; Systolic Anterior Motion; Left Ventricle.



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## INTRODUCTION

Hypertrophic obstructive cardiomyopathy (HOCM) is defined as a cardiac pathology that results in an abnormal thickening of the interventricular septum (IVS), with a disorganized array of cardiac myocytes on microscopic examination. It is due to autosomal dominant mutations in sarcomeric protein genes. It exhibits several phenotypic variations with different distribution of myocardial hypertrophy: asymmetric septal (classic HOCM), apical, neutral, concentric, and reverse septal<sup>(1,2)</sup>.

In most cases, HOCM has been classified as a non-obstructive disease, given that many patients are asymptomatic and do not show any abnormal left ventricular outflow tract (LVOT) gradient under basal conditions. LVOT obstruction (LVOTO), defined as a peak gradient >30 mmHg, defines the obstructive form of HOCM<sup>(3,4)</sup>.

Patients with HOCM are at increased risk of sudden cardiac arrest due to dynamic left ventricular outflow tract obstruction, myocardial ischemia and arrhythmias. Septal myectomy remains the gold standard therapy for patients with HOCM refractory to other treatment options<sup>(5)</sup>.

These structures can interact with the septum, causing systolic anterior motion (SAM) of the mitral valve (MV) leading to LVOTO and mitral regurgitation (MR). LVOTO in HOCM can vary from one patient to another depending on which elements are involved. For example, some patients presented with a thickened septum only. Others have abnormal length of the MV leaflet with the presence of SAM of the MV. Others present with PM hypertrophy in addition to IVS and/or LV hypertrophy<sup>(4,6)</sup>.

Controversy remains as to the optimal treatment of patients with HOCM, who remain severely symptomatic despite optimal medical therapy. Until recently, surgery was the only treatment available for these patients. Surgical revascularization of the left anterior descending coronary artery (LAD) leads to improvement of ventricular contractility and recurrence of the LVOT obstruction. Surgical septal myectomy is a standard option for patients with HOCM who are scheduled for septal reduction therapy<sup>(7,8)</sup>.

## DESCRIPTION OF THE CASE

A 47-year-old gentleman admitted to Sabah Al-Ahmed Cardiac Center at Al-Amiri Hospital in Kuwait as a case of acute coronary syndrome (ACS) (Non-ST-Elevation Myocardial Infarction (NSTEMI)). He was presented with typical chest pain of angina class II, and shortness of breath functional class (NYHA; New York Heart Association)-II, known as a smoker, with chronic kidney disease on dialysis.

Echocardiography showed HOCM with significant LVOT peak gradient was 71 mmHg at rest and increased to 86 mmHg with Valsalva maneuver. There was asymmetrical septal hypertrophy with septal thickness about 18 mm and posterior wall (PW) thickness 11 mm. The maximum LV wall thickness was

19 mm at mid-septum with normal global LV systolic function (EF~65-60%). The SAM of anterior mitral valve leaflet (AML) showed mild to moderate eccentric mitral regurgitation (MR) (Figures 1 and 2).

Coronary angiography showed normal left main with total LAD proximal occlusion, proximal and mid LCX (50%) occlusion, first (OM) mid (90%) lesion, (RCA) dominant with non-flow limiting lesions (Figure 3).

Open heart surgery (CABG) with four grafts was performed (e.g., (LIMA) to (LAD), SVG to (OM2), (RIMA) free graft to (OM1) and (SVG) to diagonal). In addition, sub-valvular myectomy one cm under right coronary cusp (RCC) using blade and cut area (3x2x1) cm using scale and left atrial appendage closure with clip 40 mm was performed. The left atrial appendage closure aimed to prevent thrombus formation in case of developing arrhythmias. The success of myectomy was assessed intraoperatively with transesophageal echocardiography. Distal anastomosis was done first then myectomy followed by the proximal anastomosis.

Surgery was completed under general anesthesia and cardiopulmonary bypass with deep hypothermic circulatory arrest three doses. The cross clamp time was 130 minutes, the mean total bypass time was 238 minutes, with easy weaning from bypass on adrenaline 50 ng/kg/min and noradrenaline 50 ng/kg/min.

Patient transferred to postoperative (ICU) and extubated after 9 hours with stable hemodynamics and complete neurological condition, with accepted drains output, and gradual weaning from inotropic support. The chest drain was removed on the 3<sup>rd</sup> postoperative day. Concurrently the removal of all lines and pacemaker cut was performed at the same day. The wound was dry with stable sternum and improvement of all symptoms as regard (NYHA) become class I and (CCS) became class I.

All results of laboratory investigations were within normal except high creatinine. A daily dialysis was done and patient was ambulated well with chest physiotherapy. The patient discharged without significant postoperative complication after 3 days in ICU and total hospital stay (9) days, on medical treatment with Plavix and aspirin and follow with nephrologist to continue dialysis after discharge.

Echocardiography was done six days after surgery showed non-significant (LVOT) peak gradient was 22 mmHg at rest increased to 42 mmHg with Valsalva maneuver with global (LV) systolic function is normal (EF~65-60%), Systolic anterior motion (SAM) of anterior mitral valve leaflet (AML) with mild to moderate eccentric mitral regurgitation (MR) (Figures 4 and 5).

Another follow up Echocardiography was done (39) days postoperatively showed non-significant (LVOT) peak gradient was 9 mmHg at rest increased to 13 mmHg with Valsalva maneuver with global (LV) systolic function is normal (EF~65-60%), and disappearance of mitral systolic anterior motion (Figures 6 and 7).

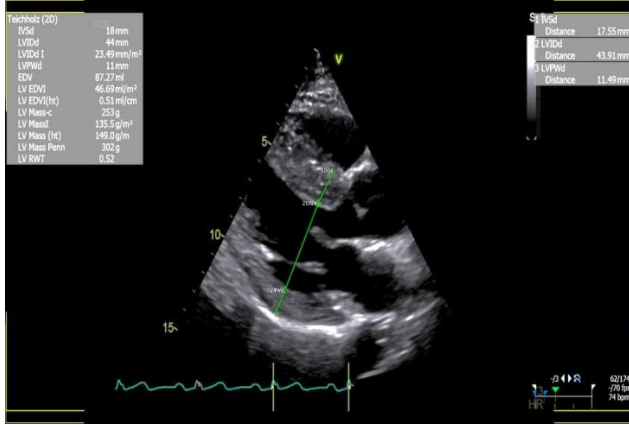


Figure (1): Preoperative LV dimensions

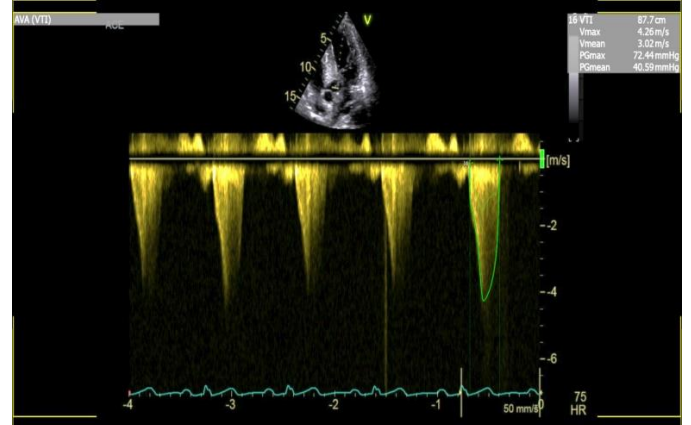


Figure (2): Preoperative LVOT gradient- Dragger shaped



Figure (3): Preoperative Coronary angiogram



Figure (4): Postoperative LV dimensions

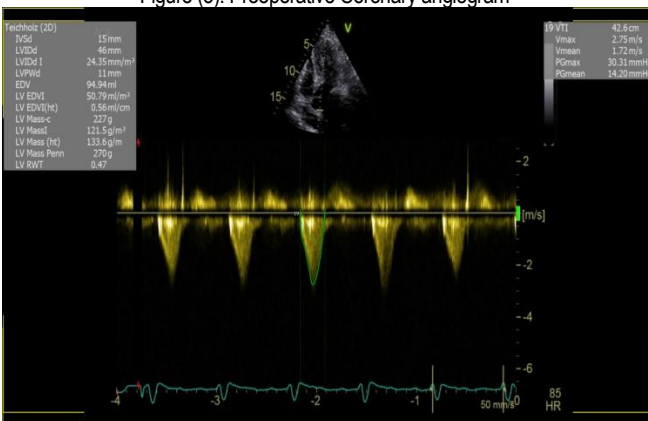


Figure (5): Postoperative LVOT gradient without Valsalva

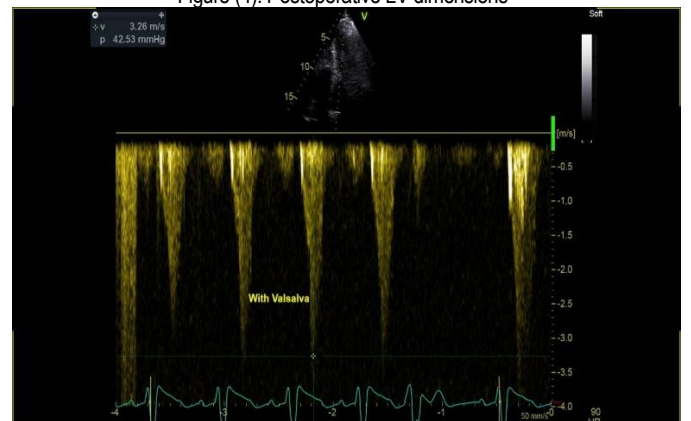


Figure (6): Postoperative LVOT gradient with Valsalva

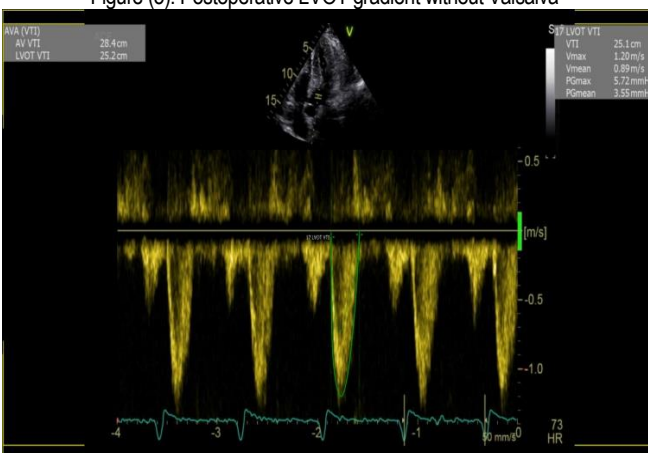


Figure (7): Late postoperative LVOT gradient

## DISCUSSION

Many studies talk about surgical myectomy in (HOCM) patients to improve symptoms, in Alireza *et al.* <sup>(9)</sup> study that conducted on Ninety-six patients treated with trans-aortic myectomy with acceptable early and late mortality rates. Improvements in functional status are seen in almost all patient's Long-term survival is excellent and cardiac sudden death is extremely rare after a good surgical treatment.

Patients are at risk of various perioperative complications from anesthesia and surgery due to the underlying complexity of their disease, but septal myectomy remains the gold standard therapy for patients with hypertrophic obstructive cardiomyopathy (HOCM) refractory to other therapy <sup>(6)</sup>.

In study done by Cui B, *et al.* <sup>(10)</sup> which occurs on seventy-six patients; the ventricular septal myotomy–myectomy can be performed successfully for the severe obstructive (HOCM) and mitral regurgitation (MR) with the low morbidity and mortality and excellent survival in the great majority of patients. But for the few patients with the intrinsic mitral valve disease, the concomitant mitral valve repair or mitral valve replacement (MVR) may be required, and (MVR) should be performed only as a priority choice for the inherent risks of prosthetic valves and anticoagulation therapy.

The benefits of mitral valve repair are better overall survival and a lower rate of thromboembolic events, in addition to extended myectomy are effective methods of surgical treatment in patients with hypertrophic obstructive cardiomyopathy <sup>(8)</sup>.

This case shows surgical treatment of (HOCM) in the form of myectomy of interventricular septum throw aortic valve below right coronary cusp with (CABG), this case associated with (SAM) which improved with proper repair of (HOCM) without intervention of mitral valve.

**Conclusion:** We conclude that surgical management is the base in treatment of (HOCM) to improve symptoms especially with associated other cardiac surgical interventions, more over mitral (SAM) usually improved after (HOCM) repair.

**Conflict of interest:** None

**Financial disclosure:** None

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