Hypertrophic obstructive cardiomyopathy (HOCM) is a rare progressive cardiac abnormality associated with dynamic systolic LVOT obstruction and diastolic dysfunction. In patients who remain symptomatic despite medical treatment transthoracic septal myomectomy or percutaneous septal myomectomy (in selected high risk patients) have been used for their surgical management. Transesophageal echocardiography (TOE) accurately characterizes the anatomic features and severity of hemodynamic obstruction, determines the coexisting mitral regurgitation, evaluate diastolic dysfunction and immediately demonstrates the surgical results of myomectomy. The surgical myomectomy involves the removal of portion of abnormal septum beneath the right coronary cusp.

The role of TOE in the surgical management of patients with HOCM is essentially a pre-pump and post-pump studies. The pre-pump study focuses on the diastolic thickness of the septum, the distance of the SAM-septal contact from the base of the right coronary cusp, the severity of LVOT obstructive gradient and the mechanism and severity of mitral regurgitation. To safely guide the surgical septal myomectomy the true septal diameter must be measured during diastole. The length of the SAM-septal contact and the distance of the right coronary cusp to the highest SAM-septal contact must be accurately measured.

The mechanism of mitral regurgitation in HOCM involves the distortion of the plane of coaptation produced by SAM of mitral leaflet. The result is systolic opening of the leaflets with a relative anterior override of the posterior leaflet and a characteristic mitral regurgitant jet posteriorly directed. If the jet is directed anteriorly or centrally other possible mechanism of mitral regurgitation should be investigated.

Diastolic dysfunction of the LV is commonly present in patients with HOCM and may include all four physiologic patterns. Although pharmacologic intervention, including lusitropic and chronotropic drugs, may assist in the management of diastolic dysfunction; yet the abnormalities commonly persists after correction of the systolic LVOT obstruction.

The post-pump study determines the success of the septal myomectomy evaluates for any potential complication and assess the competence of mitral valve. A persistent gradient across the LVOT or a provocative peak gradient of more than 36mmHg should be evaluated for further surgical myomectomy. If the septal thickness or the depth of the incision towards the apex will not permit further tissue removal, consideration is given to replacing the mitral valve. In addition to significant persistence of LVOT gradient, the most serious complications of septal myomectomy include an iatrogenic VSD, papillary muscle laceration and a coronary –ventriculo fistula resulting from incision across a left ventricular septal perforator. A septal thickness of less than 3-5 mm at point of septal myomectomy must be followed closely in the postoperative period to ensure that a VSD does not ensue.
References

