Selection of hypertrophic cardiomyopathy patients for myectomy or alcohol septal ablation

Hipertrofik kardiyomiyopatide miyektomi veya alkol septal ablasyonu için hasta seçimi

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ABSTRACT

This brief review summarizes how patients are selected for myectomy or alcohol septal ablation and reviews results for both procedures. The most recent literature is reviewed for both septal myectomy and alcohol ablation. The mechanisms for obstruction and mitral regurgitation as well as the indications for both procedures are reviewed. Septal myectomy gives a more consistent relief of the gradient with very low morbidity and mortality. The mortality for alcohol septal ablation is higher than for surgery. In addition, the need for a permanent pacemaker is higher for patients undergoing septal ablation. There maybe an increased risk for ventricular arrhythmia post ablation.

Septal myectomy is the gold standard for the invasive treatment of hypertrophic cardiomyopathy. Septal ablation should be considered for the elderly or patients with co-morbid conditions that would make surgery at increased risk. *(Anadolu Kardiyol Derg 2006; 6 Suppl 2: 27-30)*

Key words: Hypertrophic cardiomyopathy, myectomy, alcohol septal ablation

ÖZET

Bu derleme hipertrofik kardiyomiyopatide hastaların miyektomi ya da alkol septal ablasyonu için nasıl seçildiğini ve her iki işlemin sonuçlarını özetlemektedir. Septal miyektomi ve alkol ablasyonu ile ilgili en son literatür gözden geçirilmiştir. Obstrüksiyon ve mitral yetmezliğine yol açan mekanizmalar ve her iki işlem için endikasyonlar ele alınmıştır. Septal miyektomi çok düşük morbidite ve mortalite ile gradiyentte daha belirgin azalma sağlar. Alkol septal ablasyonunda görülen mortalite cerrahiden daha yüksektir. Ek olarak, septal ablasyonu uygulanan hastalarda kalıcı pil için gereksinim daha fazladır. Ablasyon sonrası ventriküler aritmi riski de artabilir. Sonuç olarak, septal miyektomi hipertrofik kardiyomiyopatinin invazif tedavisi için altın standarttır. Septal ablasyonu cerrahi açıdan yüksek riskli olan yaşlı ya da ek başka hastalıkları olan hastalarda düşünülmelidir. *(Anadolu Kardiyol Derg 2006; 6 Özel Sayı 2: 27-30)*

Anahtar kelimeler: Hipertrofik kardiyomiyopati, miyektomi, alkol septal ablasyonu

Introduction

Hypertrophic Cardiomyopathy is a disease of the heart muscle first recognized by Dr. Donald Teare (1) and Lord Russell Brock (2) in 1957. In some, but not all patients, it is genetically based. It occurs in 1 in 500 people (3). It can occur in young children as well as elderly people. Patients maybe asymptomatic or have severe symptoms of left ventricular (LV) outflow tract obstruction which includes shortness of breath, chest pain dizziness or syncope. The first symptom can be sudden death (4). The manifestations of the disease can be worse in the young than the elderly (5). Approximately 70% of patients suffer from left ventricular outflow tract obstruction at rest or with provocation (6). Obstruction is the result of a complex interaction between the septum and the mitral valve leaflets and its supporting structures, the chordae tendineae and the papillary muscles (7-9). In addition, many obstructed patients have significant mitral regurgitation due to systolic anterior motion of the mitral valve (10-12).

The first line of treatment for the obstructive form of the disease is medical treatment with either beta blocker (13-17), calcium channel blockers (verapamil or diltiazem) (18, 19) or disopyramide (20-23). When patients become refractory to medical treatment, invasive therapy is necessary. The two forms of invasive therapy are septal myectomy and alcohol septal ablation. The latter has also been called PTMSA (Percutaneous Transluminal Septal Myocardial Ablation) (24), TASH (Transcoronary Ablation of Septal Hypertrophy) (25), and NSRT (Nonsurgical Septal Reduction Therapy) (26).

Before considering, which invasive form of treatment should be used a thorough understanding of the mechanisms for outflow tract obstruction and mitral regurgitation is required.

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Mechanisms of Obstruction

Early in the history of the disease, it was proposed that left ventricular outflow tract obstruction was the result of Venturi forces being exerted on the leaflets by the rapid flow of blood through the narrow left ventricular outflow tract. This may be the case when the outflow tract is quite narrow as in older patients. These patients frequently also have calcification of the mitral annulus that may displace the mitral valve anteriorly towards the septum, resulting in marked narrowing of the outflow tract. In many patients, however, either because of large billowing mitral leaflets and/or anteriorly displaced papillary muscles, there are drag forces that result in anterior motion of the mitral valve (9, 27, 28). In addition, there are patients who have anomalous direct attachment of a papillary muscle to the anterior leaflet (29).

Mechanisms of Mitral Regurgitation

Mitral regurgitation may be the result of the systolic anterior motion (SAM) of the mitral valve leaflets or there may be intrinsic mitral valve disease. If the mitral regurgitation is related to SAM the mitral regurgitant jet is directed posterior and laterally (11). If the regurgitation is related to intrinsic mitral valve disease, the jet will be central or anteromedially directed. It is particularly important to know the mechanism of the regurgitation. When the regurgitation is related to the SAM then it will be improved or eliminated when the septum is surgically thinned. However, when there is co-existent mitral valve disease it will not be improved with septal reduction therapy alone. In addition to jet direction, the mitral valve anatomy must be completely visualized. This may require a transesophageal echocardiography if the transthoracic study is not ideal.

Surgery for Hypertrophic Cardiomyopathy

The gold standard for medically refractory disease is a septal myectomy. It has been done for more than forty years. Early results of surgery had operative mortality in the range of 8% (30). Subsequent surgical series have had a mortality of <2% (31,32). Smedira and colleagues, in 324 patients, had no hospital deaths (33). The results of surgery are clearly improved from earlier times because of better anesthesia, myocardial protection during cardiopulmonary bypass and intra-operative transesophageal echocardiography (34) The anesthesia and myocardial protection reduce the risk of ischemia during surgery. The intra-operative transesophageal echocardiography allows for the surgeon and the cardiologist to be sure that the septal myectomy and mitral valve surgery, if required, are adequate before the patient leaves the operating room. The latter has significantly reduced the need for re-operation. The series by Ommen and colleagues (31) showed that patients with successful myectomy live as long as the age-adjusted general population.

Alcohol Septal Ablation for Hypertrophic Cardiomyopathy

We feel that elderly patients, or those with severe co-morbid conditions who would be at increased surgical risk, should be considered for alcohol ablation. They should be NYHA function class III or IV with a gradient of 50 mm Hg at rest or with provocation. These would be patients, however, who have no co-existent valvular heart disease or coronary artery disease. We have considered patients for ablation because of morbid obesity, diabetes, chronic obstructive pulmonary disease or renal failure (35). In order for the ablation to be successful there must be a septal perforator of sufficient size and location to cause an infarction of the septum in the area of SAM - septal contact in order to reduce the left ventricular outflow tract obstruction. When the septum is less than 18 mm, intrinsic mitral valve disease should be strongly suspected. Usually this means the leaflets are elongated or anteriorly displaced. If the septum is greater than 25 mm the chance for successful, meaningful, septal reduction by alcohol will be reduced.

The first three cases of septal ablation were first described by Sigwart (36) after first recognizing that spontaneous myocardial infarction could reduce the left ventricular outflow tract obstruction. Over the past 11 years, greater than 3500 patients have undergone the procedure.

First, a coronary angiogram is done to insure there is no significant intrinsic coronary artery disease. A temporary pacemaker is placed in the right ventricle should the patient develop complete heart block. A transthoracic echocardiography is performed in the catheterization laboratory to define the location of SAM septal contact. Then a small catheter is placed in the left anterior descending coronary artery with the help of a guiding catheter. It is then usually advanced into the first septal perforator. The balloon is inflated. An echocardiography contrast agent is then injected to determine where the myocardial infarction will occur. It must be determined that the contrast only goes to the septum near the point of mitral-septal contact and not elsewhere; for example, not to the right ventricle, LV papillary muscles or in the LV free wall. Once it is determined that the catheter is in proper position, 1-3 cc of absolute ethyl alcohol is slowly injected down the catheter. As the alcohol is injected echocardiography is repeated and a very bright area is seen where the infarction will occur. The balloon is inflated for 10 minutes, then carefully removed. Usually, with initial balloon inflation the gradient drops significantly. It has been observed that at about five days the gradient goes back up and then slowly falls over the next few weeks to months. The initial drop is thought to be the result of initial stunning. The redevelopment of the gradient may be the result of edema in the infarcted area; then, the subsequent drop in the gradient may be the result of formation of scar and retraction of the infarcted area.

There has been early enthusiasm for septal ablation. It is clearly less invasive than surgery and many patients recover quickly. A number of studies have shown positive results in terms of symptom reduction. However, on closer scrutiny there are problems to be considered. The reported mortality with the procedure is higher than with a septal myectomy. There is less complete relief of the gradient and the need for permanent pacemaker is higher (25, 26, 37-39). In our experience the technique is successful 80% of the time (35).

In addition to these early studies there have been single center non-randomized studies comparing alcohol ablation to septal myectomy. All have shown better relief of gradient, lower inci-

dence of heart block and lesser mortality in the surgical group (35, 40). One study showed better exercise tolerance post myectomy (41). There also remains the worry of sudden death after ablation. The incidence is not clear, but there are individual case reports of sudden deaths after ablation. In many of the series at least one sudden death has occurred and sometimes more (42, 43, 44) There are also series of patients who were considered at high risk for sudden death who had an automated implantable cardioverter-defibrillator (AICD) implanted: the firing rate in the first year post ablation was high - 8% (45).

Advantages of a Septal Myectomy

The relief of the gradient is immediate and usually permanent. Need for a permanent pacemaker is less than 3% if there are no preexisting conduction abnormalities. Co-existing coronary artery disease and mitral valve disease can be dealt with. There is no scar that remains and the patient is thus at less risk for ventricular arrhythmia.

Disadvantages of a Septal Myectomy

Surgery requires an experienced surgeon, who may not be locally available. The risk is higher in the elderly patients. The incidence of post-operative atrial fibrillation is 22-30%. Mild aortic insufficiency has been reported after surgery. The recovery is longer with surgery than with alcohol ablation.

Advantages of Septal Ablation

The benefits of the ablation are that a major surgical procedure can be avoided. There is no post-operative pain and very little risk of infection or need for blood transfusion. The hospital stay tends to be shorter and therefore there is possibly less expense. There is a quicker return to daily activities. There is a very low incidence of post ablation atrial fibrillation.

Disadvantages of Septal Ablation

Variability in the location and size of the septal perforators is significant. This will, therefore, limit the number of potential candidates for the procedure. In one series of Seggeweiss 3% of the patients could not have ablation because of problems with perforator anatomy (38). Provocable obstruction may remain. The incidence of permanent pacemaker insertion is 10-23%. There remains the concern about the development of ventricular arrhythmia, and there have been reports of sudden death and AICD shocks, days to weeks after ablation.

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