Surgical treatment of hypertrophic cardiomyopathy (Düsseldorf experience)

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Summary: Surgical treatment appears to be the therapy of choice in hypertrophic, obstructive cardiomyopathy, at least in patients who are in the advanced stage of the disease and refractory to medical treatment. In addition, strong evidence has been obtained that surgical treatment improved the prognosis at least in patients who required only a myectomy via a transaortic approach.

Surgical techniques

Surgical treatment of hypertrophic obstructive cardiomyopathy started with the first successful operation performed in 1958 by W. Cleland in London (Bentall et al., 1965). The operation consisted of resection of septal myocardium.

Different approaches to the bulging hypertrophied ventricular septum have been used. Special techniques are reported for the transaortic, trans-left-ventricular and the left transatrial approach as well. A few surgical groups also used the trans-right-ventricular approach.

Following the concepts of Braunwald & Morrow (1960), anatomical pathophysiological studies established the importance of hyperactivity of the bulbo-spiral muscle. Simple myotomy using the transaortic approach seemed to be the safest method to most surgical groups. The incision was made from just below the commissure of the right- and left-coronary cusps of the aortic valve towards the apex of the heart. But after a few years experience, most surgeons preferred myocardial resection, sometimes with an additional myotomy. The transaortic approach became the method of choice (Schulte et al., 1981).

It must be mentioned that the resection should be as extensive as possible to give space enough for at least 2 cm², almost to the level of the origin of the anterior mitral papillary muscle, and that, in addition, the thickened endocardium should be removed. After the resection, one should pay attention to atypical chordae of the mitral valve. They may sometimes be an additional reason for the systolic anterior movement of the aortic mitral leaflet and should be removed. When the hypertrophy is predominantly in the mid-ventricle, the muscle resection has to be completed beyond the base of the anterior papillary muscle.

As in all procedures affecting haemodynamics, it is desirable to measure the operative result immediately. In hypertrophic cardiomyopathy this is especially important because of the fact that the morphology inside a relaxed heart is ‘difficult to judge’. Pressure curves both after normal beats and postextrasystolic have to be evaluated both before going on cardiopulmonary bypass and after weaning the patient from bypass allowing for sufficient time for myocardial recovery. Attention has to be paid to maintaining adequate blood volume and filling pressure. It can also be important to evaluate the varying resistance over the left ventricular outflow tract as a measure of surgical success. This can be determined from the relation of instantaneous pressure and flow.

The gradient under operative conditions may be small. However, the aortic pressure curve still shows the typical double-peak, a result of the bi-phasic flow into the aorta. After the opening of the aortic valve there is a rapid flow of short duration. Later in systole the flow volume decreases in spite of the rising pressure gradient. The flow increases again towards the end of systole, that is, during the phase when it normally recedes. Graphic construction of resistance shows only one peak which coincides with the minimum of flow. The relation of the two flow phases varies considerably with varying end-diastolic volume: this is well seen following extrasystoles. The increase of the contracting force is reflected by the rising pressure gradient and the enormously increased resistance. Following resection of muscle tissue there is no double peak in the flow-curve. Total resistance amounts to only about half of the preoperative value. After postextrasystolic beats, flow and pressure curves have the same characteristics as before surgery. However, total resistance under these circumstances is also reduced to about 50% of the pre-operative level.

Additionally, it is important to avoid misinterpreta-
tion of transient mitral regurgitation. Under operative conditions this can be easily excluded just by changing the conditions of anaesthesia, by testing with a small dose of catecholamines, or by prolongation of reperfusion as a type of assisted circulation.

Results of surgery

We have operated upon 160 patients with hypertrophic cardiomyopathy. For the majority of these cases, mitral insufficiency did not play an important role. Of these 160 cases, 15 belong to the midventricular type; 149 patients were in functional class III (NYHA) and 11 were in class IV. The mean ages and sex distribution are not different from other larger series. In all cases it was possible to abolish or to reduce the systolic gradient.

The hospital mortality in our series decreased with growing experience. For the period since 1977 the mortality was 5.0%. The overall mortality since 1963 was 7.5% (Table I) and was similar in patients with outflow tract and midventricular obstruction.

The perioperative mortality was 3.8% in simple hypertrophic cardiomyopathy, where only septal resection via a transaortic approach was performed. Our poorest results were in the small group of 9 patients with mitral lesions, predominantly regurgitation, 5 of whom died. In retrospect we believe that we were not aggressive enough in not replacing the mitral valve in patients with additional acquired mitral lesions. It was very difficult to repair the mitral valve in such cases, particularly when examination of the valve area after the usual sternum-splitting access was difficult because of massive left ventricular hypertrophy. Mitral valve replacement can be done more easily.

Causes of death and complications in patients who survived can be considered as related to the disease or to the special procedure, or non-related. Out of four patients dying from low cardiac output, three belonged to functional group IV. The mortality in this particular group was about 30%, caused only by heart failure. Three patients died from mitral regurgitation, including two of the six patients with severe acquired mitral lesions. These two patients both suffered from an atypical, midventricular lesion (Table II).

In two patients with minor procedure-related aortic valve injuries the cusps were repaired immediately with good long-term results. The septal resection caused septal perforation (USD) in six patients. In two cases the defect was detected and closed during the primary intervention. In another patient a second operation had to be performed 6 weeks later. In the remaining three patients the resulting shunt was minimal and surgical correction was not necessary. Total AV-block occurred five times; pacemaker-implantation was necessary and without complications. The occurrence of left bundle branch block is now more common, and is probably related to the more extensive myocardial resection which we now perform.

Looking at the non-related lethal and survived complications it has to be stressed that none of these can be avoided absolutely. One patient with sepsicaemia died on the fourth postoperative day, one patient with gastro-intestinal bleeding, on the tenth day. They were two of the first ten cases operated on (Table III).

Summary of the Düsseldorf experience

In summary the Düsseldorf surgical experience shows:

### Table II Hypertrophic cardiomyopathy - surgical series. Perioperative complications related to hypertrophic cardiomyopathy or special procedure

<table>
<thead>
<tr>
<th>Lethal (n)</th>
<th>Survived (n)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Low cardiac output</td>
<td>4</td>
</tr>
<tr>
<td>Sudden death</td>
<td>1</td>
</tr>
<tr>
<td>Mitral regurgitation</td>
<td>2</td>
</tr>
<tr>
<td>Mitral valve injury</td>
<td>1</td>
</tr>
<tr>
<td>Aortic valve injury</td>
<td>-</td>
</tr>
<tr>
<td>VSD</td>
<td>-</td>
</tr>
<tr>
<td>Septal infarction</td>
<td>-</td>
</tr>
<tr>
<td>Total AV-block</td>
<td>-</td>
</tr>
<tr>
<td>LBBB</td>
<td>-</td>
</tr>
<tr>
<td>RBBB</td>
<td>-</td>
</tr>
<tr>
<td>Cerebral emboli</td>
<td>-</td>
</tr>
</tbody>
</table>

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A(M)VR = aortic (mitral) valve replacement; L(R)V = left (right) ventricle; ACVBG = aortocoronary vein bypass graft.
Table III  Hypertrophic cardiomyopathy – surgical series. Perioperative complications non-related to hypertrophic cardiomyopathy or special procedures

<table>
<thead>
<tr>
<th></th>
<th>Lethal (n)</th>
<th>Survived (n)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Septicaemia</td>
<td>1</td>
<td>–</td>
</tr>
<tr>
<td>Intestinal bleeding</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Pulmonary embolism</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Rethoracotomy (bleeding)</td>
<td>1</td>
<td>4</td>
</tr>
<tr>
<td>Wound healing per secundum</td>
<td>–</td>
<td>2</td>
</tr>
<tr>
<td>Postoperative hernia</td>
<td>–</td>
<td>2</td>
</tr>
<tr>
<td>Pericardial effusion</td>
<td>–</td>
<td>1</td>
</tr>
<tr>
<td>Acute renal failure</td>
<td>–</td>
<td>2</td>
</tr>
<tr>
<td>Hepatitis B</td>
<td>–</td>
<td>3</td>
</tr>
</tbody>
</table>

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(1) Surgery can offer a method to abolish or at least to lower the pressure-gradient in typical and atypical cases of hypertrophic cardiomyopathy. (2) This can be done in otherwise uncomplicated cases with a risk of less than 2% mortality. (3) There is no special surgical risk for patients with atypical hypertrophic cardiomyopathy. (4) Surgically induced total AV-block cannot be avoided completely. (5) Indications for surgery may not yet be clarified, but from our point of view we recommend surgery for patients in functional class III (NYHA) and refractory to drug therapy before further deterioration.

The vast majority of patients who underwent surgical treatment were improved. The percentage in the group of typical hypertrophic cardiomyopathy was 83, in atypical hypertrophic cardiomyopathy 87.5. In the remaining patients a new onset of symptoms several years after a successful operation was observed. However, in none of these patients were the new symptoms more pronounced than preoperatively.

There was also a significant reduction in syncope post-operatively. During comparable follow-up periods, syncope occurred in only 2 operated patients out of 20 who had experienced this previously – a reduction of 90%.

Comparison of medical and surgical approaches

Published reports on the clinical course and the prognosis of patients with hypertrophic obstructive cardiomyopathy consider predominantly either studies in conservatively treated patients or operated patients. Little work has been carried out using comparative studies of patients with and without surgical treatment within one institution.

We have compared survival in patients who were treated surgically and medically and who were in identical functional classes (classes III and IV) of the New York Heart Association. Excluding perioperative mortality, the survival rates are significantly higher in the operative group. This difference becomes more pronounced if one compares the patients operated on since 1976 with those treated medically. No patient out of this series died postoperatively, compared to an annual mortality of about 6% in this subset of patients of class III and IV, who did not undergo surgical treatment.

Because complex ventricular arrhythmias have been shown to influence the prognosis, a study was undertaken by Borggrefe and co-workers (1983) in Düsseldorf, to analyse the influence of surgery on ventricular arrhythmias in patients with hypertrophic cardiomyopathy pre- and postoperatively. The incidence of these arrhythmias seems to be high even after operation. Whether they have the same prognostic value as in medically treated patients has to be established by further follow-up. Possibly complex ventricular arrhythmias are of lesser malignancy and better tolerated after surgical therapy.

The exercise performance of patients with and without surgery has been investigated by Lössle and co-workers (1983) in our cardiological department. The experience based on more than 200 maximal bicycle ergometer stress tests showed that exercise tests may be safely performed in patients with hypertrophic cardiomyopathy. The maximal exercise capacity was on average not changed after propranolol but increased after verapamil and, most significantly, after surgery. This could be attributed to corresponding haemodynamic changes, especially concerning cardiac output and pulmonary artery pressure. As a whole, verapamil was clinically and haemodynamically superior to propranolol, but not as effective as surgical treatment.

References


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