Eight patients with complex rheumatic heart disease underwent the MAZE procedure combined with mitral valve replacement. Some of the patients also required aortic valve surgery or tricuspid repair. There was one early death related to a stroke. The remaining patients have been followed from 3 to 19 mo. Atrial fibrillation was ablated in all patients. Six patients are in sinus rhythm and one patient in nodal rhythm. The MAZE procedure should be combined with surgery for rheumatic heart disease in selected patients because the MAZE operation has the potential to restore sinus rhythm, return atrial transport function, and obviate the need for anticoagulation.

ATRIAL FIBRILLATION is the most common chronic cardiac arrhythmia occurring in 0.4% to 2% of the population. The incidence increases with age and may affect from 4% to 10% of the population over 60 y.1-4 Atrial fibrillation is even more common in patients with rheumatic heart disease, affecting well over half of these patients.5

Atrial fibrillation is associated with an irregular and rapid heart beat, loss of atrial transport function, and an increased incidence of thromboembolism. Nonpharmacological treatment for patients who fail medical therapy has included left atrial isolation,6 AV node ablation,7 the Corridor procedure,8 and the MAZE operation.9

The MAZE operation was first described in 1987 for patients with lone atrial fibrillation8 and is the only interventional procedure that restores sinus rhythm, restores atrial transport, and reduces the risk of thromboembolism. Although the MAZE was initially recommended for patients with lone atrial fibrillation, this procedure has been performed in patients requiring mitral valve replacement or repair, during correction of congenital disorders, and in association with coronary artery bypass.10 The present study reviews our early experience with the MAZE operation for patients with complex rheumatic valvular heart disease.

Patients and Methods

Eight patients with rheumatic heart disease and chronic atrial fibrillation gave additional consent for the MAZE operation. The patients' profiles are illustrated in Table I. The patients ranged in age from 26 to 60 y. All patients had mitral valve disease associated with some degree of chronic pulmonary hypertension. Three patients had hemodynamically significant aortic valve disease and 6 patients had echocardiographic evidence of tricuspid insufficiency.

The ejection fraction was normal in the majority of patients. Half of the patients had suffered a peripheral embolus despite anticoagulation. Three of the systemic emboli were cerebral, one resulting in a permanent left-sided paralysis.
Indications for Operation

All patients were presented before the weekly Combined Cardiac Conference and accepted for operation based upon the need for valve replacement or repair. The MAZE operation was recommended for these patients with chronic atrial fibrillation because of a history of previous emboli (despite adequate anticoagulation), difficulty obtaining satisfactory follow-up for chronic anticoagulation, a desire to have more children, or because of an allergy to warfarin in one patient.

Operative Technique

Anesthesia was induced with sufentanil and maintained with midazolam, sufentanil, and pancuronium. A Medtronic Biomedicus pump (Medtronic Incorporated, Cardiovascular Systems, Roseville, MA, USA) was used for bypass, and high-dose aprotinin was administered to reduce the likelihood of postoperative bleeding. II Continuous warm, antegrade/retrograde blood cardioplegia was used in all patients.

The patients underwent a MAZE III operation.12 The incisions were carried out as described by Dr. Cox but the sequence of the incisions was modified. The right MAZE was performed on cardiopulmonary bypass and the tricuspid valve was repaired, if indicated, during the right MAZE. The heart was then arrested and the left MAZE was begun. The encircling incision was started from the right superior pulmonary vein and continued up to the level of the mitral annulus. The T-incision was then carried to within one centimeter of the mitral annulus and a cryofreeze was placed at the level of the annulus. The encircling incision and the T-incision were then closed. Mitral valve replacement was then performed, followed by the completion of the left MAZE and the septal incision. Aortic surgery, if required, was performed following completion of the mitral/MAZE operation.

Statistical Analysis

The results are reported as ± standard error of the mean. Paired Student's t-tests were used for statistical comparisons.

Results

All patients survived the operation, however, there was one death two weeks after operation. The death occurred in a 53-year-old female who had undergone an uneventful aortic valve replacement, mitral valve replacement, and MAZE. She had been returned to the intensive care unit without the need for inotropic support. Twenty-four hours later, when she was being considered for extubation, she developed an episode of ventricular tachycardia which deteriorated into ventricular fibrillation. Chest compressions were started and she was defibrillated. The cardioverted rhythm was nodal; the most common rhythm after the MAZE and the rhythm that she had been in since the operation. She remained hypotensive after cardioversion; thus, the chest was opened. The mediastinum and the right hemithorax was full of blood. A small laceration was found on the anterior wall of the ascending aorta; the laceration undoubtedly being related to the chest compressions. Although she was resuscitated successfully, the period of hypotension resulted in a stroke from which she did not recover.

The remaining patients have been followed from 3 to 19 mo (mean, 11 mo). Table 2 illustrates the type of operation performed, the early and late cardiac

<table>
<thead>
<tr>
<th>Age</th>
<th>Sex</th>
<th>Prior emboli</th>
<th>EF</th>
<th>SPAP</th>
<th>Aortic</th>
<th>Mitral</th>
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<td>AI</td>
<td>MS/MR</td>
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<td>F</td>
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<td>42</td>
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<td>MS/MR</td>
<td>TR</td>
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<td>F</td>
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<td>100</td>
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<td>MRIMS</td>
<td>TR</td>
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<tr>
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<td>M</td>
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<td>57</td>
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<td>35</td>
<td>AS</td>
<td>MS/MR</td>
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EF = ejection fraction; SPAP = systolic pulmonary artery pressure; C =

Table 1. Patient profiles.
Table 2. Operative results

<table>
<thead>
<tr>
<th>Age/Sex</th>
<th>Operation</th>
<th>Early/R</th>
<th>Late/R</th>
<th>Pacing</th>
<th>AT</th>
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<td>SIN,AF</td>
<td>SIP</td>
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<td>SIN</td>
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<tr>
<td>53/F</td>
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<tr>
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<tr>
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<td>N/AF</td>
<td>S</td>
<td>No</td>
<td>E</td>
</tr>
</tbody>
</table>

Early/R = early rhythm before discharge; Late/R = rhythm after 3 months; AT = atrial transport; MVR = mitral valve replacement; AVR* = aortic valve repair; AVR = aortic valve replacement; TVR* = tricuspid valve repair; SIN = sinus rhythm alternating with nodal rhythm; N = nodal rhythm; AF = atrial fibrillation; SIP = sinus atrial paced; S = sinus rhythm; AAI = atrial pacing; U = undetermined lost to follow-up; E = too early to assess.

Discussion

Although the MAZE procedure was described ten years ago, most centers with cardiac programs still do not offer the operation to patients with drug-refractory atrial fibrillation, and even fewer centers perform the MAZE operation in conjunction with other cardiac surgery. The most likely reason for the reluctance of many centers to adopt the MAZE procedure is related to the fact that the operation is technically demanding and may increase the duration of cardiopulmonary bypass by as much as one hour.

The MAZE operation was originally described for patients with lone atrial fibrillation by Cox and colleagues.9 One hundred and seventy-eight patients underwent the Cox/MAZE operation during an 81/2-year period from 1987 to 1996. Two thirds of these patients had an isolated MAZE operation. However, with increasing experience, the MAZE was combined with other operations12 which included coronary artery bypass (14%) or MAZE operation and mitral valve surgery (12%).

The operation has successfully ablated atrial fibrillation in 93% of patients; the remaining 7% being converted and maintained in sinus rhythm with drug therapy. Left atrial transport has been restored in 97% of patients after the MAZE III procedure and right atrial transport in 94%. There have been two late transient ischemic attacks and no procedure-related strokes.

The results of the MAZE III operation have been excellent in patients with lone atrial fibrillation but less satisfactory in patients with mitral valve disease. Kosakai and colleagues13 recently reported the only large series of patients who have undergone a MAZE procedure combined with other cardiac operations. Of the 101 patients, 86 patients (85%) had mitral valve replacement or repair. Twenty-five percent of this group had an isolated mitral valve operation. The remainder of the patients underwent tricuspid (36%), aortic (24%), aortic and tricuspid valve operations (12%), or coronary artery bypass (2%).

rhythm, the need for pacing, and the return of atrial transport. All patients were in sinus, nodal, or paced rhythm immediately after operation. At discharge, the predominant rhythm was nodal with three patients in atrial fibrillation. All three of these patients converted to sinus rhythm in the first 3 to 6 mo and no longer take antiarrhythmic medication.

Two patients remained in nodal rhythm after 6 mos. of follow-up. An attempt was made to insert an atrial lead in one of these patients. However, the atrial lead, with active fixation, could not be secured to the wall of the right atrium; the atrial appendage being removed as part of the MAZE operation. However, temporary atrial pacing during the attempted procedure produced normal sinus rhythm. Almost 11;2 years after the surgery, this patient is doing well without anticoagulation. In the second patient, we were able to secure a lead with active fixation to the wall of the right atrium. She is now in sinus rhythm with AAI pacing and has atrial transport.

Atrial dimensions were measured by echocardiography before and after operation. The anterior/posterior diameter of the left atrium (M -mode) decreased from 53 i: 3 mm to 41 i: 2 mm (P < 0.004) after operation and the left atrial diameter, measured in the apical view, decreased from 70 i: 3 mm to 50 i: 2 mm (P < 0.001). The left atrial volume, calculated by Simpson's Formula, was reduced from 127 i: 14 cc to 56 i: 7 cc (P < 0.002).
Sinus rhythm was restored in 79% of the patients. Junctional rhythm was present in 4% of patients after operation while 16% of this group failed the operation and remain in atrial fibrillation. The patients who did not resume sinus rhythm following operation had larger left atria (P < 0.02) and had experienced atrial fibrillation for a mean of 14 ± 6 years; 6 ± 7 years longer than those patients who converted to sinus rhythm (P < 0.001).

There may be two reasons for the lower incidence of sinus rhythm after operation in the series reported by Kosakai. Firstly, Kosakai and colleagues modified the Cox/MAZE incisions and therefore the results may not be comparable. Secondly, and more importantly, the majority of patients in the series from Barnes Hospital had lone atrial fibrillation and therefore the size of the left atrium was most likely smaller than the atria in the Japanese series since 43% of their patients had giant left atria. The size of the atrium has implications for a successful outcome because the MAZE operation is based upon the principle that there is a minimum mass of atrial tissue necessary to sustain a macro-reentrant circuit. The MAZE operation was designed to create multiple corridors or a maze that would conduct the electrical impulse from the SA node to the AV node. The corridors are sufficiently narrow so that the mass of atrial tissue between the incisions is less than the critical mass required to sustain a macro-reentrant arrhythmia. Since the incisions are made in the same manner in all patients, the prescribed incisions will be further apart and the atrial mass greater within the corridors in patients with giant left atria. The anatomical difference between small and giant left atria will predispose those patients with enlarged atria to failure of the operation. We measured the dimensions of the normal left atria by echocardiography in our patients, and after comparing these dimensions with giant left atria, it became apparent that the majority of the enlargement takes place on the posterior wall of the left atrium. For this reason, we enlarged the isolating pulmonary vein incision which may explain our success in ablating atrial fibrillation in all our patients.

The MAZE operation has a number of complications that are peculiar to this operation. The patients are at greater risk of bleeding, usually develop postoperative fluid retention, frequently experience transient atrial arrhythmias for the first six months after operation, and may require an atrial pacing lead.

Bleeding after the MAZE may be due to surgical or medical problems. Cox reported a 5% incidence of bleeding after the Cox/MAZE operation for lone atrial fibrillation. Kosakai reported an 8% incidence of postoperative hemorrhage for patients undergoing the MAZE operation combined with other procedures. The large number of atrial incisions increases the likelihood of hemorrhage, while the increased length of bypass required to perform the MAZE and concomitant surgery predisposes the patient to a bleeding diathesis. Our first three patients who had bleeding problems had to be returned to the operating room. One patient had a small artery bleeding in one of the posterior atrial incisions while the other two had medical bleeding. Both these patients had been on heparin for a number of weeks before operation and had right heart failure and hepatomegaly. To circumvent these potential bleeding problems, we now use high-dose aprotinin, a Medtronic Biomedicus Impeller pump, and cautery to make the atrial incisions. Since instituting these measures, we have not had to return any patients to the operating room for hemorrhage.

Fluid retention affects almost all patients in the first few days after operation. This problem is thought to be related to a decrease in atrial naturetic hormone secondary to multiple atriotomies that complete the MAZE operation. Spironolactone has been recommended to manage this problem. Although we routinely use this diuretic, we have found that continuous intravenous furosemide or bumetanide is the most satisfactory way to deal with this problem. The infusion is maintained for 2 to 3 d and then discontinued.

Transient atrial arrhythmias complicate the recovery of at least 44% of patients undergoing a MAZE operation. Atrial fibrillation or flutter, although most frequently reported early after operation, may occur at any time in the first three months. The recurrence of atrial arrhythmia is thought due to postoperative tissue edema, pericarditis, surgical trauma, and elevated catecholamines; all these factors shorten the effective refractory period which allow these arrhythmias to be sustained in a smaller mass of atrial tissue. The arrhythmias are treated in the usual fashion. Three of our patients experience postoperative atrial fibrillation/flutter early after operation but have since reverted to sinus rhythm without the need for medication.
tachycardia may unmask underlying SA node disease (24%) or the SA node may have a blunted chronotropic response (6%). In our series, two patients remained in nodal rhythm, one patient being successfully managed with an AAI pacemaker.

Limitations of the Study

The main limitation of this study is the limited numbers of patients. However, one must realize that many centers do not perform the MAZE, and the experience with the MAZE and complex rheumatic heart disease is even more limited. Despite the small number of patients in our study, the results of the MAZE for the ablation of atrial fibrillation are better than those reported by Kosakai and comparable to those reported by Cox.

Conclusions

The MAZE procedure should be recommended for selected patients with chronic atrial fibrillation who require surgery for rheumatic heart disease as the operation will restore a regular heart rhythm, return normal atrial transport function, and reduce the risk of thromboembolism. The operation is particularly beneficial for patients in Saudi Arabia because many of our patients with mitral valve disease are young and wish to have more children. Repair or replacement of the mitral valve with a tissue valve combined with the MAZE will restore sinus rhythm and obviate the need for anticoagulation. Many of our patients also have difficulty regulating warfarin or come from remote areas making regular laboratory testing impractical. Therefore, return of sinus rhythm or even nodal rhythm allows the patient to return home on aspirin rather than subjecting the patient to a lifetime of anticoagulation.

References