Effect of Ebstein’s Anomaly on Short- and Long-term Outcome of Surgically Treated Patients With Wolff-Parkinson-White Syndrome

Joyce C. Pressley, MPH; J. Marcus Wharton, MD; Anthony S. L. Tang, MD; James E. Lowe, MD; John J. Gallagher, MD; and Eric N. Prystowsky, MD

Background. Ebstein’s anomaly is the most commonly occurring congenital abnormality associated with the Wolff-Parkinson-White (WPW) syndrome. However, the effects of Ebstein’s anomaly on the risks and benefits of surgical ablation of accessory pathways in patients with WPW syndrome are unknown.

Methods and Results. This study compared the long-term outcome of 38 WPW patients with Ebstein’s anomaly undergoing accessory pathway ablation to a reference population of 384 similarly treated patients without the anomaly. Ebstein’s anomaly was mild in 21 patients (55%) and moderate-to-severe in 17 patients (45%). Sixteen patients (42%) required tricuspid valve surgery, and 23 (61%) had an atrial septal defect or patent foramen ovale repaired. Baseline clinical characteristics and preoperative clinical arrhythmias were similar in both groups. Ten-year survival was 92.4% and 91.2% for patients with and without Ebstein’s anomaly, respectively (p=NS). During a mean follow-up of 6.2±3.8 and 5.3±3.6 years, 82% of patients with and 90% without Ebstein’s anomaly had either clinically insignificant or no arrhythmias, and 18% versus 10% reported symptoms suggesting arrhythmias lasting longer than 1 minute, respectively. Atrial fibrillation was reduced postoperatively to 9% (p<0.001) in patients with and to 4% (p<0.001) in those without the anomaly. Fewer hospitalizations were reported postoperatively by 90% versus 96% of patients with and without Ebstein’s anomaly; 9.4% versus 6.0% of patients were disabled at follow-up, respectively (p=NS).

Conclusions. Patients with Ebstein’s anomaly are improved significantly after accessory pathway ablation. The presence of this anomaly should not preclude accessory pathway ablation in these patients.

(Circulation 1992;86:1147–1155)

KEY WORDS • Wolff-Parkinson-White syndrome • Ebstein’s anomaly • atrial fibrillation

Ebstein’s anomaly is the most commonly occurring congenital defect associated with the Wolff-Parkinson-White (WPW) syndrome.1 The anomaly, characterized by downward displacement of the tricuspid valve resulting in an atrialized posterior right ventricle, is frequently accompanied by other abnormalities such as atrial septal defect, patent foramen ovale, right atrial enlargement, and myocardial abnormalities.1-4 The degree of tricuspid valve regurgitation has been reported as a primary cause of the functional limitations observed in these patients,5,6 whereas right atrial enlargement and right ventricular abnormalities have been implicated in the increased incidence of atrial fibrillation and ventricular tachycardia, respectively.7,8 The clinical course of these patients is very diverse, with many patients with Ebstein’s anomaly remaining asymptomatic, whereas others experience varying degrees of cyanosis, dyspnea, fatigue, arrhythmias, and congestive heart failure.9-11 Although patients with Ebstein’s anomaly may live beyond the normal life expectancy with no functional limitations,9 there are also reports of arrhythmic deaths in these patients in the absence of significant anatomical malformation or hemodynamic impairment.10

Successful surgical ablation of accessory atroventricular (AV) pathways in patients with the WPW syndrome has been shown to eliminate AV reentrant tachycardia12-15 and also virtually to eliminate the recurrence of atrial fibrillation in patients without other heart disease.16 However, the benefit of accessory pathway ablation on the long-term arrhythmia recurrence of patients with Ebstein’s anomaly is less clear, because these patients may also have arrhythmias related to abnormalities of the atrial and ventricular myocardium as a result of their Ebstein’s anomaly.5,10,11

This study assessed long-term survival, arrhythmia recurrence, activity limitations, and rehospitalizations of patients with WPW syndrome and Ebstein’s anomaly after surgical ablation of their accessory pathway and compared these with data from a group of patients without Ebstein’s anomaly who underwent surgical correction of the WPW syndrome.

Methods

Patient Population

The study population comprised a consecutive series of 422 patients who received surgical ablation of their...
accessory pathway(s), 38 with and 384 patients without Ebstein's anomaly. The study population was selected from 600 patients referred for arrhythmias between May 1968 and November 1988 who subsequently had an accessory pathway electrophysiologically documented. Forty-nine (8%) of the 600 patients studied for WPW also had Ebstein's anomaly. Eleven (22%) of the 49 patients with Ebstein's anomaly and 167 (30%) of the 551 patients without the anomaly were treated with methods other than surgical ablation of their accessory pathway and were excluded. Of the 11 Ebstein's anomaly patients who were excluded, nine (82%) had mild Ebstein's anomaly and two (18%) had moderate-to-severe Ebstein's anomaly. Of the two patients with moderate-to-severe Ebstein's anomaly, one was judged to be a poor surgical candidate, and the other refused surgery.

Ebstein's anomaly was diagnosed by echocardiography in 65% of patients using established criteria and was confirmed in all patients at the time of surgery. Echocardiography was not in widespread clinical use for patients referred before 1976. Severity of the Ebstein's anomaly was classified according to functional criteria. Twenty-one patients (55%) had mild Ebstein's anomaly and were New York Heart Association (NYHA) functional class I or II. They had mild or no tricuspid regurgitation and, with the exception of their arrhythmia, had minimally significant or no symptoms. Seventeen patients (45%) were NYHA functional class III or IV with moderate-to-severe tricuspid regurgitation. All had physical activity limitations, fatigue, and/or dyspnea in addition to symptoms of their arrhythmia.

Surgical Technique

The surgical techniques used in these patients evolved over an 18-year period and underwent a variety of modifications from the original epicardial approach devised by Sealy and his associates. In the past, after completion of intraoperative mapping, cold potassium cardioplegic arrest was used for division of all accessory AV pathways. The current endocardial technique uses cold potassium cardioplegic solution only for patients with left-sided pathways. Dissection of all right-sided pathways, including those in the posterior septal space, is performed under normothermia with the heart beating and normally perfused. In patients also undergoing closure of a patent foramen ovale or an atrial septal defect, the heart is briefly fibrillated, closure completed, and then defibrillated before interruption of the accessory pathway. In patients undergoing repair or replacement of the tricuspid valve, the accessory pathway is ablated before valve surgery. Of the 16 patients undergoing tricuspid valve surgery, four had tricuspid valve repairs, including one each of a cleft posterior leaflet repair, an anterior leaflet repair, an anterior leaflet repair with resuspension of the posterior leaflet, and a tricuspid annulus plication. The other 12 patients underwent tricuspid valve replacements. A Hancock porcine valve was used in five patients, a Carpentier valve in five, an Inoescue-Shiley xenograft in one, and a Starr-Edwards prosthesis in one.

Follow-up assessment was accomplished by mailed questionnaire designed to assess general health and activity levels, symptoms indicative of arrhythmia recurrence, antiarrhythmic drug use, cardiac-related hospitalizations, and emergency room and physician office visits. Patients reporting symptoms of arrhythmias after surgery were asked to describe the relative frequency and severity before and after surgery as well as the shortest, longest, and typical episode. Patients not responding to mailed questionnaires were interviewed by telephone. Referring physicians were contacted for documentation of any symptoms suggestive of arrhythmias occurring during long-term follow-up and copies of ECGs, Holters, and/or electrophysiological restudies were obtained. The causes of death were determined from referring physicians, hospital records, and death certificates.

Follow-up was achieved in 95% of patients with and in 90% of patients without Ebstein's anomaly. Mean follow-up was 6.2±3.8 years for patients with Ebstein's anomaly and 5.3±3.6 years for those without the anomaly. There were no significant differences in age, coexisting disease (other than Ebstein's anomaly), or success of surgical ablation for patients lost to follow-up compared with those whose current status was documented.

Definitions

Atrial fibrillation was classified as new when it was documented to occur more than 3 months after surgery in patients without prior documented atrial fibrillation. For purposes of analyzing long-term follow-up data, patients undergoing surgery twice were considered to have undergone a successful procedure if the accessory pathway was divided at the second operation. An unsuccessful procedure occurred when conduction remained over an accessory pathway after attempted division of the pathway or if it became apparent that another pathway not previously identified was present. Postoperative accessory pathway function was assessed using a combination of intraoperative electrophysiological testing, postoperative electrophysiological testing generally between postoperative days 6 and 9, serial 12-lead ECGs, and continuous ECG monitoring. Electrophysiological testing was also repeated during long-term follow-up in selected patients who were known or suspected surgical failures. Improvements in all quality of life measurements (including frequency and severity of arrhythmias and decreases in hospitalizations) are measured in percent of patients reporting improvements rather than decreases in the number of events. Perioperative mortality was defined as death occurring from any cause during surgery or at any time after surgery but before hospital discharge. Sudden death was defined as mortality occurring within 1 hour of onset of symptoms or unwitnessed deaths. Late mortality was defined as deaths occurring after hospital discharge.

Statistics

Statistical analyses used the conventional methods of Fisher's exact or χ² for testing differences in discrete variables, the Kaplan-Meier method for survival analysis, and the Mann-Whitney U test for comparison between groups. A value of p<0.05 was considered significant. All confidence intervals for odds ratios were calculated at the 95% level.
TABLE 1. Characteristics of Patients With and Without Ebstein's Anomaly

<table>
<thead>
<tr>
<th></th>
<th>Ebstein's anomaly</th>
<th></th>
<th></th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Present (n=38)</td>
<td>Absent (n=384)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mean age at surgery (years)</td>
<td>26.3±12.3</td>
<td>31.0±14</td>
<td>NS</td>
<td></td>
</tr>
<tr>
<td>Mean duration of arrhythmia (years)</td>
<td>15.0±9.4</td>
<td>13.7±12.4</td>
<td>NS</td>
<td></td>
</tr>
<tr>
<td>Mean number of antiarrhythmic drug trials</td>
<td>3.0±1.5</td>
<td>3.1±1.6</td>
<td>NS</td>
<td></td>
</tr>
<tr>
<td>History of cardiac arrest</td>
<td>2 (5.3%)</td>
<td>58 (15.1%)</td>
<td>NS</td>
<td></td>
</tr>
<tr>
<td>Other heart disease</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mitral valve prolapse</td>
<td>5 (13.2%)</td>
<td>36 (9.4%)</td>
<td>NS</td>
<td></td>
</tr>
<tr>
<td>Coronary artery disease</td>
<td>1 (2.6%)</td>
<td>17 (4.4%)</td>
<td>NS</td>
<td></td>
</tr>
<tr>
<td>Cardiomyopathy</td>
<td>3 (7.9%)</td>
<td>15 (3.9%)</td>
<td>NS</td>
<td></td>
</tr>
<tr>
<td>Other congenital disease</td>
<td>3 (7.9%)</td>
<td>9 (2.3%)</td>
<td>NS</td>
<td></td>
</tr>
<tr>
<td>Documented atrioventricular reciprocating tachycardia</td>
<td>29 (76.3%)</td>
<td>269 (70.0%)</td>
<td>NS</td>
<td></td>
</tr>
<tr>
<td>Documented atrial fibrillation</td>
<td>16 (42.1%)</td>
<td>165 (43.0%)</td>
<td>NS</td>
<td></td>
</tr>
<tr>
<td>Mean follow-up (years)</td>
<td>6.2±3.8</td>
<td>5.8±3.6</td>
<td>NS</td>
<td></td>
</tr>
</tbody>
</table>

Results

Patient Population

The baseline characteristics of patients with and without Ebstein's anomaly are shown in Table 1. Mean age at time of surgery, duration of arrhythmia, distribution of presenting clinical arrhythmias, and the presence of other associated heart disease did not differ significantly between the two groups (Table 1). Twenty-eight Ebstein's anomaly patients (74%) underwent an associated surgical procedure at the time of their accessory pathway ablation. Tricuspid valve surgery was performed in 16 patients (42%), atrial septal defect repairs in 17 (45%), and closure of a patent foramen ovale in six (16%).

Pathway distributions are shown in Table 2. Patients with Ebstein's anomaly more frequently had multiple pathways (50% versus 15%, p<0.001) and their pathways more commonly occurred in a right (78.9% versus 21.6%, p<0.001) and posteroseptal (57.9% versus 23.7%, p<0.001) location than patients without Ebstein's anomaly. In fact, 18 patients (47%) with Ebstein's anomaly compared with five (1%) without the anomaly had the combination of a posteroseptal and right freewall accessory pathway (p<0.001). Three patients (7.9%) with Ebstein's anomaly and 241 (62.8%) of the patients without the anomaly, respectively, had left-sided pathways (p<0.001). One patient with mild Ebstein's anomaly had a single left-sided pathway. The remaining two patients with Ebstein's anomaly and left-sided pathways had congenitally corrected transposition of the great vessels with the accessory pathway located on the same side as the morphological tricuspid valve. One of these patients had a single accessory pathway; the other had a total of three pathways located in the left free wall, posteroseptal, and anteroseptal positions. Mean cycle length of AV reciprocating tachycardia was significantly longer in patients with than in those without Ebstein's anomaly (358.8 msec versus 310.9 msec, p<0.001). The shortest RR interval of preexcited complexes during atrial fibrillation was similar between the two groups (Table 2). Success of surgical accessory pathway ablation was not different in the two groups (92% and 90%), respectively.

Perioperative Mortality

Two of the 38 patients with Ebstein's anomaly and 13 of the 384 patients without Ebstein's anomaly died after surgery, for a perioperative mortality of 5.3% and 3.4%, respectively (p=NS). Both of the deaths in the Ebstein's anomaly group occurred in patients aged 15 years and younger who were also undergoing surgery for associated cardiac disease. Causes of perioperative death in the Ebstein's anomaly group included hypotension, pneumonia, and cardiopulmonary arrest in a

TABLE 2. Baseline Electrophysiological Characteristics of Patients With and Without Ebstein's Anomaly

<table>
<thead>
<tr>
<th></th>
<th>Ebstein's anomaly</th>
<th></th>
<th></th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Present</td>
<td>Absent</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Multiple pathways (% patients)</td>
<td>19 (50.0%)</td>
<td>58 (15.1%)</td>
<td>&lt;0.001</td>
<td></td>
</tr>
<tr>
<td>Pathway distribution (% patients)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Right</td>
<td>30 (78.9%)</td>
<td>83 (21.6%)</td>
<td>&lt;0.001</td>
<td></td>
</tr>
<tr>
<td>Posterosetal</td>
<td>22 (57.9%)</td>
<td>91 (23.7%)</td>
<td>&lt;0.001</td>
<td></td>
</tr>
<tr>
<td>Left</td>
<td>3 (7.9%)*</td>
<td>241 (62.8%)</td>
<td>&lt;0.0001</td>
<td></td>
</tr>
<tr>
<td>Anterosetal</td>
<td>3 (7.9%)</td>
<td>28 (7.3%)</td>
<td>NS</td>
<td></td>
</tr>
<tr>
<td>Mean cycle length at electrophysiological study (msec)</td>
<td>358.8±65.9</td>
<td>310.9±47.8</td>
<td>&lt;0.0001</td>
<td></td>
</tr>
<tr>
<td>Atrioventricular reciprocating tachycardia</td>
<td>215.3±52.5</td>
<td>218.5±58.2</td>
<td>NS</td>
<td></td>
</tr>
<tr>
<td>Shortest preexcited RR during atrial fibrillation</td>
<td>215.3±52.5</td>
<td>218.5±58.2</td>
<td>NS</td>
<td></td>
</tr>
</tbody>
</table>

*Two of three patients had congenitally corrected transposition of the great vessels.
2-year-old patient in preoperative NYHA class I with a large ventricular septal defect, and incessant supraventricular tachycardia and low cardiac output after tricuspid valve replacement in a 14-year-old patient with Ebstein's anomaly class III failure. In the patients without Ebstein's anomaly, nine of the 13 perioperative deaths (69.2%) occurred in patients with severe cardiac disease: dilated cardiomyopathies (four patients), coronary artery disease (two patients), chronic obstructive pulmonary disease (one patient), extreme concentric left ventricular hypertrophy (one patient), and a large ventricular septal defect (one patient). Causes of death in the patients without Ebstein's anomaly included low cardiac output in 10 patients, arrhythmia in one, pancreatitis in one, and bleeding with subsequent respiratory and renal failure in one.

Of the 12 patients with Ebstein's anomaly undergoing tricuspid valve replacement, perioperative death occurred in one of two patients aged 15 years or younger compared with none of the 10 patients over the age of 15 years. Mortality in the Ebstein's anomaly patients not undergoing tricuspid valve replacement was one of five (20%) for those 15 years and under and 0 of 21 for those over 15 years of age. Mortality for the group without Ebstein's anomaly was three of 60 (5.0%) for those 15 years and under and 10 of 324 (3.1%) for those over 15 years, respectively. There has been no perioperative mortality in the 13 patients with Ebstein's anomaly undergoing accessory pathway ablation in the last 5 years. Perioperative mortality for all surgical accessory pathway ablation is approximately 1.0% in the past 5 years.

Long-term Outcome

Survival was greater than 90% for patients with and without Ebstein's anomaly at both 5 years (92.4% versus 93.5%) and 10 years (92.4% versus 91.2%), respectively (Figure 1). Survival in the 12 patients with Ebstein's anomaly undergoing both tricuspid valve replacement and accessory pathway ablation was slightly lower (84.3%) by year 5, but both the 5- and 10-year survival figures remained stable at that level. Although four patients in the Ebstein's anomaly group without valve replacements who underwent valve repairs, survival in this group remained at 95.8% 10 years after surgery (Figure 1).

One of the two deaths (50%) occurring during follow-up in the Ebstein's anomaly group and five of the 20 deaths (25%) in the group without Ebstein's anomaly occurred in patients who were known to have cardiomyopathy and decreased ventricular function at the time of surgery. Both of the long-term deaths in the Ebstein's anomaly group occurred in patients with moderate-to-severe Ebstein's anomaly and tricuspid valve replacements (Hancock porcine and Starr-Edwards valves). One was a 29-year-old man with severe cardiomyopathy who died 4.2 years after surgery following attempted cardioversion for atrial arrhythmias in his community hospital. The other was a 51-year-old woman with unsuccessful surgical ablation of her accessory pathway who had sudden cardiac death 12 years after surgery. Both Ebstein's anomaly patients were on antiarrhythmic drugs at the time of death. Of the 20 deaths occurring during long-term follow-up in patients without Ebstein's anomaly, five were due to sudden cardiac deaths (three with associated cardiomyopathies, one with unsuccessful pathway ablation, and one with a history of ventricular tachycardia). Three additional patients died of complications related to prior existing cardiomyopathies, two of noncardiac causes, one of coronary artery disease, one of stroke, and eight of undocumented cause.

Arrhythmias During Follow-up

Compared with their preoperative status, 87% of patients with and 92% of patients without Ebstein's anomaly reported a decrease in the frequency of symptoms suggestive of arrhythmias (p<0.001), and 96% versus 93% of patients reported a decrease in the severity of symptoms of arrhythmias (p<0.001), respectively. Forty percent of patients with and 50% of patients without Ebstein's anomaly reported no symptoms of arrhythmias after accessory pathway ablation (p=NS) (Figure 2). Of the remaining patients, approximately 40% in both groups reported symptoms of palpitations lasting only seconds (Figure 2). Documentation from local physicians primarily demonstrated premature ventricular and atrial complexes in this latter group. Six patients (18%) with Ebstein's anomaly and 32 patients (10%) without the anomaly reported symptoms of arrhythmias lasting longer than 1 minute.
(p = NS). Of the six patients with Ebstein's anomaly and symptoms of sustained arrhythmias, three continued to have preexcitation: one with successful accessory pathway ablation had a residual Mahaim fiber participating in reciprocating tachycardia and two with unsuccessful surgery experienced recurrent reciprocating tachycardia or atrial fibrillation. In the three patients with Ebstein's anomaly and symptoms of sustained arrhythmias with successful pathway ablation, atrial flutter and atrial fibrillation were documented in one each. In the third patient, no arrhythmia could be documented. Of the 32 patients without Ebstein's anomaly reporting symptoms of arrhythmias lasting longer than 1 minute, 14 had atrial fibrillation, five had atrial flutter, five had recurrent reciprocating tachycardia, and eight had no documented arrhythmias.

Preoperatively, a similar percentage of patients with and without Ebstein's anomaly experienced atrial fibrillation (Figure 3). Preoperative atrial fibrillation was not different in the group of patients with mild compared with those with moderate-to-severe Ebstein's anomaly (43% versus 41%, respectively). During long-term follow-up of the Ebstein's anomaly group, atrial fibrillation decreased significantly from 42% (16 patients) preoperatively to 9% (three patients) postoperatively (p < 0.001). One of these experienced new atrial fibrillation, and two with a history of atrial fibrillation preoperatively continued to have the rhythm postoperatively. The patient with new atrial fibrillation had mild Ebstein's anomaly and preexcitation over a residual Mahaim fiber. The two patients who continued to experience atrial fibrillation had moderate-to-severe Ebstein's anomaly, one with unsuccessful accessory pathway ablation. In the patients without Ebstein's anomaly, atrial fibrillation was reduced from 43% preoperatively to 4% (14 patients) postoperatively (p < 0.001). Two patients without Ebstein's anomaly experienced new atrial fibrillation. One was a 63-year-old woman with hypertension, and the other continued to have preexcitation after unsuccessful pathway ablation.

The odds of a patient with Ebstein's anomaly reporting symptoms of a sustained arrhythmia during long-term follow-up was 2.3 times (confidence interval, CI, 0.8–5.2) that of the patient without Ebstein's anomaly. In patients alive at follow-up, the odds of an Ebstein's anomaly patient having documented atrial fibrillation during follow-up was 2.0 times (CI, 0.6–7.5) that of the patient without the anomaly. When patients with Ebstein's anomaly were compared with those without the anomaly, the odds of a patient alive at follow-up having documented atrial fibrillation during follow-up was 1.2 (CI, 0.1–9.2) for the patient with mild Ebstein's anomaly and 3.4 (CI, 0.7–16.4) for patients with moderate-to-severe Ebstein's anomaly.

**Quality of Life**

The number of patients on antiarrhythmic therapy decreased significantly from nearly 100% observed preoperatively to 26% (eight patients) postoperatively in the Ebstein's anomaly group and to 22% (68 patients) in patients without Ebstein's anomaly (Figure 4). Within the Ebstein's anomaly group, four patients (33%) in the moderate-to-severe group were treated postoperatively with antiarrhythmic therapy as compared with four patients (21%) with mild Ebstein's anomaly (p = NS). Patients with mild Ebstein's anomaly were similar to patients with no Ebstein's anomaly in the number of patients who remained on antiarrhythmic therapy after surgery (21% versus 22%, respectively).

There was a marked reduction in cardiac-related hospitalizations after surgery for 90% of patients with Ebstein's anomaly and for 96% without Ebstein's anomaly (Figure 4). The majority of patients in both groups were employed or full-time students, with only three patients (9%) with Ebstein's anomaly and 19 patients (6%) without Ebstein's anomaly on medical disability at follow-up (p = NS). All of the disabled patients with Ebstein's anomaly were in the moderate-to-severe category with tricuspid valve replacements. In addition, one of these had developed cardiomyopathy and one had unsuccessful accessory pathway ablation. Of the 19 patients without Ebstein's anomaly who were on disability, three had coronary artery disease, three had unsuccessful pathway ablation, four had cardiomyopathy, and nine had unknown reasons.

**Discussion**

**Major Findings**

Patients with Ebstein's anomaly and the WPW syndrome who undergo surgical ablation of accessory pathways, similar to those without the anomaly, show significant long-term improvement in the incidence of postoperative arrhythmias, including atrial fibrillation. Despite the myocardial and other existing structural...
abnormalities present, the majority of patients with Ebstein's anomaly either had no symptomatic arrhythmias postoperatively or arrhythmias of short duration (less than 60 seconds). The occurrence of atrial fibrillation was significantly reduced in both groups after surgery. Fewer patients received antiarrhythmic medications postoperatively, and hospitalizations were significantly lowered. This is accomplished with an operative mortality that is slightly but not significantly higher than that observed for accessory pathway ablation in patients without Ebstein's anomaly but significantly lower than that reported in many studies during the same time period for correction of Ebstein's anomaly alone.24,25 The majority of patients in both groups were alive and not disabled at follow-up.

Perioperative Mortality

The mortality experience in this study would suggest that there is little increased risk of death in patients undergoing simultaneous corrective procedures for both the WPW syndrome and Ebstein's anomaly. In fact, the presence of both anomalies may lead to corrective surgery earlier in the natural history of Ebstein's anomaly, thereby decreasing the overall operative risk. Although there were several hemodynamically unstable patients in this series, many of the patients presented for corrective arrhythmia surgery before hemodynamic deterioration from their Ebstein's anomaly forced surgical correction. Although this series includes all except two of the most serious cases referred to us, it was our policy surgically to ablate accessory pathways of patients with life-threatening or difficult-to-control arrhythmias regardless of whether severe functional cardiac disability related to Ebstein's anomaly was present. Corrective procedures for Ebstein's anomaly were performed when indicated by the functional symptoms of the patient as an associated procedure at the time of accessory pathway ablation. This policy may account, in part, for our relatively low operative mortality of 8.3% with tricuspid valve replacement, a rate that is approximately one-third that of other series24,25 of tricuspid valve replacements during a similar time frame. Carpenter et al26 noted the importance of hemodynamic factors and cardiac size as factors affecting mortality suggesting that earlier repair may decrease the risk of tricuspid valve replacement in patients with Ebstein's anomaly. Oh et al27 reported an increased tendency to perform tricuspid valve repair rather than valve replacement and an operative mortality of 7.7%. During the last 5 years of our series, tricuspid valve repairs were performed exclusively with no operative mortality.

Hansen et al28 suggested that corrective procedures for Ebstein's anomaly be performed at the appearance of heart failure because after the initial signs of failure, there is rapid deterioration, with death ensuing within a few years. He also reported the optimal timing of the operation to be when the patient is above the age of 15 years, although it may become mandatory earlier in patients with severe Ebstein's anomaly and hemodynamic compromise. An international cooperative study29 from the 1970s reported age to be a critical determinant of survival, with an overall surgical mortality rate of 60% for patients ages 1–15 years versus half that for patients over 15 years of age. Both of the patients with Ebstein's anomaly in our series experiencing operative mortality were under 15 years of age. Much of the increased perioperative mortality with younger patients may be related to a greater severity of Ebstein's anomaly that necessitates earlier surgical intervention and to the increased difficulty in performing surgical procedures on smaller hearts.

Atrial Fibrillation

There was a marked decrease in the occurrence of atrial fibrillation after surgical accessory pathway ablation in the patients with Ebstein's anomaly regardless of whether or not concomitant surgical repair of their Ebstein's anomaly was performed. This suggests that the etiology of atrial fibrillation in many of these patients may be related more to the presence of their accessory pathway than to primary or secondary atrial myocardial abnormalities seen with Ebstein's anomaly. These data are consistent with the significant decreases in atrial fibrillation observed in patients without Ebstein's anomaly after successful accessory pathway ablation as shown by Sharma et al16 and confirmed in this study.

Although ablation of the accessory pathway significantly decreases the frequency of long-term postoperative atrial fibrillation in the Ebstein's anomaly patient, atrial fibrillation still tends to occur more frequently than in patients with a normal heart. Depending on their functional class, patients with surgical ablation of their accessory pathway and Ebstein's anomaly had a two- to threefold increased risk of developing atrial fibrillation postoperatively compared with patients without Ebstein's anomaly. Thus, residual hemodynamic and structural abnormalities facilitate the occurrence of atrial fibrillation in some patients. Despite this, it is important to note that preoperative atrial fibrillation did not recur in 13 (87%) of the 15 patients with Ebstein's anomaly after accessory pathway ablation.

Long-term Survival and Quality of Life

Late results of Ebstein's anomaly patients undergoing corrective surgery are reportedly influenced by atrial and ventricular arrhythmias, conduction disturbances, degenerative changes in the artificial valves used for replacement and intrinsic primary myocardial disease.24,25 Ten-year survival was excellent in patients with and without Ebstein's anomaly, exceeding 90% in both groups. In patients undergoing concomitant tricuspid valve replacement, 10-year survival was 84%. Although the majority of patients with and without Ebstein's anomaly were alive and not disabled at follow-up, three patients with Ebstein's anomaly (9.4%) were on medical disability at the time of postoperative follow-up, and two additional patients not alive at follow-up had experienced disabilities before death. All of these patients had surgery after the appearance of significant hemodynamic compromise.

In patients with mild Ebstein's anomaly, survival and long-term disabilities approximate those observed in patients with the WPW syndrome alone. Although a positive association is seen between increasing severity of Ebstein's anomaly and mortality, incidence of arrhythmias, and prevalence of disabilities, patients with moderate-to-severe Ebstein's anomaly still demonstrate remarkably better quality of life after accessory pathway ablation. While it is also clear that a significant portion
of this enhancement in quality of life is due to correction of WPW-related arrhythmias, nearly three fourths of patients in the moderate-to-severe category also had simultaneous valve replacement, making it difficult to distinguish between improvements attributable solely to hemodynamic status versus those caused by abolition of WPW-related arrhythmias.

Most individuals view good health as a key component in overall quality of life. Consequently, there is an increasing trend toward examining illnesses and medical interventions in terms of their impact on the quality of life of the patient. The majority of patients in this study experienced cardiac arrhythmias of varying degrees of frequency and severity before surgery. Untreated or unsuccessfully treated, these arrhythmias provide interruptions in activities associated with everyday living, including participation in recreational, social, educational, and work-related endeavors. In addition, arrhythmias may be the source of considerable psychological and financial stress to the patients and their families. The significant decreases in the frequency and severity of arrhythmias, the frequency of hospitalizations, the need for antiarrhythmic drugs, and activity limitations appear to have significantly improved the quality of life of patients with and without Ebstein’s anomaly. Thus, patients with Ebstein’s anomaly and WPW syndrome, even those in whom the defect is moderate to severe, should not be denied the benefits of ablation of the accessory pathway.

**Baseline Electrophysiological Characteristics**

The electrophysiological differences between patients with and without Ebstein’s anomaly are consistent with previous reports that have detailed the unique electrophysiological properties of patients with this anomaly. Smith et al previously described the relatively long cycle lengths of orthodromic reciprocating tachycardia and ventriculo-atrial conduction times over the accessory pathway and the predominance of right and posteroseptal pathways in patients with Ebstein’s anomaly. In addition, multiple accessory pathways occurred in nearly half of the patients with Ebstein’s anomaly, indicating the need for careful endocardial and epicardial mapping of these patients. The most common combination was that of posteroseptal and right free wall pathways.

An interesting finding in the current study population was an Ebstein’s anomaly patient with a single left-sided pathway. This is a rare occurrence, although Becker et al reported a case of histological identification of a clinically unsuspected left posterior accessory pathway in an Ebstein’s anomaly patient with a second right anterolateral pathway. The diagnosis of a single left-sided accessory pathway in a patient with mild Ebstein’s anomaly seen in this study was made preoperatively from a preexcited ECG and confirmed by electrophysiological study and epicardial mapping. Less surprising, two additional patients with congenitally corrected transposition of the great vessels also had “left-sided pathways” to their systemic, but anatomic, right ventricles.

It was also unexpected that the incidence of preoperative atrial fibrillation was similar between patients with and without Ebstein’s anomaly and between patients with mild as compared with those with moderate-to-severe Ebstein’s anomaly in this series despite similar duration of symptoms of arrhythmias and age at time of surgery. Atrial fibrillation in WPW patients identified at electrophysiological study tends to originate in the right atrium regardless of pathway location, presumably in response to anatomic and functional properties of the right atria during reciprocating tachycardia. It was anticipated that the presence of Ebstein’s anomaly in patients with WPW would predispose to electrophysiological alterations in the right atrium that would facilitate the occurrence of atrial fibrillation. However, an increased risk of atrial fibrillation preoperatively was not seen in patients with versus those without Ebstein’s anomaly. Initially, it was believed that this may reflect the fact that half of the patients in this series had no or minimal functional sequelae from their Ebstein’s anomaly; however, there were no differences found in the incidence of documented atrial fibrillation between patients with mild and those with moderate-to-severe Ebstein’s anomaly.

**Classification of Ebstein’s Anomaly**

Ebstein’s anomaly patients in this study were grouped into two categories, mild or moderate-to-severe, according to whether or not they experienced function-limiting symptoms. It has been suggested that functional and hemodynamic status are more accurate predictors of surgical outcome in the Ebstein’s anomaly patient than anatomical malformations alone. Cabin et al noted prolonged survival in a patient who lived symptom free to 68 years of age with an anatomically severe form of Ebstein’s anomaly, and others have noted serious symptoms and/or sudden death in Ebstein’s anomaly patients lacking gross malformations of the heart. Because of the wide variations in the clinical course of the Ebstein’s anomaly patient, we chose functional disability as a more accurate representation of the current state of the disease. Combining the moderate and severe categories of Ebstein’s anomaly created the larger subgroup of 17 patients that provided for more meaningful data analysis. It also eliminated the problem of retrospectively categorizing patients with “moderately severe” Ebstein’s anomaly into a moderate or severe category. Classification into mild or moderate-to-severe could be made with confidence, whereas differentiating moderate and severe was less inherently clear from the retrospective data available.

**Study Limitations**

Recent data suggest that late electrophysiological study may have added additional information relative to return of accessory pathway function. However, because of the worldwide distribution of patients in this study and the cost of travel and repeat studies in a largely successfully ablated group of patients, it was neither feasible nor cost effective to obtain late follow-up electrophysiological studies in patients who were asymptomatic or who had singular, limited, and/or brief episodes of symptoms suggestive of arrhythmias. The majority of patients who experienced symptoms reported palpitations lasting less than 60 seconds. ECG recordings revealed premature atrial or ventricular contractions corresponding to symptoms in most cases, or the palpitations were of such negligible concern that further evaluation was not pursued by the patient and/or his physician. Furthermore, the electrophysio-
logical study is not a specific method for confirming the diagnosis of atrial fibrillation or atrial flutter unrelated to the Wolff-Parkinson-White syndrome, which explained a large portion of sustained palpitations in our study and may likewise have explained those patients reporting sustained symptoms without residual accessory pathway function in the study by Davis et al.

It is possible that during long-term follow-up, the use of antiarrhythmic drugs for symptomatic atrial or ventricular ectopy or for arrhythmias unrelated to the Wolff-Parkinson-White syndrome may have also suppressed recurrence of arrhythmias in some cases of undiagnosed accessory pathway function. However, because the practice occurred in a minority of study patients and in a similar percent of patients in each group, an effect, if any, should be uniform across treatment groups and would not be expected to change the findings of this study.

Conclusions

Nearly three fourths of Ebstein's anomaly patients also underwent an associated surgical procedure related to their Ebstein's anomaly at the time of accessory pathway ablation. The recent development of the closed-chest technique of radiofrequency catheter ablation is likely to benefit those patients with mild disease and no indication for structural repairs; however, the results of this study are less generalizable for that group of patients whose WPW-related arrhythmias precipitated early surgical correction of their Ebstein's anomaly. Further studies are needed to determine the optimal role of surgical and catheter ablation therapies in patients with Wolff-Parkinson-White syndrome who have variable degrees of severity of Ebstein's anomaly.

Acknowledgments

We would like to thank Bettie McCade for assistance in preparation of the manuscript, Benjamin Burks for assistance in patient follow-up, and Laura Cook, RN, for her care of these patients in the electrophysiology laboratory.

References

Effect of Ebstein's anomaly on short- and long-term outcome of surgically treated patients with Wolff-Parkinson-White syndrome.

J C Pressley, J M Wharton, A S Tang, J E Lowe, J J Gallagher and E N Prystowsky

Circulation. 1992;86:1147-1155
doi: 10.1161/01.CIR.86.4.1147

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/86/4/1147