Advantages and disadvantages of one-stage and two-stage surgery for arrhythmias and Ebstein’s anomaly

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Abstract

Objective: To evaluate efficacy of one-step and two-step surgery approach in patients with arrhythmias combined with Ebstein’s anomaly.

Methods: Fifty-three patients with Ebstein’s anomaly combined with tachyarrhythmias (58.5% men, 41.5% women, mean age 21.6 ± 10.7 years) were operated on. In group A (32 patients), one-step surgical correction (simultaneous intraoperative elimination of arrhythmias and congenital heart defect repair) was performed, whereas in group B (21 patients), two-step surgery was performed with initial elimination of arrhythmogenic substrate by transcatheter radiofrequency ablation (first step) and following surgical repair of congenital heart defect (second step).

Results: In group A, total hospital mortality was 3.1% (1 patient) due to initial severe condition of this patient. One-step surgery was effective in 93.5% of cases. Mortality was not observed in group B. Efficacy of transcatheter radiofrequency ablation was 76.2%.

Conclusions: one-stage and two-stage surgery of arrhythmias and Ebstein’s anomaly are highly effective. First step of surgery of combined pathology reduces cardiopulmonary bypass time, complications and mortality while performing the second step of congenital heart defect surgery. However, simultaneous approach (one-step surgery) is better in terms of arrhythmia elimination.

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Keywords: Ebstein’s anomaly; Congenital heart defect; Arrhythmias; Wolff-Parkinson-White syndrome; Open heart surgery; Radiofrequency ablation

1. Introduction

Congenital heart diseases (CHD) are very frequently combined with cardiac arrhythmias. Accessory atrio-ventricular (AV) pathways are revealed in 6-24% of patients with Ebstein’s anomaly, double AV pathways—in 15-20% of them [1-3]. Hence, if successful surgery of CHD is not completed by arrhythmogenic substrate elimination, there is no guarantee of avoiding postoperative rhythm disorders and sudden cardiac death in patients with the combined pathology [4]. Sudden cardiac death and tachyarrhythmias (TA) are registered in 14% of patients with CHD despite adequate antiarrhythmic therapy [Sabirov 1995, personal communication]. Implementation of open methods of surgical correction of arrhythmias allowed extending indications for simultaneous surgery of CHD and rhythm disorders. It resulted in reduction of mortality and improvement of surgery. Introduction into clinical practice of closed methods of arrhythmia elimination and their enhancement made it possible to use them for transvasal destruction of arrhythmogenic area as a first step of surgery in patients with the combined pathology.

The aim of our study was to evaluate advantages and disadvantages of simultaneous and stage-by-stage surgical correction of Ebstein’s anomaly and TA.

2. Materials and methods

Fifty-three patients with Ebstein’s anomaly and concomitant TA (58.5% men, 41.5% women), at the age of 7-46 years (mean age 21.6 ± 10.7 years), were operated on in Arrhythmology Department of Bakoulev Scientific Center for Cardiovascular Surgery during the period 1995-2003.

We have been performing radiofrequency catheter ablation (RFA) of accessory AV pathways before the surgery of Ebstein’s anomaly since 1995. Hence, all patients were divided into two groups:

- Group A—surgical correction of Ebstein’s anomaly and TA was performed simultaneously (32 patients);
- Group B—TA was eliminated by RFA, as a first stage in the stage-by-stage treatment of the combined pathology (21 patients).
Patients of these two groups differed by the severity of hemodynamical disorders caused by aforementioned CHD. In group A, 65% of patients were admitted to the hospital because of severe clinical condition attributable to Ebstein’s anomaly, whereas 35% of patients were admitted to the hospital due to complaints associated with rhythm disorders. In group B, leading causes of hospitalization were cardiac arrhythmias, while symptoms determined by Ebstein’s anomaly were insignificant. During the interval between arrhythmia occurrences hemodynamical and clinical disorders down to Ebstein’s anomaly were mild, therefore quality of life of these patients was almost normal.

On admission to the hospital patients of both groups suffered from fatigue, rapid palpitation and breathlessness. Thirty percent of patients from group A had edema additionally to aforementioned symptoms, 31% had liver enlargement and 28% had cyanosis. Most patients of group A were in NYHA II-III functional class, while in group B patients were mostly in NYHA I-II (Fig. 1). Ebstein’s anomaly was combined with different types of tachyarrhythmias, most frequently with Wolff-Parkinson-White syndrome (Table 1). Average duration of history of cardiac arrhythmias was 9 years (8.9 ± 8.69 years). In 17 patients (belonged to groups A and B), TA episodes were accompanied by syncopes, 2 patients survived sudden death and in 2 other patients it was essential to use electrical cardioversion for arrhythmia elimination.

For verification of CHD and arrhythmias in order to subsequently select appropriate surgical tactics, all patients underwent following investigational procedures: standard ECG, X-ray, echocardiography, 24 h holter monitoring, multichannel body surface potential mapping, transcatheter and intraoperative electrophysiological study. During echocardiography most important targets for examination were: diameter of atrIALIZED part of right ventricle (RV), diameter of intrinsic tricuspid valve (TV) annulus, displacement of tricuspid valves towards RV apex, degree of TV insufficiency (Fig. 2). Echo characteristics of patients of both groups were mainly as follows: average TV insufficiency 2.8 ± 0.8 (from grades 1-4) and mean RV ejection fraction 45.2 ± 6.4% (from 35 to 58%). In group B, right heart chamber characteristics were less than in group A, what indicated less severe abnormalities in this group (Table 2). In 11 patients atrial septal defect was revealed, in 6 patients patent foramen ovale and in 1 patient perimembranous ventricular septal defect was revealed (these 18 patients belonged to groups A and B).

Table 1

<table>
<thead>
<tr>
<th>Types of TAs</th>
<th>Number of patients</th>
<th>Total number</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wolff-Parkinson-White syndrome</td>
<td>26</td>
<td>43</td>
</tr>
<tr>
<td>AV nodal re-entrant tachycardia</td>
<td>3</td>
<td>7</td>
</tr>
<tr>
<td>Ectopic supraventricular tachycardia</td>
<td>3</td>
<td>3</td>
</tr>
<tr>
<td>All</td>
<td>32</td>
<td>53</td>
</tr>
</tbody>
</table>

Main indications for surgical treatment of combined pathology were: heart failure due to Ebstein’s anomaly and/or arrhythmias, long lasting tachyarrhythmias resistant to medical therapy, hemodynamically significant rhythm disorders (history of syncopes), history of at least one episode of ventricular fibrillation, paroxysmal atrial fibrillation with fast antegrade conduction (ventricular rate ≥ 200 beats/min).

3. Statistics

Data are expressed as the mean value ± SD. The Student paired t-test was used to compare two continuous variables. For differences, a value of $P<0.05$ was considered statistically significant.

The event-free survival was estimated by the Kaplan-Meier method, and differences among groups were assessed by means of the long-rank test. The chi-square and Fisher exact tests were used to compare qualitative variables between the two groups. All tests were two-tailed, and $P\leq 0.05$ was considered statistically significant.

Quality of life was assessed by Short-Form-36 (SF-36) questionnaire.

4. Results

Among revealed Wolff-Parkinson-White (WPW) syndrome cases, 64% were manifested type, 14.3% were concealed type, 14.3% were intermittent type. In 2 patients, Maheim path occurred. Right side location of AV accessory pathways was observed in 23% of cases, posterior was observed in
26.6% of cases, posterior-septal was observed in 36% of cases and anterior-septal was observed in 9.5% of cases. Only in 2 patients accessory pathways had left side location, herewith in one case Ebstein’s anomaly was combined with corrected transposition of great arteries. Multiple AV accessory pathways were revealed in 28.5% of patients.

In group A multichannel epicardial mapping with subsequent arrhythmogenic substrate elimination was performed as a first step of surgery of the combined pathology. In patients with WPW syndrome elimination of AV accessory pathways was carried out by Sealy procedure [9]. In 5 patients with right-posterior location of AV accessory pathways, Sealy operation was completed by cryoablation of arrhythmogenic substrate. Surgical isolation of AV node was used in patients with AV nodal tachycardias. Surgical excisions of arrhythmogenic area or its cryoablation were applied in patients with ectopic atrial tachycardias (Table 3). CHD correction (TV replacement by bioprosthesis and atrial septal defect closure with a patch) was conducted after arrhythmia elimination.

In group B with 70% of patients under the age of 20, insignificant hemodynamic abnormalities due to Ebstein’s anomaly allowed using RFA for arrhythmia elimination as a first step of surgery. RFA was performed under permanent monitoring of heart rhythm, PQ interval, power, temperature, impedance-controlled energy delivery and fluoroscopy supervision (Fig. 3).

Efficacy of accessory pathways elimination was checked out: in one-stage surgery—after the total recovery of the heart at the end of surgery, and in stage-by stage surgery—right after RF energy application and at the end of RFA procedure.

Surgical correction of Ebstein’s anomaly in 8 patients from group B was performed after successful elimination of TA by RFA in 3 months–2.5 years period. Mean cardiopulmonary bypass (CPB) and aortic cross-clamping time during this type of surgical approach were significantly lower than during simultaneous surgery (P < 0.0001). In two-stage surgery of TA and Ebstein’s anomaly mean CPB time and aortic cross-clamping time were 88.5 ± 10.2 and 54.3 ± 10.3 min, respectively, whereas during one-stage surgical correction of combined pathology same parameters were as follows: 118.8 ± 29.9 and 74.4 ± 18.5 min, respectively. Thus, time difference in CPB was 25–30 min and in aortic cross-clamping the time difference was 15–20 min.

In group A, hospital mortality was 3.1% (1 patient). It should be mentioned that this patient had an initial severe myocardial dysfunction as a result of long lasting Ebstein’s anomaly and TA (ectopic supraventricular tachycardia). Main reason of death was acute heart failure in intra- and

### Table 2

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>Group A</th>
<th>Group B</th>
<th>P</th>
</tr>
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<tbody>
<tr>
<td>Right atrium diameter, mm</td>
<td>45–80 (63.9 ± 11.2)</td>
<td>44–68 (56.7 ± 6.4)</td>
<td>&lt; 0.05</td>
</tr>
<tr>
<td>Dislocation of tricuspid valves, mm</td>
<td>10–60 (33 ± 14.3)</td>
<td>13–38 (25.2 ± 7.7)</td>
<td>&lt; 0.05</td>
</tr>
<tr>
<td>Tricuspid valve annulus diameter, mm</td>
<td>45–62 (53.6 ± 5.2)</td>
<td>32–62 (43.2 ± 10.7)</td>
<td>&lt; 0.05</td>
</tr>
<tr>
<td>Cardiotoracic index, %</td>
<td>46–62 (55.7 ± 5.4)</td>
<td>45–69 (52.7 ± 6.4)</td>
<td>&lt; 0.05</td>
</tr>
</tbody>
</table>

### Table 3

<table>
<thead>
<tr>
<th>Types of antiarrhythmic surgery</th>
<th>Number of patients (n)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sealy operation and its modification</td>
<td>21</td>
</tr>
<tr>
<td>Sealy operation and its modification+cryoablation</td>
<td>5</td>
</tr>
<tr>
<td>Arrhythmogenic substrate cryoablation</td>
<td>1</td>
</tr>
<tr>
<td>AV node surgical isolation</td>
<td>3</td>
</tr>
<tr>
<td>Surgical excision of arrhythmogenic zone</td>
<td>2</td>
</tr>
<tr>
<td>Total</td>
<td>32</td>
</tr>
</tbody>
</table>

Fig. 3. X-ray and Electrograms during RFCA of right-sided posterior-septal AV accessory pathways.
patients with and without CHD [5,6]. However, in CHD substrate identification and RFA technique do not differ in anomaly and concomitant arrhythmias. Arrhythmogenic become first-line therapy for patients with Ebstein’s revolutionized treatment of tachyarrhythmias and has date back to 1986 [6,7,10–13]. Since that time RFA has correction of combined pathology. First reports about efficiency of simultaneous and stage-by-stage surgical protocol), what allowed postponing performance of major manipulations was 8.6 (from 2 to 30). There were no complications except TA recurrences.

In this study in group B RFA was efficient in 76.2% of patients. These patients reported improvement in quality of life, increase of exercise tolerance (assessed by BRUCE protocol), what allowed postponing performance of major surgery for CHD for a best possible period of time. Among five unsuccessful RFA, in three cases multiple AV accessory pathways were revealed.

Thus, it can be assumed that one-stage and two-stage surgery for arrhythmias and Ebstein’s anomaly are highly effective. However, TA recurrences are observed in both groups; they mostly occur in patients with multiple and septal AV accessory pathways.

5. Discussion

There are different opinions concerning rationality and efficiency of simultaneous and stage-by-stage surgical correction of combined pathology. First reports about transcatheter elimination of WPW syndrome in CHD patients date back to 1986 [6,7,10–13]. Since that time RFA has revolutionized treatment of tachyarrhythmias and has become first-line therapy for patients with Ebstein’s anomaly and concomitant arrhythmias. Arrhythmogenic substrate identification and RFA technique do not differ in patients with and without CHD [5,6]. However, in CHD patients number of RF energy delivery manipulations, their duration and fluoroscopy time are higher than in patients without CHD, due to technical difficulties caused by cardiac anatomical abnormalities. For instance, in some studies average number of RF energy delivery manipulations during RFA of isolated AV accessory pathways was 5 and fluoroscopy time was 52 min, whereas in combined pathology (CHD with concomitant TA) average number of RF energy delivery manipulations was 6–9 (ranged from 1 to 44), mean fluoroscopy time was 70 min and mean duration of whole procedure was 7.5 h [6,7].

RFA for AV accessory pathways elimination is generally less efficient in patients with CHD, than in patients without CHD [8]. In both categories of patients in this study TA recurrences were observed mostly after RFA of multiple (21%) and septal (17%) AV accessory pathways. TA recurrences occurred more often in patients with corrected transposition of great arteries (67%) and then in patients with Ebstein’s anomaly (23%). That was a result of technical difficulties during RFA due to those complex CHD. [8] Based on literature data TA recurrences after RFA in patients with Ebstein’s anomaly occur in 7–30% of cases, what is generally explained by the cautious performance of this procedure in order to avoid complete AV block [4,7,12].

Some investigators obtained positive results of simultaneous surgery for complex CHD and arrhythmias both in early and late postoperative periods [14,15]. Although, Van Hare [4] considers that in patients with Ebstein’s anomaly preoperative RFA of AV accessory pathways is more preferable than surgical excision of arrhythmogenic area. In these cases the CHD itself does not preclude from arrhythmia elimination and there is vivid improvement in patients condition afterwards [16]. Hebe and co-authors [6] conducted simultaneous surgery only in severe patients necessitating urgent surgical correction of CHD and having high possibility of developing complications during closed procedures. In cases, when immediate surgery of CHD was not required, they usually carried out RFA of arrhythmogenic substrate as a first step of surgical approach. In cases of initially failed RFA of TA in patients with Ebstein’s anomaly and TA, surgical treatment of combined pathology is indicated in time-interval, individual for each patient, depending on CHD and arrhythmia severity.

Intracardiac recordings and RFA are complicated in CHD patients due to atypical location of the conduction system of the heart. However, considering these facts and individual approach to cardiac anatomy of each patient helps to minimize risk of accidental damage of conduction system and thus increase the efficacy of the procedure [7].

Advantage of RFA is determined by its ability to be repeated several times with less risk for the patient compared to re-do ‘open heart’ surgery. RFA of arrhythmogenic area reduces cardiopulmonary bypass time, complications and mortality while performing surgical correction of CHD as a second step of surgery of combined pathology.

When choosing different options of surgery in patients with Ebstein’s anomaly and concomitant TA, the following factors should be taken into account: patients current clinical condition, level of anatomical and hemodynamic changes caused by the CHD and patients age.

Table 4 Efficacy of simultaneous surgery (in early postoperative period) depending on tachyarrhythmia type

<table>
<thead>
<tr>
<th>Types of Tachyarrhythmias</th>
<th>Number of patients</th>
<th>Efficacy (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wolff–Parkinson–White syndrome</td>
<td>26</td>
<td>92.3</td>
</tr>
<tr>
<td>AV nodal re-entrant tachycardia</td>
<td>3</td>
<td>100</td>
</tr>
<tr>
<td>Ectopic supraventricular tachycardia</td>
<td>2/1</td>
<td>66.6</td>
</tr>
<tr>
<td>All</td>
<td>31/1*</td>
<td>93.5</td>
</tr>
</tbody>
</table>

* Success rates are calculated for 31 patients because 1 patient died after surgery due to progressive RV dysfunction and ectopic supraventricular tachycardia recurrence.
6. Conclusions

- Simultaneous and stage-by-stage surgical correction of arrhythmias in patients with Ebstein’s anomaly both allow achieving positive antiarrhythmic results; one-stage surgery is indicated for patients with tachyarrhythmias and a progressive heart failure due to Ebstein’s anomaly; ‘Open’ surgery is more effective than radiofrequency catheter ablation, though performance of the latter enables to reduce cardiopulmonary bypass time in high risk patients; in cases of frequent preoperative ‘malignant’ tachyarrhythmia events and lack of clinical manifestations of Ebstein’s anomaly, radiofrequency catheter ablation is more preferable as a less traumatic, relatively safe and a fairly effective procedure, which allows postponing surgical repair of Ebstein’s anomaly for a best possible period of time.

References


Appendix A. Conference discussion

Dr J. Melo (Carnaxide, Portugal): What have you observed in those patients with the two-stage approach regarding the lesions induced by radiofrequency? Were they located in the exact and precise place where they are expected to be? Were there scars, clots? Can you comment on your findings?

Dr Bockeria: That is really a very good question for surgeons, because when you open the heart after, let’s say, one or two weeks after radiofrequency ablation you really see a huge area looking like a scar. In fact, a scar, and normally it’s located in between the coronary sinus orifice and the area where the posterior leaflet of the tricuspid valve should be located. Sometimes the scar area is really very big and partially occupies the posterior wall of the right ventricle. That view is not attractive.

Dr Lorenzo Menicanti (Segrate, Italy): Did you consider the economical cost of these two types of strategies?

Dr Bockeria: No, we did not because we are a poor country, so it doesn’t matter. But it is definitely much cheaper to do surgery, of course, because the equipment and disposables for radiofrequency ablation is really very expensive. But it is paid by the patient and not by the Ministry.