Surgical treatment of Wolf–Parkinson–White syndrome during plastic operations in patients with Ebstein’s anomaly

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Abstract

Objective: Ebstein’s anomaly is the most common pathology associated with the accessory conduction pathways. Methods: From January 1990 to August 1999 48 patients underwent surgical repair of Ebstein’s anomaly by various plastic techniques. The pathways were identified and characterized at preoperative electrophysiologic mapping in 17 (34.5%) patients. There were seven males and ten females. The patients’ age ranged 6–35 years (mean 12.7 ± 2.1 years). Five patients were in NYHA class II and were in 12 in NYHA class III. The cardiothoracic ratio ranged from 0.59 to 0.69 (mean 0.65 ± 0.08). Tachycardia was present in 15 patients with Wolf–Parkinson–White (WPW) syndrome. Atrial septal defect was present in 12 patients (70.6%). Accessory conduction pathways were in the right posterior septal area in seven patients, in right posterior septal area and free wall in seven patients and in right free wall in three. The pathways were successfully ablated in all patients during cardiopulmonary bypass. Results: There were no deaths, no relapses. Follow-up ranged from 4 months to 7 years (mean 4.7 ± 1.1 years). There were no late deaths, but one patient required successful tricuspid valve replacement because of severe tricuspid insufficiency. At follow-up 57.8% of patients were in NYHA class I. Conclusions: Preoperative electrophysiologic study allows to identify the accessory conduction pathways in patients with Ebstein’s anomaly. The combined approach of tricuspid valve repair and surgical ablation of accessory atrioventricular connections has been proved safe and effective. This operative procedure allows to improve functional results in patients with Ebstein’s malformation and concomitant accessory conduction pathways. © 2000 Elsevier Science B.V. All rights reserved.

Keywords: Ebstein’s anomaly; Wolf–Parkinson–White syndrome; Plastic operations; Accessory pathways

1. Introduction

Ebstein’s anomaly (EA) is a complex and relatively rare heart malformation (approximately 1% of all congenital heart diseases). The essential anatomic defect of EA is the displasia and hypoplasia of the tricuspid valve (TV), which causes a marked right ventricular failure.

Carpentier classification of this pathology is widely used in the clinical practice [1]. This classification is based on the mobility of the anterior leaflet of TV and on the contractility of the right ventricle (RV).

Different operative procedures were proposed for the total correction of TV dysfunction in patients with EA, including plastic operations or TV replacement [2–4].

Wolf-Parkinson-White (WPW) syndrome accompanies EA more frequently than any other congenital heart disease – it is seen in 10–29% of patients with this pathology [5]. Inherent to this syndrome tachycardias not only worsen patient’s clinical status and natural course of the disease, but also aggravate an early postoperative period. In connection with this the question of a simultaneous correction of the lesion and elimination of arrhythmia acquires especial significance. Fulfilment of that stage of the operation that deals with arrhythmia elimination prolongs the extracorporeal circulation time and at present time when in the majority of the patients with WPW syndrome accessory conduction pathways usually can be eliminated with help of ablation catheter technique [6], the question arises about advisability of a simultaneous elimination of accessory pathways during the TV correction.

To clarify this question we analyzed our experience of the plastic procedures in patients with AE and concomitant WPW syndrome.
2. Material and methods

From January 1990 to August 1999 a surgical correction of AE was performed in 48 patients using different plastic methods.

All patients, in spite on presence or absence of complaints on heart palpitations in their previous history, underwent electrophysiological studies (EPI). As result of EPI 24 accessory conduction pathways were found in 17 (34.5%) of 48 patients with EA. The degree of preoperative tricuspid valve incompetence is shown in Table 1. There were seven males and males females. The patients ages ranged from 6 to 35 years (mean 12.7 ± 2.1 years). Five patients were in NYHA class II and 12 were in NYHA class III. The cardio-thoracic ratio ranged from 0.59 to 0.69 (mean 0.65 ± 0.08). Fifteen patients had in their past history fits of tachycardia; in two patients with accessory conduction pathways tachycardia was absent. Accessory conduction pathways were localized in the following manner: in seven patients in the right posterior septal area; in seven in the right posterior septal and free wall area; in three patients in the free wall. During the EPI a detailed study was performed with the aim to specify the mechanisms of tachycardia and localization of accessory atrioventricular connections. Twenty-one of 24 accessory pathways possessed conductivity in an antegrad and in retrograde directions Two had conductivity only in a retrograde (ventriculo-atrial) direction and one only in an antegrad direction. Orthodromic atrioventricular tachycardia with accessory conduction pathways participation was provoked in 16 patients.

Longitudinal plication of the atrialized part of the RV was used in all cases together with the certain method of annuloplasty procedure: in one patients with A type of pathology the De Vega method was used, in two patients with the same type of pathology our own method of valve reconstruction was used (septal leaflet enlargement with the help of pericardial patch); the Danielson method was used in eight patients and the Carpentier method in six patients. All operations were performed with the use of extracorporeal circulation and moderate hypothermia (24–27°C). Cardiopulmonary bypass time ranged from 58 to 132 min (mean 92 min) and aortic cross-clamping time from 26 to 93 min (mean 45 min.).

Accessory pathways localization during operation was determined exclusively on the base of preoperative EPI data. Accessory pathways ablation in the first four patients was performed according to the routine method [7] by the way of atrial myocardium incision with the following tissue dissection of an atrio-ventricular groove on the length of its all right heart contour, and, if it’s necessary, with the dissection of the posterior pyramidal space on the arrested heart in conditions of cardioplegia. Later on we refused from this extensive manipulation, and in the last 13 patients we limited zone of the dissection to 1.5 cm even in cases with multiple accessory conduction pathways. We performed pacing and recording with the use of temporary wire electrodes placed on a right atrium and a right ventricle to assess an effectiveness of accessory pathway ablation immediately at the operating room and later before discharge on the 7th–9th day postoperatively. All 17 accessory conduction pathways were ablated from the first attempt and in no case it was necessary to use extracorporeal circulation for the second time in order to ablate pathways. Time used for accessory conduction pathways ablation from the beginning of atrial myocardium incision and to the completion of the suture line on the site of this incision, ranged from 9 to 20 min (mean 15 ± 3.0 min).

3. Results

There were no hospital deaths and relapses.

Follow-up ranged from 4 months to 84 months (mean 55 ± 13 months). There were no late deaths. Postoperative function of the tricuspid valve was assessed with the help of echo-Doppler in all 17 survived patients. The extent of residual or recurrent tricuspid valve incompetence is shown in Table 2.

One patient underwent successful TV replacement because of severe tricuspid incompetence.

The functional improvement in remote postoperative period (from 4 months to 84 months (mean 55 ± 13 months)) is shown in Table 3.

In no one case we observed atrioventricular block. There were no cardiac rhythm disturbances in the remote period in all 17 patients whom accessory pathways ablation was performed. No one of them did not take antiarrhythmic drug treatment.

| Table 1 | Grades of preoperative tricuspid valve incompetence |
|-----------------|-----------------|-----------------|-----------------|-----------------|
| Grade of TV insufficiency | 0 | 1+ | 2+ | 3+ |
| Patients (n = 17) | – | 1 | 7 | 9 |
| % | 5.8 | 41.2 | 53 |

| Table 2 | Grades of postoperative tricuspid valve insufficiency |
|-----------------|-----------------|-----------------|-----------------|-----------------|
| Grade of TV insufficiency | 0 | 1+ | 2+ | 3+ |
| Patients (n = 17) | 8 | 6 | 2 | 1 |
| % | 47 | 35.3 | 11.8 | 5.9 |

| Table 3 | Dynamics of NYHA functional class after AE correction 4.7 ± 1.1 years postoperatively |
|-----------------|-----------------|-----------------|-----------------|-----------------|
| NYHA Class | I | II | III | IV |
| Patients (n = 17) | 9 | 6 | 1 | 1 |
| % | 53 | 35 | 6 | 6 |
4. Discussion

Patients with EA are predisposed to heart arrhythmias because of abnormal cardiac anatomy [8,9]. For patients with tachyarrhythmias there is a typical tendency to cardiac hemodynamic decompensation. Heart rhythm disturbances together with RV dysfunction, tricuspid insufficiency and an atrial septal defect lead to the increase right-to-left blood shunting, hypoxemia, and reduced cardiac output [10]. Thus, anatomical and electrophysiological changes in patients with EA are interconnected factors of the hemodynamic disturbances gravity and of the clinical course of the disease.

Preoperative electrophysiological study becomes essential in patients with EA, it should include documentation of the mechanisms of tachycardia and localization of atrioventricular connections, both of which are important in surgical planning.

Successful development of the catheter ablation with the help RF current energy essentially had changed the role of a surgical methods in the treatment of arrhythmia, connected with the presence of accessory conduction pathways. At present time there are two alternative tactics of patients treatment with EA and concomitant WPW syndrome: surgical ablation of accessory pathways during the TV reconstruction or its replacement or the catheter ablation of pathways before operation. It is not expedient to postpone accessory pathways elimination on the second stage after EA repair, because arrhythmia can seriously complicate the course of operation itself as well as the course of an early postoperative period. Comparison of the efficiency and of the safety of each of the mentioned methods will help to choose the preferable variant of the treatment tactics.

Unlike catheter ablation of accessory conduction pathways in patients without concomitant pathology, where the efficiency results exceed 95% [5], results of catheter ablation of pathways in patients with EA are noticeably worse (76%) [11,12]. To a considerable extent this can be explained by difficulties in the determination of the fibrous ring position TV on the base of a standard electrophysiological criteria. This obstacle is absent, when there is possibility of a visual control.

We eliminated accessory conduction pathways in all patients with EA, and in remote period we observed no conduction recurrence through this pathways. In that way efficiency of operative elimination of accessory pathways in cases of EA is at present day superior to catheter ablation.

The main shortcomings of the surgical elimination of accessory conduction pathways is comparison with the catheter ablation is bigger trauma, as sternotomy is needed for its fulfillment, as well as cardiotomy and extracorporeal circulation. In cases where surgical elimination of accessory pathways is performed as the kind of an additional stage of the operation of a total EA correction, the same surgical approach and the same extracorporeal circulation is used for both pathways elimination and plastic procedure on TV. Thus, a single ‘negative’ moment introduced by accessory conduction pathways elimination, came to a certain prolongation of the aortic cross-clamping time. In our series of observations the time spent on ablation ranged from 9 to 20 min (mean 15 ± 3 min), this hardly can be considered as a serious negative influence on the course of operation. We did not observe any complications which were directly connected with the manipulation of accessory conduction pathways elimination.

We conclude that preoperative EFI allows to identify the accessory conduction pathways in patients with EA, and it has to become a routine method. Tricuspid valve repair and concomitant surgical ablation of accessory conduction pathways as a single operative procedure allows to improve functional results in patients with Ebstein’s malformation.

References

Appendix A. Conference discussion

**Dr S. Chauvaud (Paris, France):** Our series in Paris with Professor Carpentier, in 140 patients operated on with the technique, which includes detachment of the anterior leaflet and longitudinal plication of the right ventricle, we were able to reduce rhythm disturbances from 40 to 5% without specific surgery of accessory pathways. So I don’t think that specific surgery is necessary in Ebstein’s anomaly.

My question is, how long, in time consuming are the detection of the accessory pathways and their treatment by radio frequency.

**Dr Lazorichinets:** In our institute we don’t have equipment for radiofrequency ablation, and we make surgical ablation in operating room for all cases.

**Dr R. Benetis (Kaunas, Lithuania):** It is big material about Ebstein’s anomaly in conjunction with the WPW syndrome and thanks for your work.

I have a question. How many patients from your series there were with atrial fibrillation and how many patients had supraventricular arrhythmias in conjunction with the accessory pathways?

And the second question is, I still didn’t get information what kind of surgical techniques or ablation techniques you have used on your patients.

**Dr Lazorichinets:** I don’t have now data for paroxysmal tachycardia or atrial tachycardia now. And our surgical technique is very simple. For right side excuse me, I would like to show you my slide with this technique for surgical ablation. I don’t have this slide.

**Dr D.M. Cosgrove (Cleveland, OH, USA):** Why don’t you just describe your technique in lieu of not having the slide.

**Dr Lazorichinets:** Okay.

**Dr Cosgrove:** In the meantime are there other questions?

**Dr Benetis:** I withdraw my question.