Electrophysiological Study in Ebstein's Anomaly With no Evidence of Accessory Pathway

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Introduction

Ebstein's anomaly (EA) is characterized by low implantation of the tricuspid valve, and corresponds to less than 1% of all congenital heart defects[1-3]. It is often associated with other malformations, such as ventricular septal defect, and Wolff-Parkinson-White syndrome often accompanies.

Its clinical manifestation depends on the severity of the anatomical changes, and the indications for surgery have been well established[4,5].

When anomalous pathways are associated with EA, in one third of the cases the electrocardiographic pattern can differ from the classic one (short PR interval and presence of delta wave), because of slow intra-atrial conduction or anomalous pathway of long and slow conduction[6].

Arrhythmias are usually present in up to 80% of the patients[7], with prevalence of anomalous atrioventricular pathways ranging from 0 to 44%[1-3]. Despite the universal acceptance of electrophysiological study (EPS) in symptomatic patients or in those with apparent accessory pathway on the electrocardiogram (ECG), data on asymptomatic patients with no electrocardiographic evidence of pre-excitation are scarce in the literature.

Because of postoperative implications (difficult access to certain left atrial areas after surgical repair, risk of tachyarrhythmias in the intra- and postoperative periods, and likelihood of intraoperative elimination of accessory pathway), we report a case in which preoperative EPS was performed in a patient with EA and no classical evidence of tachyarrhythmias.

Case Report

The patient is a 43-year-old female, who sought the emergency service with findings of ischemic stroke. She reported progressive dyspnea, orthopnea and atypical chest pain for four years. On physical examination, in addition to neurological changes, a systolic murmur was audible on all valve areas (++/4). The ECG showed right bundle-branch conduction disorder and no evidence of ventricular pre-excitation (Figure 1 - II). Chest X-ray showed significant cardiomegaly (Figure 1 – I). The echocardiogram identified EA with moderate tricuspid regurgitation, 19-mm *ostium secundum* atrial septal defect (ASD), and left ventricular ejection fraction of 64%. The diagnosis of paradoxical embolism was presumed, and the surgical repair of EA and ASD was indicated.

Preoperative EPS was performed after assessing the possible deleterious consequences of not identifying an anomalous pathway, such as higher risk of perioperative severe arrhythmias, missed opportunity of intraoperative ablation in case of refractoriness, and more difficult percutaneous access to certain areas of the tricuspid ring after surgical repair. During atrial stimulation, sudden incremental conduction through an anomalous pathway occurred with slow atrioventricular conduction positioned at the posteroseptal region of the tricuspid ring. Antidromic and orthodromic tachycardias were induced, the later with right bundle-branch block (RBBB) pattern. After ablation, at the atrioventricular fusion site, ventricular pre-excitation disappeared, and classical RBBB appeared (Figure 2). Elimination of the accessory pathway was confirmed with adenosine.

Surgery was performed for ASD and EA repair using the cone technique. The patient was discharged asymptomatic on the sixth postoperative day.

Discussion

This case report raises the hypothesis that performing preoperative EPS might benefit patients with EA and no evidence of anomalous conduction pathway. The patient reported no palpitations and her ECG did not suggest pre-excitation.

A peculiar finding was the lack of RBBB, revealed after ablation with the elimination of the accessory pathway. This suggests the presence of accessory pathway of slow atrioventricular conduction, contributing to QRS complex depolarization, a natural ventricular resynchronization form usually seen in patients with EA[6]. Iturralde et al[6] have found that 38% of the patients with EA and documented supraventricular tachycardia had a minimal or questionable evidence of ventricular pre-excitation on baseline ECG, and 100% of that group had no RBBB pattern. On the other hand, 93% of the patients with EA of the control group, with no evidence of tachycardia, had RBBB. Absence of that on V1 on baseline ECG showed 98% sensitivity and 92% specificity for the anomalous pathway diagnosis[6]. Thus, finding an accessory pathway is highly probable when performing an EPS.

Keywords

Ebstein Anomaly / surgery; Cardiac Electrophysiology; Embolism, Paradoxical.

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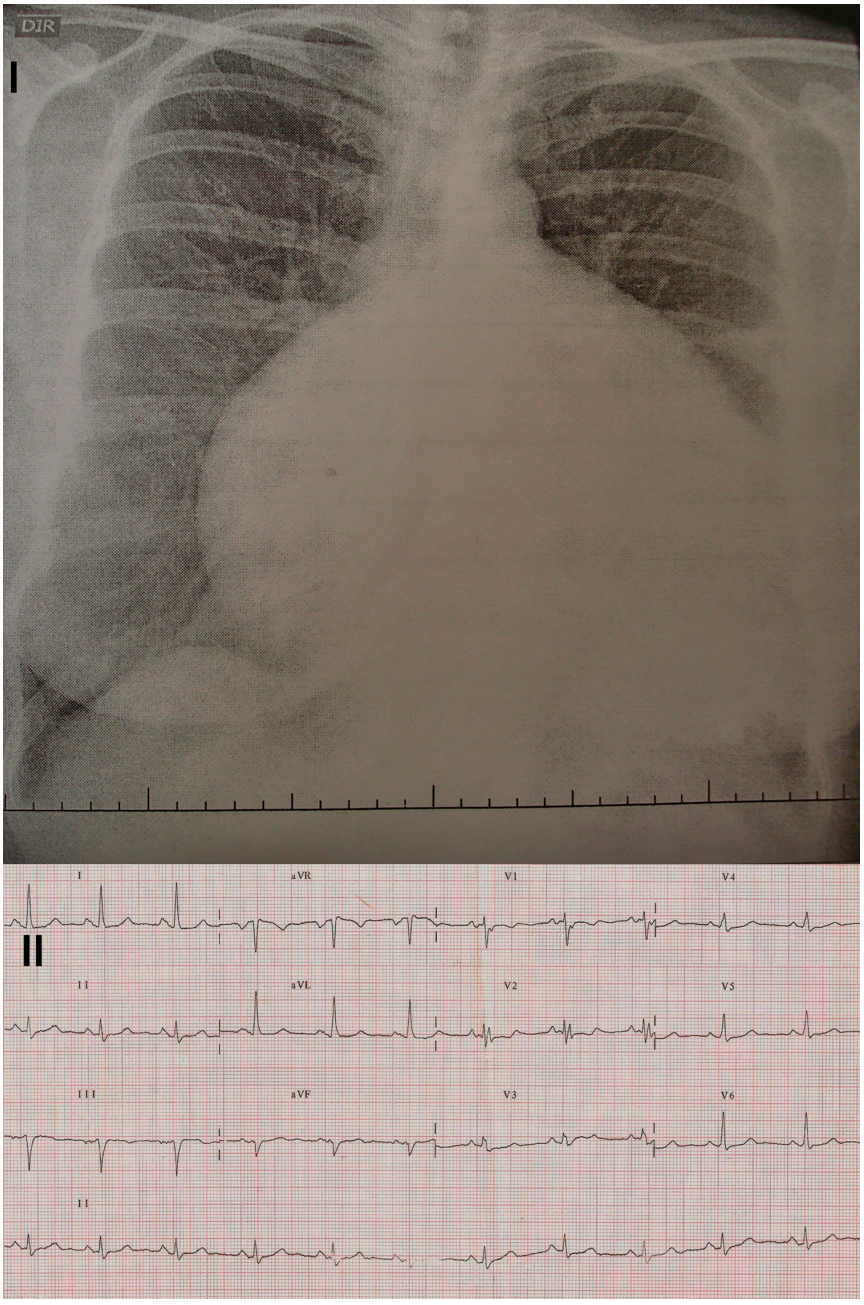

Figure 1 – *I: Chest X-ray evidencing important cardiac area enlargement. II: ECG before ablation, with no evidence of pre-excitation.*

The presence of untreated accessory pathway in patients with EA can lead to a reserved prognosis even in individuals undergoing surgical repair[7]. Huang et al[7] have reported that 20% of the patients with history of supraventricular tachycardia progressed to sudden death despite the surgical repair for EA. No death due to that cause in the group undergoing preoperative EPS was observed.

A more aggressive attitude in patients with EA is justified by the high prevalence of arrhythmias (up to 80%)[7], via either the atrioventricular anomalous pathway, or the potential arrhythmia circuits generated in the postoperative period [atrial fibrillation (AF) and incisional flutters]. The sudden death mechanism in EA is usually attributed to AF and high ventricular response via an accessory pathway[8]. Thus, the identification and previous ablation of the accessory pathway would be highly desirable.

Another justification for the preoperative search for accessory pathways in EA would be the possibility to eliminate that pathway during surgery[4,5]. Catheter ablation

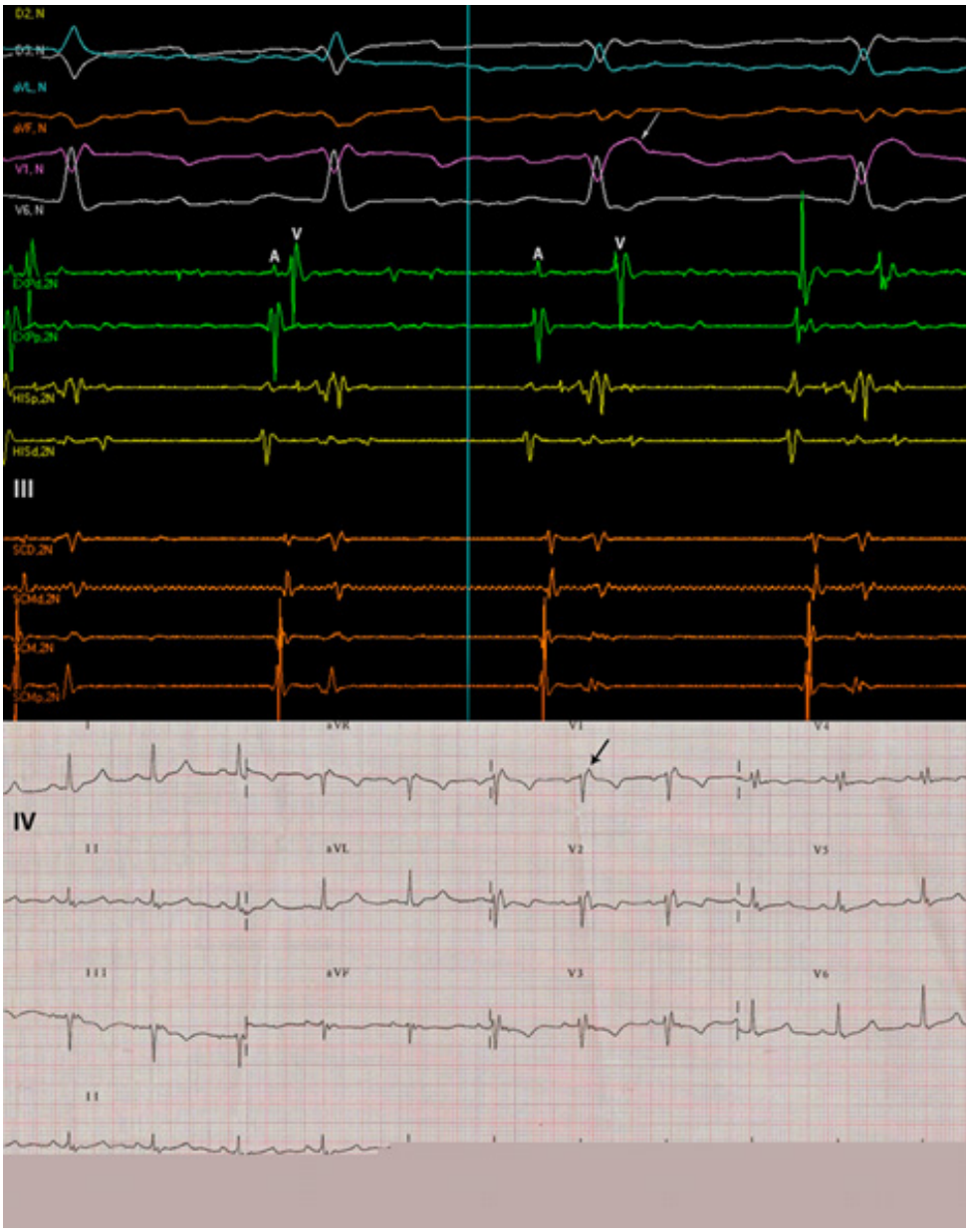

Figure 2 – *III: Continuous electrograms during ablation, showing elimination of the anomalous pathway during radiofrequency application (1st intracavitary electrogram). A: atrial electrogram. V: ventricular electrogram. Note atrioventricular fusion that fades after ablation (arrow). IV: ECG after ablation. Note QRS enlargement and progression of the conduction disorder to right bundle-branch block (arrow).*

has significantly lower success rates (around 80%)[2] than those of the general population (around 95%). That results from the mapping difficulty due to the low tricuspid valve implantation[2]. The preoperative non- identification can imply missing a unique opportunity for surgically sectioning an anomalous pathway with previous inefficient ablation.

A third justification for preoperative EPS would be that the surgery currently considered the most anatomical (cone surgery)[9] can exclude potential areas of anomalous pathways with plication of the atrialized portion of the right ventricle. This would make catheter access to that heart region impossible, a usual ablation site for accessory pathways in those patients (septal and posteroseptal regions). In addition, patients with EA have abnormal ventricular irritability during catheter manipulation in the intra- and postoperative periods, making them more susceptible to arrhythmias with sudden death risk[7].

Despite the insufficient number of studies to support a recommendation for preoperative EPS in all patients

scheduled for surgical repair of EA, considering the relatively low risk of EPS and the huge potential benefit if an anomalous pathway is found, that more aggressive attitude should be considered in the management of asymptomatic patients[10].

Conclusion

This case report suggests that preoperative EPS can be useful to patients with EA and no evidence of anomalous conduction pathways. In asymptomatic patients with normal ECG, an additional hint about masked accessory pathway (observed in our patient) is the lack of RBBB. However, further studies about that population of patients are required to assess the real dimension of the benefit of that procedure.

Author contributions

Conception and design of the research: Oliveira L, Freitas AKE, Mehta N, Ortiz MR, Mulinari LA, Cunha CLP; Acquisition of data: Oliveira L, Freitas AKE, Mehta N, Mulinari LA; Analysis and interpretation of the data and Writing of the manuscript: Oliveira L, Freitas AKE, Mehta N; Critical revision of the manuscript for intellectual content: Mehta N, Ortiz MR, Mulinari LA, Cunha CLP.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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Study Association

This study is not associated with any thesis or dissertation work.

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