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Original article

Cardiac arrest and ventricular arrhythmia in adults with Ebstein anomaly and left ventricular non-compaction

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ABSTRACT

Background: Ebstein anomaly is a complex, congenital heart defect that is associated with a variety of cardiac abnormalities. Studies found a similar sarcomere gene mutation in patients with Ebstein anomaly (EA) and patients with isolated left ventricular non-compaction (LVNC).
Aim: We aimed to show the prevalence of LVNC and its potential relationship with severe cardiac events (VT – ventricular tachycardia, cardiac arrest) in adult patients with EA.
Methods: We conducted a retrospective search of our institutional database from 2010 to 2014 for patients with EA and reviewed patients' medical records (age, sex, clinical presentation, electrocardiographic, echocardiographic, and CMR – cardiac magnetic resonance features). We reviewed echocardiograms and CMR scans for concomitant morphological abnormalities (LVNC, PDA – patent ductus arteriosus, VSD – ventricular septal defect, ASD – atrial septal defect, mitral valve prolapse, BAV – bicuspid aortic valve, CoA – coarctation of aorta).
Results: The studied group consisted of 84 consecutive patients (mean age 38 ± 15 years, 50 women) with EA.

We found four patients (4.8%) with LVNC, two of them had cardiac arrest, one had VT, and one was symptomless, but had QTc prolongation in Holter recordings. Concomitant abnormalities were VSD (4.8%), PDA (1.2%), CoA (1.2%), mitral valve prolapse (1.2%), and BAV (2.4%). The most common anomaly was ASD type II – 23 patients (27.3%) and WPW – Wolff-Parkinson-White's syndrome – 9 patients (10.7%).
Conclusions: Non-compaction is a notable abnormality in adult patients with EA and it may affect their prognosis. Although other concomitant lesions were more common, only patients with LVNC suffered from cardiac arrest or ventricular arrhythmia.
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Introduction

Left ventricular non-compaction (LVNC), a rare cardiomyopathy with a poor prognosis, accompanies congenital heart disease in up to 12% of cases [1]. Ebstein anomaly (EA) is linked with LVNC genetically [2–4]. Non-compaction of the ventricular myocardium is characterized by numerous excessively prominent trabeculations and deep intertrabecular recesses [5]. Patients with LVNC and arrhythmias seem to have worse outcomes compared to those with similar forms of rhythm disturbance but without LVNC. The presence of ventricular arrhythmias is an independent risk factor for mortality [2,5–7].

Objectives

We aimed to show the prevalence of LVNC and its potential relationship with severe cardiac events (VT – ventricular tachycardia, cardiac arrest) in adult patients with EA.

Patients and methods

We conducted a retrospective search of our institutional database from 2010 to 2014 for patients with EA and reviewed

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patients' medical records (age, sex, clinical presentation, electrocardiographic, and echocardiographic data). Standard two-dimensional echocardiography with Doppler and color Doppler, resting electrocardiographic (ECG) and Holter monitoring are performed at least annually in every patient with EA. Two independent echocardiographers, blinded to each other's results and the clinical data, screened echocardiographic studies and focused on non-compaction.

All patients enrolled in the study were diagnosed with EA in childhood and, as adults, met the echocardiographic criteria of apical displacement of the septal leaflet of the tricuspid valve. This was observed following the insertion of the anterior leaflet of the mitral valve by at least 8 mm/m^2 of body surface area [8].

A universally accepted definition of non-compaction is lacking. In our study, we employed the echocardiographic criteria most widely used and first published by Jenni et al. [9]. These criteria defined LVNC as a two-layer structure, with a thin, normally compacted layer (C) and a markedly thickened non-compacted layer (NC) (with a ratio of NC/C > 2). Excessively prominent trabeculations, and deep intertrabecular recesses were measured at end-systole in parasternal short-axis views.

In revised echocardiograms, we searched for accompanying concomitant morphological abnormalities such as LVNC, persistent ductus arteriosus (PDA), ventricular septal defect (VSD), atrial septal defect (ASD), mitral valve prolapse (MVP), bicuspid aortic valve (BAV) and coarctation of the aorta (CoA). In ECGs, ECG Holter monitoring and clinical data we looked for: the prevalence of supraventricular (supraventricular tachycardia-SVT, atrial fibrillation or flutter - AF) and ventricular arrhythmia, bradyarrhythmia, and other rhythm disorders such as ventricular pre-excitation syndrome (WPW) or pacemaker implantation.

Statistics

Descriptive statistics include frequencies and percentages for categorical data and mean ± SD values for continuous data.

Results

The studied group consisted of 84 consecutive adult patients (59% women, mean age 38 ± 15 years, median 34.5, range 16–71 years) with EA. Nineteen of them (22.6%) had tricuspid valve replacement, 3 (3.6%) tricuspid valve plasty, 3 (3.6%) a Glenn operation, and in 3 (3.6%) cases percutaneous ASD closures with an Amplatzer septal occluder were performed. Based on medical history, and the results of ECGs and Holter monitoring, we established the prevalence of concomitant rhythm disorders and the number of implanted devices (pacemakers/ICD implantable cardiac defibrillator) (Table 1).

Two echocardiographers (MK, ML) independently found only 4 patients (4.8%) with LVNC. One diagnosis of LVNC was confirmed by magnetic resonance imaging. The echocardiographic characteristic is listed in Table 2.

Although concomitant lesions other than LVNC were more common, only patients with this abnormality suffered from cardiac arrest or ventricular arrhythmia. Among two cardiac arrest survivors, one had a decreased left ventricular ejection fraction (40%) and a history of WPW ablation, the other had an ICD implanted in the secondary prevention of cardiac arrest. The third patient with LVNC had non-sustained VT in Holter recordings. The fourth was symptomless during two years of observation, but with a QTc prolongation observed in Holter recordings. In a patient with QTc prolongation we observed it in Holter recording, QT was 425 ms, QTc was 506 ms, and heart rate at this point was 85/min. Table 3. In 3 patients with LVNC, 5 segments were affected: apex, apical lateral, mid inferolateral, apical inferior, and mid inferior. In one patient, we saw 6 segments with noncompacted layers: apex, apical septal, apical anterior, apical lateral, mid anterior, and mid lateral. All segments met Jenni's criteria: a maximal end systolic ratio of non-compacted to compacted layers of >2 (Fig. 1).

Table 1
Baseline characteristics of studied population (n = 84) patients. The prevalence of accompanying morphological abnormalities and concomitant rhythm disorders and device implantation.

Age	Mean 38 ± 15 yrs Number	Median 34.5 (16–71 yrs) % of overall EA patients
Female	50	59
Caucasian	84	100%
Surgery procedures		
TVP	3	3.6
TVRb	15	17.9
TVRm	4	4.8
AMPL	3	3.6
Glenn op.	3	3.6
Associated cardiac lesions	**Number**	**% of overall EA patients**
ASD	23	27.3
VSD	4	4.8
PDA	1	1.2
MVP	1	1.2
BAV	2	2.4
CoA	1	1.2
LVNC	4	4.8

2 patients have more than one associated lesion.
TVP, tricuspid valve plasty; TVRb or TVRm, tricuspid valve replacement with biological or mechanical valve; Glenn op., Glenn's procedure; AMPL, percutaneous ASD closures with Amplatzer septal ocluder; ASD, atrial septal defect; VSD, ventricular septal defect; PDA, persistent ductus arteriosus; MVP, mitral valve prolapsed; BAV, bicuspid aortic valve; CoA, coarctation of the aorta; LVNC, left ventricular non-compaction; EA, Ebstein's anomaly.

Table 2
Echocardiographic parameters.

Echcardiographic parameters	n = 84
LVEDd (mm)	41.5 ± 7.3
LVEF (%)	57.8 ± 11.6
RVIT (mm)	50.5 ± 9
RAA	40.8 ± 15
TR (grade 1/2/3/4)	28/11/26/19

LVEDd, left ventricle end-diastolic diameter; LVEF, left ventricular ejection fraction; RVIT, right ventricular inflow tract; RAA, right atrium area; TR, tricuspid regurgitation.

Discussion

Chronic symptoms in EA are mainly related to right heart morphology and function. In our study, we focused not only on right-sided abnormalities but also on left-side pathology. We recognized LVNC as a notable abnormality in patients with EA (4.8%) – the second most concomitant lesion in the study (Table 1). Surprisingly, in our study the prevalence of LVNC in EA patients was even lower than had been previously reported (from 4.8% to 16–18%) [10,11]. A probable explanation for this is that our patients were older than those in previous studies. In the cited series, the age range was broad – from birth and it covered the pediatric or adolescent population [10,11]. Other features such as the percent of operated patients, and the percentage of implanted pacemakers were similar to previous reports [10,12]. Some studies on EA follow-up do not mention LVNC [12]. It is likely that patients with serious forms of LVNC associated with EA died earlier [11]. LVNC was reported to be less common in the adult EA population than in the pediatric one [12–14]. The presence of LVNC had not always

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