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CASE REPORT
Rare congenital heart diseases

Pregnant woman with Ebstein's anomaly

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Abstract

A 34-year-old female with Ebstein's anomaly presented to our centre at 19th week of her first pregnancy with a history of dyspnea, palpitations and syncopes. Echocardiogram showed typical signs of Ebstein's anomaly with severe tricuspid regurgitation, large atrial septal defect with left-to-right shunt and elevated right ventricular systolic pressure. She was prescribed metoprolol and anticoagulation with low-molecular-weight heparin. The pregnancy course was uneventful. She delivered by vaginal route in 36th week of pregnancy a healthy male baby weighing 2140 g. On the 6th day after delivery she experienced tachycardia, dyspnea, dizziness. Clinical examination and additional studies revealed only slight limb oedema and low haemoglobin level. After treatment with furosemide and prescription of folic acid and iron supplementation she was discharged home in a good clinical condition after 4 days. JRCD 2012; 1: 13–17

Key words: Tricuspid valve regurgitation; Atrial septal defect; Pulmonary hypertension

Case presentation

A 34-year-old female with Ebstein's anomaly diagnosed at early childhood presented to our centre at 19th week of her first pregnancy. **Her main symptoms** included syncopes, with last episode at 6th week of pregnancy, dyspnea and occasionally palpitations. She was at New York Heart Association (NYHA) functional class II during first examination.

Physical examination revealed systemic blood pressure of 110/80 mm Hg, heart rate (HR) of 78 beats per minute (bpm), and body mass index (BMI) of 24.9 kg/m^2. Her oxygen saturation (SpO_2) in room air was decreased to 90%, however no signs of cyanosis were seen. On auscultation a holosystolic murmur along the low left sternal border was heard.

Blood reports were within normal limits.

ECG which is presented in Figure 1 demonstrated:

- sinus rythm at 71 bpm with first degree atrio-ventricular (a-v) block (PQ = 0.22s),
- right axis deviation,
- prolonged QTc to 458 ms,
- mitral P
- signs of right ventricle (RV) hypertrophy (S in V_6 >3 mm)
- non-specific interventricular conduction disturbances.

In 24-hour ambulatory (Holter) ECG monitoring maximal HR was 142 bpm, minimal 57 bpm, and mean 74 bpm. There were 124 isolated supraventricular premature beats, 4 episodes of supraventricular tachycardia (SVT) (longest run at rate of 124 bpm), and 8 ventricular premature beats.

Transthoracic echocardiogram (TTE) showed typical signs of Ebstein's anomaly such as: caudal (inferior) displacement of the septal and posterior leaflets of the tricuspid valve (>8mm/m^2) with severe tricuspid regurgitation (TR) and right atrial (RA) and RV enlargement (areas of 89.1 and 74.6 cm^2, respectively) with small left ventricle (area = 17 cm^2). Additionally, large atrial septal defect (ASD II) with left-to-right shunt and estimated right ventricular systolic pressure (RVSP) of 46 mmHg were detected (Figure 2, 3, 4).

At 6-minute walk test the patient walked 420 m with increase of heart rate from 77 bpm before exercise to 109 bpm at the end of the test and increase of SpO_2 from 93% to 96% after exercise.

Condition of the fetus, assessed by gynecological examination with fetal ultrasonography, was considered normal.

Literature review

Ebstein's anomaly (EA) is a rare congenital heart defect with a prevalence of 0.3–0.5% (without any gender predilection) characterized primarily by abnormalities of the tricuspid valve and right ventricle [1]. The survival of patients with EA during childhood and adolescence has improved dramatically: after 35 months of diag-

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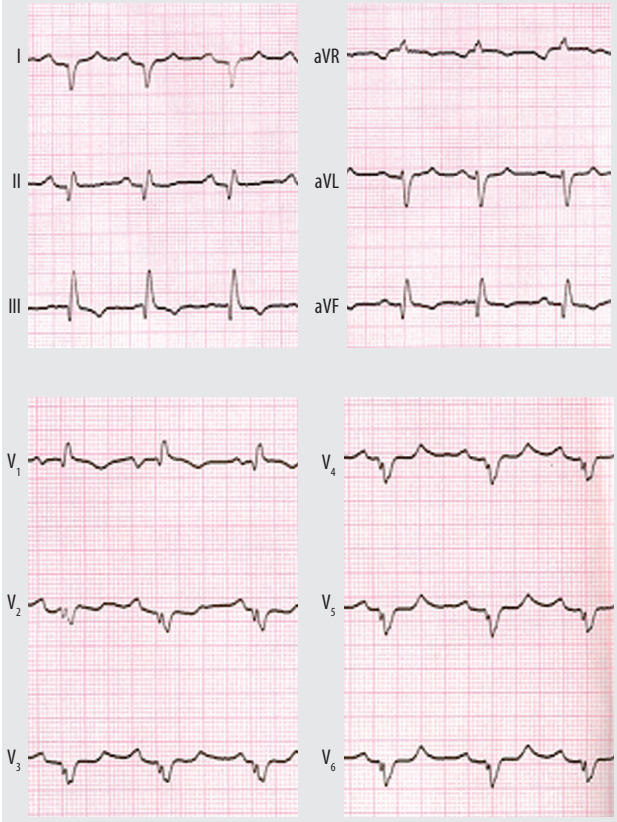

Figure 1. ECG: Right axis deviation, first degree a-v block (PQ = 0.22 s), prolonged QTc = 458 ms, mitral P, RV hypertrophy and non-specific interventricular conduction disturbances

nosis, the Kaplan–Meier survival probability remains stable at 80% (95% CI: 0.72–0.89) [2]. Survival into adulthood is common, there were also described survivals up to 85 years [3]. EA may manifest clinically at any age and has a highly variable clinical course. Adults often present with cyanosis, dyspnea, palpitations, decreasing exercise tolerance, fatigue [4-9]. In the presence of an interatrial communication the risk of paradoxical embolization, brain abscess and sudden cardiac death increases. Exercise tolerance is dependent on heart size and oxygen saturation [8-11].

EA is a malformation characterized by adherence of the septal and posterior leaflets of tricuspid valve to the underlying myocardium, apical displacement of the functional annulus >8 mm/m^2 body surface area, dilation of the atrialized portion of the RV, redundancy fenestrations and tethering of the anterior leaflet and dilation of the right atrio-ventricular junction [9,12]. The spectrum of the malformation may range from minimal displacement of the septal and posterior leaflets to imperforate membrane between the inlet and trabecular zones of the RV [9].

According to the Carpentier anatomic classification of EA [13] the patient was classified as B/C, according to the Celermajer echocardiographic grading score [8], the patient was encountered to grade 3 (ratio of 1.2). The ratio provides information about prognosis, when the ratio is 1.5–100% mortality is likely, contrarily a ratio of 0.1 indicates 92% survival. There are no publications regarding its use in pregnancy [5,14].

An interatrial communication (ASD, Patent Foramen Ovale) is present in 80–94% of patients with EA [15,16]. This patient had ASD II with left-to-right shunt. Accessory pathways (Wolff–Parkinson–White syndrome) are commonly associated with EA (6–36%) and may lead to supraventricular tachycardia [5,9]. First degree heart block is found in up to 50% of patients (may relate to right atrial dilation and stretch), right bundle branch block (RBBB) is a typical finding [3,17,18]. Our patient didn't have any signs of pre-excitation, ECG revealed first degree a-v block.

Other cardiac defects associated with EA include ventricular septal defect, pulmonary outflow obstruction, patent ductus arteriosus, coarctation of aorta, parachute mitral valve, cleft anterior leaflet of the mitral valve, mitral valve prolapse, bicuspid aortic valve, corrected transposition of the great arteries, subaortic stenosis, tetralogy of Fallot, and left ventricular non-compaction [9,19,20].

This anomaly does not have any effect on fertility, even in women with cyanosis [4]. According to current guidelines women with EA without cyanosis and heart failure are encountered to World Health Organization (WHO) risk class II and usually tolerate pregnancy well [21].

In opposite, symptomatic patients with cyanosis and/or heart failure should be treated before pregnancy or counselled against pregnancy. The haemodynamic problems seen during pregnancy depend on the severity of the TR and the functional capacity of the RV.

Heart failure, stroke, arrhythmias, paradoxical embolism can occur even in the asymptomatic patients. The presence of arrhythmia or cyanosis in the mother is associated with increased maternal and fetal risk, and needs closer maternal and fetal monitoring during pregnancy and delivery [22].

While pregnant patients with EA are usually acyanotic, those with interatrial shunting can develop shunt reversal and cyanosis in pregnancy. The parameters of bad prognosis include SpO_2 <85% and Hb >18 g/dl; these are related to increased fetal loss rate. Mild cyanosis is associated with increased premature deliveries, low birth weight and thromboembolic complications [23].

The largest series reporting the outcome of 111 pregnancies in 44 women with EA was described by Connolly and Warnes [4]. In this report 16 patients were cyanotic, 20 had an interatrial communication (ASD/PFO). Majority (76%) of pregnancies resulted in live birth, 89% were delivered vaginally, 11% by cesarean section. The mean birth weight of infants born to cyanotic women was significantly lower than of newborns of acyanotic women (2530 vs. 3140 g, $p < 0.001$) [4].

Donelly *et al.* [21] described 42 pregnancies in 12 patients with EA. Most of them were well tolerated, resulting in 36 live births. Mild dyspnea in 3rd trimester of pregnancy was common, 1 patient had paroxysmal atrial tachycardia (during 1st pregnancy) and atrial fibrillation (2nd pregnancy), and 1 patient – right heart failure.

Chopra et al. analyzed the outcome of 8 pregnancies in 4 patients with EA, one of them had right heart failure, two had arrhythmia [5].

In patients with EA the physiological changes of pregnancy may aggravate haemodynamic consequences. In the pregnant women with impaired RV size and function, the increased stroke volume

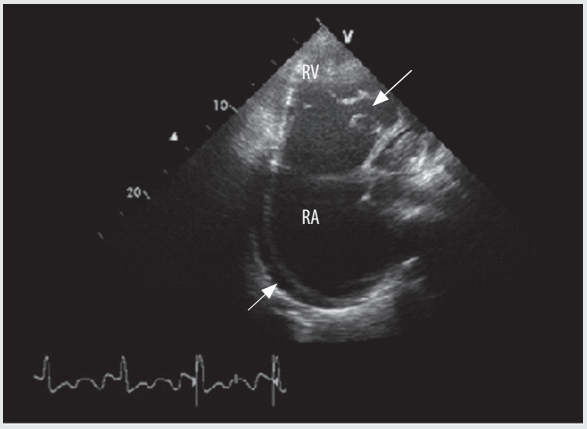

Figure 2. Echocardiography. Four chamber view. Apical displacement of septal and posterior leaflets of the tricuspid valve (25.5 mm, 8.8 mm/m^2) (big arrow). Small pericardial effusion (small arrow). RA – right atrium, RV – right ventricle

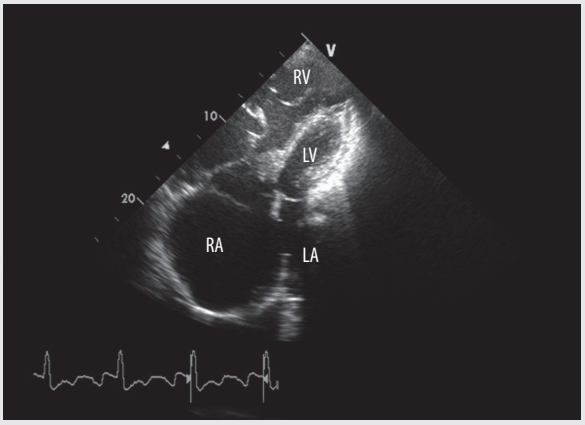

Figure 3. Echocardiography. Four chamber view. RA and RV enlargement (areas of 89.09 and 74.6 cm^2, respectively). Compression of the LV (area = 17 cm^2). RA – right atrium, RV – right ventricle, LA – left atrium, LV – left ventricle

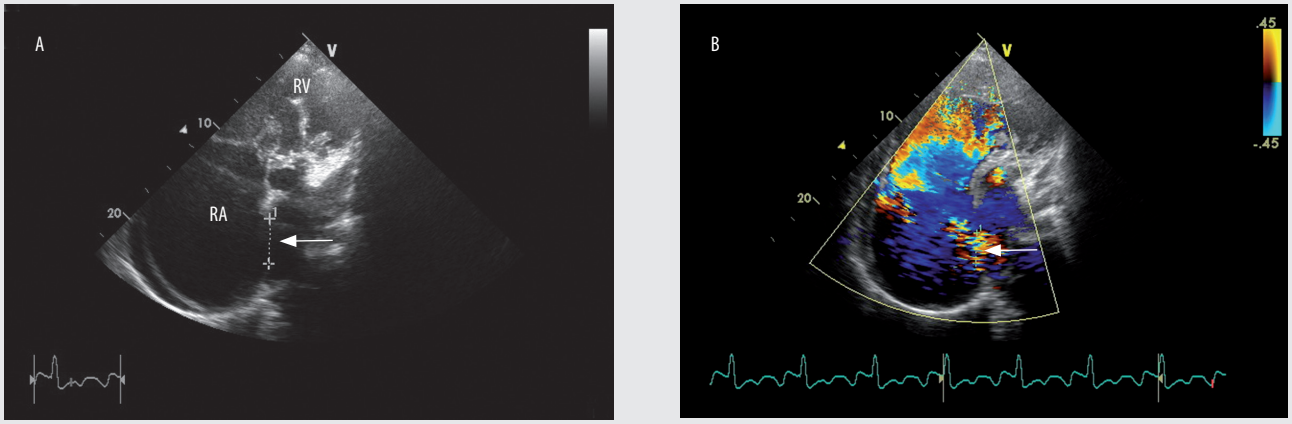

Figure 4. Echocardiography. Four chamber view. Large atrial septal defect (ASD II) with left-to-right shunt (arrow). RA – right atrium, RV – right ventricle. A. 2D imaging. B. Colour Doppler imaging

(SV) may be poorly tolerated and results in worsening tricuspid regurgitation, raised atrial pressures and increased right-to-left shunting [24]. No correlation was demonstrated between symptoms and the degree of displacement of the tricuspid valve leaflets [22,25]. Even severe TR with heart failure can usually be managed medically during pregnancy. Medical treatment includes therapy for heart failure and arrhythmia. In this patient, beta-blocker therapy with metoprolol 50 mg/day (due to prolonged QTc interval and syncopal symptoms) and anticoagulant therapy with low-molecular-weight-heparin (LMWH) were used. Palpitations and supraventricular arrhythmias documented in Holter monitoring in the patient with two clinically relevant non-major risk factors (heart failure, female, CHA_2DS_2-VASc = 2) constituted the indication for anticoagulation. Moreover, the patient presented pulmonary hypertension secondary to the large atrial septal defect with left-to-right shunt. Additional risk factors are the procoagulant state related to pregnancy, sedentary lifestyle and weight increase. All of these factors may exaggerate risk for thromboembolic events, i.e. paradoxical embolism.

There were reported an increased risk of fetal loss, prematurity and low birth weight in the babies born to mothers with cyanosis [4,22].

The preferred mode of delivery is vaginal in almost all cases [21]. The management during labor should avoid all factors leading to congestive heart failure, cyanosis and arrhythmias [5,26]. To maintain normal sinus rhythm during labour, adequate pain relief in the form of epidural analgesia is helpful and can be upgraded to anesthesia if cesarean section is indicated [27-29]. During the second stage of labour, Valsalva maneuver causes an increase in intrathoracic pressure, increase in right-to-left shunt, therefore assisted vaginal delivery is indicated [5,30].

The risk of congenital heart disease in offspring is reported in 4–6%, and familial EA in 0.6% [4,31]. There were no congenital

abnormalities recognized in baby of our patient. Endocarditis prophylaxis in the peripartum period is not indicated for the pregnants with EA [32].

Patient management and follow up

The patient was prescribed long acting metoprolol 50 mg/day and anticoagulant therapy with LMWH (Enoxaparine 2×60 mg sc). She was closely monitored with clinical examination and echocardiography every month until delivery. There were no changes noted on RV size and function on repeated echocardiograms. Serial NT-proBNP were as follows: 203.9 pg/ml, 123.1 pg/ml, 148.7 pg/ml (normal range <125 pg/ml). The course of pregnancy was uneventful.

Patient delivered vaginally in the 36th week of pregnancy a healthy male baby, weighing 2140 g and the Apgar score of 10. On the 6th day after delivery she experienced tachycardia, dyspnea, dizziness. She was referred to cardiac department. On admission her heart rate was 80 bpm, blood pressure 90/60 mm Hg. On physical examination a slight lower limbs oedema was observed, which resolved after furosemide 20 mg iv. ECG, echocardiography were performed – no significant changes were recognized, there were no signs of right heart failure, no change in hemodynamic factors. SpO_2 was stable 94%. Laboratory tests revealed anaemia (Hb = 9.9 g/dl, Ht = 30.2%, RBC = 3.25×10^6/mm^3). Other parameters: D-dimers 0.75 mg/l, Na^+ = 138 mmol/l, K^+ = 4.8 mmol/l, CRP = 3.0 mg/l, total protein = 54 g/l, albumins = 30 g/l, fibrin degradation products <5.0 ug/ml (were within normal ranges). NT-proBNP was elevated (825.8 pg/ml, 451 pg/ml). She was treated with long acting metoprolol, furosemide, LMWH, folic acid, iron supplementation. The patient was consulted with cardiac surgeon and she was accepted for surgical correction of EA nevertheless she decided to postpone the operation about 1 year. After 4 days she was discharged home.

Algorythm

Before pregnancy:

Symptoms: cyanosis, right heart failure, poor exercise tolerance NYHA III-IV – pregnancy contraindicated

Echocardiogram (Celermajer classification): impaired RV size and function, right-to-left shunting, pulmonary hypertension, Celermajer grade 4 – pregnancy contraindicated

ECG, Holter monitoring – any severe arrhythmias should be treated before pregnancy

oxygen saturation <85% – pregnancy contraindicated

During pregnancy:

Patients with EA without cyanosis, heart failure, pulmonary hypertension and arrhythmia (WHO class II): clinical and echocardiographic monitoring of patient – every trimester of pregnancy and after delivery

Patients with EA and pulmonary hypertension and supraventricular arrhythmia: close clinical and echocardiographic monitoring of patient – every 2–4 weeks during 1st and 2nd trimester of pregnancy, after 28th week – once a week, in tertiary centre having experience with patients with congenital heart diseases. Holter monitoring, serial oxygen saturation, BNP (NT-proBNP) are recommended. Close obstetric and fetal ultrasonography monitoring.

Medical treatment: heart failure and arrhythmia therapy based on clinical status

Delivery: assisted vaginal delivery, epidural analgesia. Cesarean section in case of maternal or fetal complications.

Conflict of interest: non declared.

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