Case reports

Ebstein's anomaly and pregnancy: a case report

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Ebstein's anomaly, an uncommon malformation of the tricuspid valve, has an extremely variable natural history, depending on a wide spectrum of pathological features. We here described a case of a patient with Ebstein's anomaly who gave birth to 2 healthy unaffected full-term infants after two successful pregnancies; a third pregnancy miscarried at the 11th week. The anomaly was diagnosed during childhood, was not associated with other cardiac anomalies, cyanosis or preexcitation and the echocardiographic degree of severity was low (grade 1). All of these factors are considered predictors of a good survival and prognosis. During the pregnancies, no arrhythmias, cyanosis or signs of cardiac failure were observed and the patient's NYHA functional class (I) remained unchanged. Our case is the only published case of two successful term pregnancies in Ebstein's anomaly, it confirms the importance of echocardiographic evaluation and that the probability of maternal and neonatal events may be predicted from the baseline characteristics of the mother. Pregnancy is well tolerated in the absence of important maternal cardiomegaly, cyanosis and arrhythmias and in those patients with mild cardiac dysfunction as evaluated at echocardiography and a low NYHA class, but it is associated with an increased risk of abortions and prematurity.

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Ebstein's anomaly is characterized by an apical displacement of the septal leaflet of the tricuspid valve. The clinical manifestations of the disease vary markedly depending on other pathological features and the presence of other associated anomalies. If the deformity of the tricuspid valve is severe, congestive heart failure with hydrops fetalis, intrauterine death or congestive heart failure at birth may occur[1]. Fetal and neonatal presentation is associated with a poor outcome[2]. The patients surviving till adolescence may present with different degrees of the anomaly depending on the right ventricular function. Little information is available about the course and outcome of pregnancy in women with Ebstein's anomaly. The aim of our report was to evaluate and confirm the prognostic value of the baseline maternal characteristics on the course and outcome of two pregnancies.

Case report

The patient, A.S., 36 years old, born following an uneventful pregnancy and with a normal growth pattern had no history of cyanosis or of intolerance to physical effort. The family history was negative for mental disorders or therapy with lithium carbonate to the patient's mother[3]. In 1975, when the patient was 9 years of age, clinical examination revealed the presence of a cardiac murmur and an ECG showed right bundle branch block. On the basis of this finding a cardiac anomaly was suspected. Cardiac catheterization showed only a minimal pressure gradient between the right ventricle and pulmonary artery and the relative cardiac volume was 356 ml/m^2 (normal value 324 ± 28 ml/m^2). The right ventricular outflow tract was slightly enlarged and a mild pulmonary stenosis was diagnosed. During hospitalization, a chest X-ray showed a regular transparency of the lungs with a regular vasculature and no cardiomegaly. Blood analysis was within normal limits. Afterwards the patient enjoyed good health and grew normally without symptoms. In 1987, at 21 years of age, a two-dimensional echocardiogram showed the presence of Ebstein's anomaly and revealed apical displacement of the insertion of the septal leaflet of the tricuspid valve with severe regurgitation, enlargement of the right chambers and a minimal mitral valve prolapse. Chest X-ray had remained unchanged with

respect to the previous one (September 1, 1975). Drugs were not prescribed to the patient but periodical echocardiograms and visits were advised. At the age of 27 years, the patient became pregnant for the first time. In February 1983, echocardiography showed a shorter septal leaflet of the tricuspid valve as well as an apical displacement of 2.5 cm (15.9 mm/m^2) below the valvar annulus and a deformed, but normally positioned posterior leaflet (Fig. 1). The right ventricle was moderately dilated without hypertrophy, the right atrium was dilated and a severe tricuspid regurgitation was found with a highest jet velocity of 2 m/s; the inferior caval vein had a normal size with respiratory collapse. Blood analysis was within normal limits and the hemoglobin level was 11.5 mg/dl. During pregnancy the patient never presented with symptoms of heart failure and a Holter ECG carried out during the third month confirmed the absence of arrhythmias. A caesarean section was performed at 38 weeks of gestation (W.G.); the child was in good health weighing 3350 kg and with an Apgar index of 9. On July 24, 1996 the patient underwent a clinical examination that revealed normal cardiac sounds and holosystolic tricuspid regurgitation. The liver was not enlarged. The echocardiographic picture had remained unmodified with respect to 1993 (Table I). In July 1996, after a surgical examination, it was decided that surgical intervention was not necessary and the patient was prescribed drugs for the prophylaxis of endocarditis. On November 18, 1996, during a periodical check-up, the patient was prescribed digoxin for the prophylaxis of tachyarrhythmias in case she became pregnant again. In May 1997, the patient was again pregnant and in the second month an echocardiogram was performed with the same results as previously. Blood analysis was within normal limits and the hemoglobin level was 11.8 mg/dl; it was decided to continue with digoxin (0.125 mg/daily). Holter ECG, performed during the fourth month of gestation, did not reveal any arrhythmias. On December 16, 1997, at the seventh month of pregnancy, the patient without symptoms was again submitted to cardiologic evaluation, with a satisfactory clinical outcome and an unchanged echocardiogram (Table I). Her hemoglobin level had remained unmodified at 11.8 mg/dl. Fetal

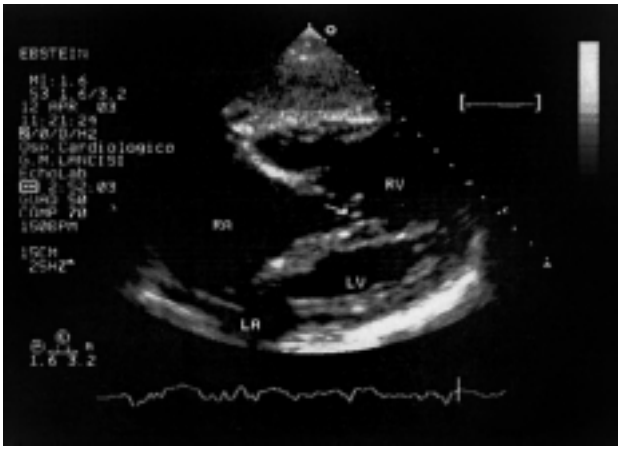

Figure 1. *Four-chamber subcostal view. LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle. The* arrow *shows the apical displacement of the septal tricuspid leaflet.*

Table I. Echocardiographic data.

	February 1993	July 1996	December 1997	February 2000	January 2002	April 2003
Age (years)	27	30	31	34	36	37
No. pregnancies	1st	2nd	2nd	0	3rd	0
Months of pregnancy	7th	2nd	7th	–	2nd	–
Echocardiographic parameters						
LV end-diastole (cm)	4.0	4.0	3.9	3.9	4.0	3.9
LV end-systole (cm)	1.7	1.9	1.9	2.1	1.7	1.5
Fractional shortening (%)		38	50	46.2	57	61.5
IVS end-diastole (cm)	1.0	0.9	0.9	0.9	0.9	0.9
IVS end-systole (cm)	–	–	–	1.4	1.4	1.5
PW end-diastole (cm)	1.0	0.9	0.9	0.7	0.7	0.7
PW end-systole (cm)	–	–	–	1.4	1.5	2.0
LVM (g)	–	–	–	90	93.1	90
LVMI (g/m^2)	–	–	–	58.2	59.2	55.4
LA end-systole (cm)	2.8	3.3	3.3	2.1	3.0	3.4
AO-R end-diastole (cm)	2.6	2.4	2.4	2.6	2.8	2.7
RV end-diastole (cm)	–	4.0	4.1	3.9	3.9	3.9
IVC	Normal	Normal	Normal	Normal	Normal	Normal
IVC-RC	Preserved	Preserved	Preserved	Preserved	Preserved	Preserved
TVR	Massive	Massive	Massive	Massive	Massive	Massive
Vmax v–a/dx (m/s)	2	2	2	2	2	2

AO-R = aortic root; IVC = inferior vena cava; LA = left atrium; LV = left ventricle; LVM = left ventricular mass; LVM = left ventricular mass index; PW = posterior wall; RC = respiratory collapse; RV = right ventricle; TVR = tricuspid valve regurgitation; Vmax v-a/dx = maximal velocity of the right ventriculo-atrial jet.

echocardiography was normal, with a normal tricuspid valve. In February 1998, a caesarean section was performed at 38 weeks of gestation; a child weighing 3200 kg and with an Apgar index of 9 was delivered. At clinical evaluation performed on February 22, 2000 the patient's cardiac status was found to be good, without cyanosis or arrhythmias. In December 2001 the patient had her third pregnancy. The echocardiographic picture had remained unmodified and the hemoglobin level was normal (14.2 mg/dl). On February 18, 2002, at 11 weeks of gestation, the pregnancy ended in a spontaneous abortion.

Discussion

The maternal and fetal outcome of pregnancy in Ebstein's anomaly has been previously described in several series[4-8]. The reported number of successful pregnancies is small and there is little information about such pregnancies[4,5,8]. Analyzing these data it appears that a pregnancy may be well tolerated even though it is believed that this anomaly is associated with an increased risk of miscarriages, early neonatal deaths, premature births and of congenital heart disease in the offspring of such patients (6-14%)[5]. Pregnancy may be well tolerated after tricuspid valve repair, replacement or following the correction of associated anomalies even though the increased stroke volume and cardiac output occurring during pregnancy may cause an enlargement of the right ventricle with annulus dysfunction and worsening tricuspid regurgitation[5]. A significantly lower weight is found in the newborns of cyanotic vs acyanotic women with Ebstein's anomaly, both in full-term and in pre-term infants[6]. Arrhythmias and other factors as well as the increasing heart rates, catecholamines, the decreasing peripheral vascular resistances and enhanced right ventricular overload may lead to heart failure in pregnancy. On the whole, pregnancy in women with congenital heart disease is not associated with an increased mortality rate but female patients with Ebstein's anomaly who reach child bearing age have a considerable morbidity due to congestive heart failure, thromboembolic complications and rhythm disorders[7]. The probability of maternal or neonatal cardiac events may be predicted from the baseline characteristics of the mother: overloading of the right ventricle is the most severe risk factor for maternal cardiac complications while hypoxia and arrhythmias are the most dangerous risks to the fetus[6]; the most important predictor of an unsuccessful outcome is an increased hemoglobin value and a decreased arterial oxygen saturation[6,9,10]. Hypoxemia frequently causes intrauterine deaths, premature births and low birth weights if the arterial oxygen saturation at rest is < 85%[6]. Supraventricular arrhythmias may recur and be resistant to drug therapy, causing recurrent miscarriages and intrauterine deaths[6]. The frequency of arrhythmias is due to the natural history of this anomaly including preexcitation and gradual enlargement of the right atrium[5]. Our patient with Ebstein's anomaly had only a mildly deformed tricuspid valve that remained asymptomatic well into adulthood. The anomaly was diagnosed after the incidental finding of a cardiac murmur and was not associated with other cardiac anomalies, cyanosis or preexcitation. The Celermayer echocardiographic grade of severity was 0.34 (grade 1) and therefore the situation was considered good[9]. A surgical intervention was not found necessary because the patient was in the NYHA functional class I and had no cyanosis or arrhythmias. The patient had two successful pregnancies without complications; the weights of the full-term infants were normal as they would have been in pregnancies of any non-cyanotic mother[5]. The offspring were healthy with a normal index of vitality and did not have congenital heart diseases. The third pregnancy miscarried at the 11th week of gestation. During the second and the third pregnancy the patient was taking digoxin for the prevention of atrial fibrillation, with a good tolerance to the drug. During each pregnancy no arrhythmias or signs of heart failure occurred and there was no worsening in the NYHA functional class; furthermore, the patient maintained a normal hemoglobin level and oxygen saturation. These two parameters have the highest predictive value for fetal wellbeing (when the oxygen saturation is > 85% and the hemoglobin at the beginning of the pregnancy is < 20 mg/dl), while the age of the mother is less important for the prognosis[6,10]. There was no deterioration of the patient's hemodynamic status after the two successful pregnancies. The miscarriage that ended the third pregnancy was not due to the mother's clinical conditions and considering that all other factors in the pregnancy remained unchanged, a possible fetal malformation could be hypothesized. The incidence of congenital anomalies in those who miscarry is unknown, but it might be higher than in the normal population[6].

The pregnancy-associated risk in Ebstein's anomaly may therefore be assessed with an appropriate evaluation of the anatomic and functional severity. In our patient no cardiomegaly, a low Celermayer grade of severity and a NYHA functional class I, were predictors of a good maternal survival and prognosis; on the other hand, the absence of cyanosis or arrhythmias and the normal hemoglobin levels and arterial oxygen saturation were predictors for a good prognosis for the fetus. Our case report is the only published case of a woman with Ebstein's anomaly who gave birth to two healthy unaffected full-term newborns after two uneventful pregnancies. Clinical monitoring and testing inclusive of the arterial oxygen saturation allow us to keep the worst risk factors under control with a better prognosis for the fetus. The caesarean section procedure has to be performed under the surveillance of both an obstetrician and a cardiologist. Our patient was given a general anesthesia in order to avoid fetal distress.

The risks of the mother and the fetus may be reduced by the collaboration of a multidisciplinary team that includes obstetricians, cardiologists and anesthetists during the course of the pregnancy, the delivery and even during the *postpartum*. This case report shows that pregnancy is well tolerated in patients with Ebstein's anomaly who are in NYHA functional class I and have no cardiomegaly, arrhythmias or cyanosis and confirms the importance of serial echocardiographic evaluation.

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