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# **Medical Expertise**

"Development of the European Network in Orphan Cardiovascular Diseases" "Rozszerzenie Europejskiej Sieci Współpracy ds Sierocych Chorób Kardiologicznych"

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# CASE SUMMARY

24-year-old woman with Ebstein anomaly and pre-excitation syndrome (WPWs.), treated previously with RF ablation of the accessory pathway (2006, 2007) presented at our center at 27 week of her first pregnancy. Her main symptoms included palpitations and fatigue, without dyspnea, chest pain or syncope.

Transthoracic echocardiography showed typical signs of Ebstein's anomaly, such as inferior displacement of the septal leaflet of the tricuspid valve (45 mm), right atrial (RA) and right ventricle (RV) enlargement (areas, 33.0 and 27.6 cm<sup>2</sup>, respectively), good RV function, no evidence of interatrial communication.

24-hour Holter ECG monitoring revealed sinus rhythm of maximal HR 124 beats/min, minimal HR 68 beats/min, and mean HR 81 beats/min, pre-excitations signs, no significant arrhythmias.

Propafenon (75 mg 3 times a day) and  $\beta$ -blocker therapy with metoprolol (23.75 mg/d) were used.

# DISCUSSION

Ebstein anomaly (EA) is a rare congenital cardiac defect with the prevalence of 0.3% to 0.5% in the general population, characterized primarily by abnormalities of the tricuspid valve and right ventricle (RV) (1).

An interatrial communication (ASD, persistent foramen ovale) and accessory pathways (Wolff–Parkinson–White syndrome) are commonly associated with EA (respectively, 80% -





94% and 6%–36%) (2, 3) and may lead to supraventricular tachycardia (4, 5).

Małopolska

Hemodynamic abnormalities observed during pregnancy depend on the severity of tricuspid regurgitation and the functional capacity of the RV. In pregnant women with impaired RV size and function, the increased stroke volume may be poorly tolerated and result in worsening of tricuspid regurgitation, raised atrial pressures, and increased right-to-left shunting. Even severe tricuspid regurgitation with heart failure can usually be managed medically during pregnancy. Treatment includes therapy for heart failure and arrhythmia (4). An increased risk of fetal loss, prematurity, and low birth weight in babies born to mothers with cyanosis have been reported (6). Symptomatic patients with cyanosis and/or heart failure should be treated before pregnancy or counseled against pregnancy.

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Women with EA without cyanosis and heart failure are classified as class II according to the World Health Organization (WHO) risk classification and usually tolerate pregnancy well (4). The presence of arrhythmia in the mother is associated with increased maternal and fetal risk. The patients with interatrial shunting can develop shunt reversal, cyanosis and paradoxical embolism during pregnancy (7, 8).

The preferred mode of delivery is vaginal in almost all cases (4). The risk of congenital heart disease in the offspring is reported in 4% to 6% of the cases (6, 9). Endocarditis prophylaxis in the peripartum period is not indicated in pregnant women with Ebstein's anomaly.

#### **EXPERT'S OPINION**

Systematic clinical, echocardiographic, ECG and Holter ECG monitoring is indicated. The medical therapy with propafenon (FDA C) and metoprolol (FDA C) should be continued.

In the case of clinical deterioration, RV dysfunction in TTE, life threatening arrhythmias earlier cesarean section should be performed.

When the tolerancy of pregnancy is good the preferred mode of delivery is vaginal. The management during labor should aim at eliminating factors leading to congestive heart failure, cyanosis, and arrhythmias. To maintain normal sinus rhythm during labor, adequate pain relief in the form of epidural analgesia is helpful (10). Assisted vaginal delivery is indicated.

Endocarditis prophylaxis in the peripartum period is not indicated in this patient.







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### CONCLUSION

Patients with EA, without cyanosis, heart failure, atrial communication usually tolerate pregnancy well (WHO risk classification II), nevertheless, participation of multidisciplinary team is recommended to establish the treatment.

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