

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

A 60-year-old woman with uncorrected total anomalous pulmonary venous return and atrial septal defect is presented. She was admitted to the hospital because of arrhythmias - recurrent supraventricular tachycardia, ablation was performed, but the procedure was unsuccessful. The cardiac CT revealed total anomalous pulmonary venous return and atrial septal defect (25x23mm). The patient was in WHO functional class III. The distance in 6-minutes walking test was 410m with desaturation (pulse oximetry after the test decrease from 87% to 75%). Maximum oxygen consumption in cardiopulmonary exercise test was only 9,4ml/kg/min. Echocardiography showed enlarged right atrium (area 48cm²) and right ventricle (RVOT prox 54mm), paradoxical movement of intraventricular septum, severe tricuspid regurgitation, while left ventricular size and ejection fraction were normal. The right heart catheterization revealed pulmonary hypertension (mean pulmonary pressure was initially 31mmHg and 25mmHg after NO), the pulmonary vascular resistance was normal (2,4 WU).

EXPERT'S OPINION

Uncorrected total anomalous pulmonary venous return and atrial septal defect in a patient 60 years of age is a rare finding, because surgical correction is performed in infancy in most of the cases. This patient has symptoms - recurrent supraventricular arrhythmias and dyspnea (NYHA f.cl III). There is significant overload of the right chambers of the heart with Qp/Qs around 4:1 and severe secondary tricuspidal regurgitation, still there are no signs of irreversible changes of pulmonary circulation, RV function is preserved. So, the surgical correction in this case could be performed if the patient will agree with this method of treatment. Concerning the medicament treatment because of the recurrent supraventricular tachycardia episodes - the dose of propafenon must be corrected and another electrophysiological investigation should be proposed if medicament treatment will be



ineffective. Oral anticoagulants still have no indications in this case. |

CONCLUSION

|The surgical correction in this case could be performed if the patient will agree with this method of treatment. Because of the recurrent supraventricular tachycardia episodes - the dose of propafenon must be corrected. |