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

This is a rare case of 60 - years old woman with TAPVR and ASD II diagnosed in early childhood. Restriction of physical exercise tolerance and rhythm disturbances were observed but patient did not undergo surgical correction of the defect because of lack of informed consent of her parents and herself. In 2013 she was admitted to the cardiology department because of episodes of supraventricular tachycardia. The unsuccessful attempt of ablation was made and angio-CT was performed confirming the previous diagnosis of TAPVR. Since February 2013 exercise tolerance worsening was also noted. The euthyroid goiter and propensity for depressiveness was also diagnosed. In clinical assessment clubbing of the fingers, discreet cyanosis and murmur of 2/6 in Levine scale were noted. In lab tests NT-proBNP was elevated. ECG showed episode of supraventricular tachycardia, and I degree AV block. In the X-ray picture signs of pulmonary venous congestion and right heart enlargement are visible. Exercise tests showed clear exercise tolerance restriction. Echocardiography revealed RV dysfunction with marked RA enlargement and severe TV regurgitation. Angi-CT showed typical picture of supracardiac TAPVR with big vertical vein draining into the innominate vein and SVC. Hemodynamic assessment showed increased saturation in innominate vein, SVC and right heart, increased RV pressure (half of systemic pressure), increased pulmonary artery pressure, Qp:Qs 3.8:1 although the pulmonary resistance was low (2.4 UW). After the NO2 administration the mPAP increased from 11 to

25 mmHg.

LITERATURE REVIEW

TAPVR is a rare congenital heart defect (0.4 - 1.4% of all congenital heart defects), although in above patient it's most frequent variant was diagnosed (1, 2). Most of the patients require surgical correction in neonatal period or within the first few months of life (3). When not treated, symptoms of the defect depend mainly on the degree of pulmonary venous return obstruction, RV function and pulmonary artery bed resistance (4, 5). There are many cases of succesful corrective surgery for TAPVC in adults reported in the literature (6, 7, 8, 9, 10). Patients with untreated TAPVC often may develop symptoms of pulmonary hypertension because of the high blood flow through the pulmonary circulation (2, 3).

EXPERT'S OPINION

There are favorable conditions for the correction of the defect. Pulmonary resistance is low. Proper addressing of the TV regurgitation and residual interatrial communication is necessary. Correction of the defect may be considered after informed consent of the patient.

CONCLUSION

Although this is the rare case of congenital heart defect encounterd in 60 year-old woman, the hemodynamic conditions are favorable. The correction of the defect may be considered after informed consent of the patient. The poor RV function as a risk factor should be taken into account and tricuspid valve plasty as well as small residual interatrial communication should be done.

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