Medical Expertise

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

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This is a patient aged 57 years with pulmonary arterial hypertension associated with congenital heart disease in the form of atrial septal defect diagnosed at the age of 49 years. At present she is in WHO functional class III. When she was 50 she suffered from ischemic stroke. Furthermore, she has several risk factors for cardiovascular diseases including diabetes, hypertension, hypercholesterolemia and varicose veins of the lower extremities. In laboratory studies there is significantly elevated NT-proBNP level. Distance in 6-minute walk test is short - 340 m. In ECHO there are enlarged both atria and right ventricle, paradoxical movement of the interventricular septum. In right heart catheterization average pulmonary artery pressure is 44 mmHg, pulmonary vascular resistance is 9.7 Wood units.

The patient is a candidate for the targeted treatment of pulmonary arterial hypertension. The drug of choice is bosentan. In view of the criteria of the National Health Fund inclusion of this drug in patients after stroke is not possible despite there is no justification for such criteria in the literature nor in the summary of product characteristics for bosentan. In the absence of the

possibility of the use of bosentan I suggest to start treatment with sildenafil.

References:

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