

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

- Pulmonary hypertension was diagnosed for 57 year old female with atrial septal defect type II 8 years ago. Total hysterectomy with oophorectomy was performed at that time. One year later (7 years before) ischemic stroke of the right hemisphere with hemorrhagic transformation was diagnosed. The patient has some risk factors – such as arterial hypertension, dyslipidaemia, DM, varicose veins.
- The investigations revealed bidirectional shunt at the level of interatrial septal defect, mostly from left to right, severe pulmonary hypertension with overload of right ventricle, with high vascular resistance (9,7 Wood Units) RV lateral wall contractility was preserved, though high values of NT proBNP showed the signs of RV dysfunction.
- Medicament treatment is going on with specific treatment of PH – Sildenafil 25 mgx2. Still the patient remains in NYHA Class f. 3, other options of the treatment are of great importance..

EXPERT'S OPINION

Investigations: The vasoreactivity test should be performed. MRI should give additional information about RV function. The diagnosis of pulmonary embolization should be ruled out. Detail neurological investigation should be performed – its not clear the reason of the stroke, the neurological status of the patient at the moment.

Treatment options: Its not clear the duration of specific treatment of pulmonary hypertension and changes of clinical status of the patient with sildenafil. I would advise to augment dosage of sildenafil. If deterioration of the clinical status is obvious with this treatment, the possibility of adding endothelin antagonist should be reasonable.