





# **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

The presented patient is a young, 28-year-old male after the correction of tetralogy of Fallot in 1989, closure of a residual VSD in 1992 and transcutaneous balloon dilatation angioplasty of stenotic pulmonary artery branches in 2000. He leads an active life; his level of physical activity classifies him as NYHA I class. A single episode of fainting while skiing cannot be possibly associated with his heart defect. Similar fainting spells are observed in healthy individuals when they are in the mountains; examinations and tests performed in the described patient do not indicate either circulatory failure or any significant heart rhythm disturbances. In echocardiography, major deviations from the norm include moderate pulmonary valve incompetence with a low, most likely flow-associated gradient of approximately 28 mm Hg. A similar gradient is observed across the moderately incompetent tricuspid valve. Echocardiography and MRI show increased right ventricular volume. In order to determine the final recommendations for surgical intervention, it would be necessary to calculate both the left and right ventricular volume. Diagnostic management should also include pulmonary arterial branches, especially in view of the fact that in 2000, the patient was subjected to balloon dilatation angioplasty of one stenotic branch. Right heart catheterization combined with MRI data seem to be the most reliable method of assessing the tricuspid valve, the right ventricular morphology and function, the outflow tract from the right ventricle and the morphology of the pulmonary arteries. Prior to heart catheterization, it is necessary to re-diagnose the patient with respect to conduction and cardiac rhythm disturbances, so that possible electrophysiological procedures could be performed in parallel with catheterization. At present, the patient compensates for a moderate tricuspid and pulmonary valve incompetence, but the situation is expected to deteriorate in the future. The right ventricular volume is significantly increased and both valves in the right heart are incompetent, therefore, the patient should be cautiously followed-up and in case his right heart function deteriorates, he will be qualified for pulmonary artery valve implantation.







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#### DISCUSSION

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#### CONCLUSION

The patient should be cautiously followed-up and in case his right heart function deteriorates, he will be qualified for pulmonary artery valve implantation.

