

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 51 years old male with multilevel scoliosis from childhood and with quickly progressive decrease of physical activity and shortness of breath during daily functioning. Paroxysmal atrial fibrillation (AF) from 35 years old, permanent AF from year 2008. Laboratory findings revealed severely elevated BNP to 3581 pg/ml with normal markers of kidney and liver function. ECG and Holter monitoring showed atrial fibrillation without ventricular arrhythmias. Echocardiography revealed non dilated LV with signs of hypertrophy, enlarged LV atrium, slightly diminished LV ejection fraction (LVEF - 45%) with elevated pressure in LA (E/E' – 17). Hemodynamic investigation confirmed the echo results. Cardiopulmonary exercise test showed VO₂ max decrease to 12,33 ml (kg*min), Endomyocardial biopsy revealed hypertrophy with focal disorganization of cardiomyocytes and only small focuses of fibrosis in subendocardial region

EXPERT'S OPINION

At the moment pharmacological treatment of heart failure must be continued, maybe higher doses of diuretics could be used. Frequent follow up evaluation should be recommended for the patient.

The patient is a likely candidate for heart transplantation in the future, especially if VO₂/kg max decrease is detected.

CONCLUSION

1. The patient should undergo frequent follow-up evaluation with TTE and CPX check-up.
2. Heart transplantation should be considered if the patient's condition deteriorates (especially if VO₂/kg max parameter decreases).



REFERENCES

ACCF/AHA 2011 guidelines for the diagnosis and treatment of hypertrophic cardiomyopathy: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines. J Thorac Cardiovasc Surg. 2011 Dec