

## 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 present case concerns a 50 year old man with cardiomyopathy. Probably the heart disease is caused by a viral infection, which he had 8 years ago. Since that time, he has had atrial fibrillation, which required an ICD implantation. Medical history of the patient was presented during heart team consultation and sildenafil was recommended. During treatment there was a significant improvement in heart failure as well as a reduction in pulmonary resistance. However, the patient still shows severe mitral valve insufficiency in echocardiography study.

### DISCUSSION

The heart failure (e.g., cardiomyopathy ) can lead to the secondary pulmonary hypertension (PH) and right heart failure. There is a lot of papers talking about the positive effect of treatment with sildenafil in patients with pulmonary hypertension.

One of them is the work of Ramani et al (1) in 2010, who found that sildenafil is an effective treatment for these patients, although they suggest that further research is needed. Adir and colleagues (2) indicates that the left ventricular failure is the main cause of secondary pulmonary hypertension, while Singh et al (3) emphasize that the fixation of pulmonary hypertension in many patients, prevents the performance of a heart transplant .

They also point out that this problem affects both adults and children. In the group of 24 children the oral administration of sildenafil significantly improved the function of the right ventricle. The good results were confirmed by invasive tests.

Sansone et al (4) indicate that the right ventricular dysfunction after heart transplantation is a major complication in patients who have previously demonstrated PH. The authors emphasize that the use of sildenafil in transplant recipients may be beneficial and have good prognosis , Oral administration of sildenafil reduces pulmonary resistance and pulmonary artery pressure and increases cardiac output. But at the same time the authors mentions that the experience of oral administration of inhibitors of 5 - phosphodiesterase is

still limited.

Polish authors also contributed the study of sildenafil in transplant patients. The team of Zakliczyński et al (5) evaluated the efficacy and safety of sildenafil in 6 patients with pulmonary hypertension prepared for a heart transplantation. In 3 patients after 1 month of treatment, normalization of pulmonary artery pressure was noted. In other 2 patients pressure drop was satisfactory in the sodium nitroprusside test, while 1 patient presented obstructive hypertension. It has been shown that sildenafil treatment was well tolerated. The same group presented, that the use of sildenafil improves short, and long-term prognosis in patients after heart transplantation (6). Group De Santo et al (7) presented a group of 13 patients with right ventricular failure and pulmonary hypertension after heart transplantation who were treated with sildenafil. They found a positive effect of sildenafil on improving both right ventricular function and a reduction in pulmonary artery pressure.

Presented here the case of 50 year old patient with CMP is fascinating because it shows a spectacular positive effect of sildenafil. This may improve pulmonary vascular resistance and gives a better prognosis for the patient waiting for HTX. One cannot ignore also the role of intensive medical therapy as well as ICD implantation. These two factors together caused the patient's condition dramatically improved.

## EXPERT'S OPINION

In my opinion there are two aspects: one is the basic disease (CMP) and the second is the improvement the efficiency of the heart as well as the improvement of lung function (blood flow) after sildenafil administration. This is associated with a decrease in pulmonary resistance and increased stroke volume of the left ventricle. Good clinical outcome is also influenced by intensive cardiac treatment.

## CONCLUSION

I think that the function of the left ventricle should be observed, during sildenafil therapy. Surgery for mitral valve can be very effective. However, if left ventricular systolic function will not improve significantly, one should consider a heart transplant, especially at the time when pulmonary resistance is low. It might make a good prognosis for the patient especially if he will be qualified for the HTX

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