

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

Authors presented a case of a 20 year old male after Rastelli operation due to double outlet right ventricle, after closure of the atrial septal defect with Amplatzer septal occluder, and with persistent left superior vena cava. The main question that the authors asked is the reason for decreased exercise capacity in this patient being in functional class II.

LITERATURE REVIEW

Rastelli repair of double outlet right ventricle involves creation of intraventricular tunnel between aorta and ventricular septal defect and placing a conduit between the right ventricle and pulmonary artery. The risk factors for conduit insufficiency include: smaller size, younger age of implantation, pulmonary artery stenosis, transposition of the great arteries, process of sterilization and conservation. The main complications in patients with conduit include: mature out of the conduit, progressive stenosis, aneurysms and pseudoaneurysms. The main symptoms are dyspnea, palpitations, syncope and sudden cardiac death.

Measurement of stenosis of the conduit in echocardiography is usually unreliable therefore estimated pressure in the right ventricle is used to assess its severity. Cardiovascular magnetic resonance and computed tomography are suitable to assess the level of stenosis. Right heart catheterisation is required when surgical treatment is considered. The indications for intervention include increase in right ventricular systolic pressure >60 mmHg in symptomatic patients. In asymptomatic patients with severe right ventricular tract obstruction and/or pulmonary regurgitation intervention should be considered when at least one of the following features is present:

- decrease in exercise capacity as measured by spiroergometry
- progressive dilation of the right ventricle
- progressive dysfunction of the right ventricle
- progressive tricuspid regurgitation (at least moderate)
- systolic pressure in the right ventricle >80 mmHg

- persistent supraventricular and ventricular arrhythmias

The preferred method of treatment is percutaneous balloon dilatation of the conduit or implantation of pulmonary valve.

EXPERT'S OPINION

In catheterisation the gradient through the conduit was 15 mmHg and the systolic blood pressure in the right ventricle was 30 mmHg. Unfortunately there is much inconsistency between the Fick and thermodilution measurement of the cardiac output, probably some repeated measurements should have been made. Despite the homograft calcifications and stenosis of 45% seen in multislice computed tomography hemodynamic data do not support the need for intervention. However strict echo monitoring is necessary. Severe tricuspid regurgitation seems to be one of the reasons for reduced exercise capacity. I would recommend repeated measurements of Vmax.

CONCLUSION

Observation with monitoring of echocardiographic right ventricular systolic pressure and exercise capacity is recommended.

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