Medical Expertise*

"Development of the European Network in Orphan Cardiovascular Diseases"
„Rozszerzenie Europejskiej Sieci Współpracy ds Sierocych Chorób Kardiologicznych"

Title: Young adult after surgical correction of double outlet right ventricle.

RCD code: IV-6.O

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[* The expertise should be written in English]

Medical Expertise

CASE SUMMARY

This is a case of a 20-year-old male with double outlet right ventricle. At the age of 4 he underwent a Rastelli procedure with a Gore-Tex patch used to reconstruct the left ventricle outflow tract (LVOT) and a pulmonary homograft implanted to restore the outflow from the right ventricle (RV). Subsequently, at the age of 12, the patient underwent a successful percutaneous correction of atrial septal defect with an Amplatzer Septal Occluder. Two years later he developed persistent atrial flutter and electrical cardioversion was required to reinstate the sinus rhythm (SR).

He first presented at the age of 18. He was then asymptomatic, in SR with intermittent 2nd degree atrio-vetricular block and RBBB. Echocardiography revealed moderetly depressed left ventricular ejection fraction (LVEF, 45%) with normal end-diastolic volume (EDV, 127ml) and trivial mitral regurgitation. The right ventricle was dilated but normally contracting (right ventricular outflow tract - RVOT - 31mm, right ventricular inflow tract – RVIT - 55mm, tricuspid annular plane systolic excursion - TAPSE - 22mm), the right atrium enlarged (RA area 30cm2) and a severe TR was present with calculated RVSP of 46mmHg. The patch separating the LVOT from RV was competent and there was no leak at the level of interatrial septum either. Additional testing showed maximal oxygen consumption of 24ml/kg and NT-proBNP plasma concentration of 95pg/ml and the patient was then treated conservatively. Currently, at the age of 20, he was readmitted due to heart failure symptoms in NYHA class II with NT-proBNP plasma concentration of 390pg/ml. Electrical cardioversion was needed again to treat recurrent atrial flutter. Repeated echocardiography showed stable function of the left heart (LVEF 47 %, EDV 131ml, trivial MR) and progressive dysfunction of the right heart (RVOT 40mm, RVIT 56mm, TAPSE 16mm, RA area 36cm2, severe TR with RVSP of 45mmHg). The flow within the homograft was somewhat turbulent and the systolic peak gradient between the RV and the pulmonary artery (RV/PA gradient) was calculated at 20mmHg. Consequently, multi-detector computed tomography (MDCT) was performed to better visualize the suspected stenosis within the homograft. It showed only some amount of calcification in the homograft wall but not within the cusps, however. The lumen cross-section area at the level of homograft was about 45% of that at the level of pulmonary trunk just proximal to bifurcation (2.6cm2 vs 4.77cm2). Additionaly, some more vascular abnormalities were found. Firstly, there was anomalous right coronary artery, originating from the non-coronary sinus and the left main course was between the pulmonary homograft and aorta. Secondly, a mirror image type right aortic arch along with persistent left superior vena cava were present. Invasive coronary angiography ruled out the possibility of left main functional compression. Cardiac catheterization showed RV/PA maximal gradient of 15mmHg, RVSP of 30mmHg, pulmonary vascular resistance of 3.02 HRUI with ratio to systemic vascular resistance of 0.08. The cardiac index was 3.83 (Fick) and the pulmonary to systemic flow ratio 1:1.

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DISCUSSION

Adult patients who had previously undergone Rastelli type operation can be expected to develop pulmonary conduit stenosis. Its clinical features comprise of exertional dyspnoea, palpitations, syncope, and sudden cardiac death. The diagnosis may be challenging due to problematic measurements of Doppler-derived gradients across the conduit. Current guidelines underline the significance of right ventricular systolic pressure (RVSP) estimation from tricuspid regurgitation (TR) velocity. Also cardiac catheterization may be necessary for reliable assessment of conduit stenosis. Symptomatic patients with RV systolic pressure >60 mmHg and/or moderate/severe PR should undergo surgery (IC). Asymptomatic patients with severe RVOT obstruction should be considered for surgery when at least one of the following criteria is

present: 1) decrease in exercise capacity, 2) progressive RV dilation, 3) progressive RV systolic dysfunction, 4) progressive TR (at least moderate), 5) RV systolic pressure >80 mmHg, 6) sustained atrial/ventricular arrhythmias (IIaC).

EXPERT'S OPINION

Cardiac anomaly was corrected in the childhood. The homograft implanted at that time might no more be suitable. Imaging studies suggest, that a stenosis of the homograft might now be ppresent. Giving the fact that the patient is merely symptomatic, he is in class II according to WHO, close observation and follow up seems resonable at this point. Clinical assessment including cardio-pulmoanry exercise test, 6 minute walking test or WHO functional class should be performed twice yearly. Decision upon replacement of the homograft should be based on the clinical status.

CONCLUSION

Close follow-up with clinical assessment is now recommended. Surgical replacement of the homograft seems to be the treatment of choice in the future. Decision regarding interention should be based upon patient's clinical status.

REFERENCES

Baumgartner H, Bonhoeffer P, De Groot NM, et al. ESC Guidelines for the management of grown-up congenital heart disease (new version 2010). Eur Heart J. 2010 Dec;31(23):2915-57.

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Expert's signature**

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