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

There was a case study of 27-years old man with Tetralogy of Fallot. In 1987 surgical procedure was performed - probably Blalock-Taussing anastomosis. The patient had no medical documents. He had been lost in follow-up until 2012.

In 2012 the patient was hospitalized due to low exercise tolerance and dyspnea (NYHA functional class II). Physical examination revealed central cyanosis. Laboratory studies have found polycythemia, thrombocytopenia. The cardiopulmonary exercise **test** showed severely impaired exercise tolerance and maximal oxygen consumption was 15,4 ml/kg/min. Echocardiography showed Enlargement of right ventricle, hypoplastic pulmonary trunk, ventricular septal defect . Shunt between left subclavian artery and left pulmonary artery was visible. The patient had no catheterization.

DISCUSSION

Pulmonary atresia and ventricular septal defect is considered the most severe form of tetralogy of Fallot. In unoperated patients the arterial blood supply to the lungs, provided by major aorto-pulmonary collateral arteries (MAPCAs), is one of the main determinants of survival (1). Presented patient probably had Blaloc-Taussing anastomosis, a surgical procedure used to increase pulmonary blood flow. According to the ACC/AHA 2008 Guidelines for the unusual case of a patient with tetralogy of Fallot who has undergone palliation with a surgical shunt, catheterization should be performed to assess the potential for

repair. The presence or absence of additional muscular VSDs may be determined, as well as the course and anatomy of the epicardial coronary arteries. The pulmonary architecture and vascular pressure and resistance should be delineated, because pulmonary artery distortion and PAH are frequent sequelae of palliative surgical shunts. Potential catheter interventions include elimination of collateral vessels or systemic–pulmonary artery shunts (2).

EXPERT'S OPINION

For further decision about surgical treatment heart catheterization should be performed.

CONCLUSION

The patient should be qualyfy for heart cathetherization. The pulmonary artery pressure and resistance of pulmonary vascular bad should be measure.

REFERENCES

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2. ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease.

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