

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

53 year old male with a tetralogy of Fallot, after Blalock - Taussig anastomosis in 1969 and after total correction in 1986. Currently patient in NYHA class II/III, with moderate conduction and rhythm disorders, with symptoms of right ventricle insufficiency, severe pulmonary valve regurgitation and moderate tricuspid valve regurgitation, with chronic hepatitis C and symptoms of the liver cirrhosis, with thrombocytopenia, trivial residual VSD, significant patent systemic - pulmonary shunt and pulmonary hypertension.

DISCUSSION

Presented patient is after transannular correction of tetralogy of Fallot with patch enlargement of right ventricle outflow tract (RVOT). This procedure results in a significant RVOT regurgitation caused by absence of the pulmonary valve. In this particular case the patient has also patent systemic - pulmonary shunt with left to right blood flow what significantly increases the pulmonary pressure and right ventricular volume load. This results in right ventricular hypertrophy, tricuspid insufficiency, congestion in systemic veins and right atrium hypertrophy. Enlarged thickness of the wall of right atrium and right ventricle promotes a cardiac arrhythmias, which significantly increases complaints.

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EXPERT'S OPINION

| The fundamental problem of the presented patient is right ventricle insufficiency as a result of pulmonary hypertension due to persistence of left to right shunt in patent Blalock - Taussig anastomosis. The best solution for the patient should be closure of persistent Blalock - Taussig shunt. Due to reduced liver function should be avoided extensive operations in a cardiopulmonary bypass. Cardiac intervention procedure performed in the course of measurements before and after closing anastomosis seems to be safe. Due to catheterization there is possibility to assess the stenosis of proximal segment of left pulmonary artery with any invasive intervention if would be required.

I would suggest to perform cardiac catheterization with evaluation of pulmonary pressure and systemic saturation before and after closure of patent Blalock - Taussig shunt. If it would be necessary to perform procedure of enlargement of proximal left pulmonary artery. |

REFERENCES

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