

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 authors presented a case of a 53 year old patient with congenital heart disease - Tetralogy of Fallot after surgical correction with patent Blalock – Taussig. In 1969 r. – left side Blalock-Taussig shunt was performed. In 1986 r. – total correction of ToF was done. Blalock – Taussig shunt was preserved.

Concomitant conditions include: cirrhosis of liver due to chronic hepatitis C, venous insufficiency, thrombocytopenia, cardiac arrhythmias - episodes of atrial fibrillation and atrial flutter;

The Patient complains of exercise intolerance, dyspnoea – NYHA II/III class, and peripheral oedema since 06.2014r.

On echocardiography:

- enlarged right ventricle (41 mm),
- enlarged right (area 23 cm²) and left atrium (area 24 cm²),
- ejection fraction of left ventricle of 56%;
- severe pulmonary valve regurgitation (PHT 60 ms, gradient 40 mmHg) with calcifications of valve annulus and trunk.
- moderate tricuspid regurgitation (tricuspid annulus 39 mm, TAPSE 14 mm, fala S' 10 cm/s, RVSP 70 mmHg; vena cava inferior 28 mm, not movable)
- leavings of ventricular septal defect with left- right shunt.

On magnetic resonance imaging:

- enlargment of right ventricle and mild enlargment of right atrium(26,6 cm²),
- low ejection fraction of right ventricle (EF30%; EDV=250 ml; ESV=175 ml; SV=74 ml; mass 61g)
- preserved EF of the left ventricle (EF 55%);
- dyssynchronous ivs movement,
- right sided aortic arch
- pulmonary regurgitation,
- mild tricuspid regurgitation,
- ostial stenosis of the left pulmonary artery

-dilation of vena cava inferior

Right heart catheterization:

-elevated LV end-diastolic pressure (118/9/23 mmHg),

- elevated pressures in PA and the right heart (right atrium - 28/24/24 mmHg, after NO 27/23/24 mmHg; right ventricle – 90/9/25 mmHg; PA – 84/10/38 mmHg, after NO 71/22/35 mmHg)

- PVR – 94ARU; TPR – 448ARU; VSR – 972ARU.

- pressure measured in left PA – distal to Blalock – Taussig shunt was 33 mmHg. Qp/Qs 1,1:1.

In ventriculography – small shunt to right ventricle in upper part of ventricular septum.

No pulmonary thromboembolic disease, no coronary arteries disease.

DISCUSSION

In this case patent Blalock – Taussig may be evaluated analogously to the situation of patent arterial duct. Because of that it seems correctly to proceed according to actual ESC guidelines for the management of grown – up congenital heart disease (2010), which recommends PDA closure in patients with signs of LV volume overload (class I c) and in patient with pulmonary arterial hypertension but pulmonary artery pressure < 2/3 of systemic pressure or pulmonary vascular resistance < 2/3 of systemic vascular resistance (class I c)

In Tetralogy of Fallot pulmonary hypertension is rather rare (for example: in case of vast MAPCAs).

According to actual ESC guidelines for diagnosis and treatment of pulmonary hypertension (2009) adequate recording of PWP is required for the differential diagnosis of PH due to left heart disease and in rare cases, left heart catheterization may be required for direct assessment of LV end-diastolic pressure.

EXPERT'S OPINION

In this case part of systemic output goes to pulmonary circulation through patent Blalock – Taussig, causing overload of pulmonary circulation and at the same time overload of left ventricle. Therapy of PAH might be considered if only PCWP and LVEDP are confirmed that are low. It seems reasonable to perform Blalock – Taussig occlusion test. In case of good reaction closure of Blalock – Taussig could be considered.

The case should be consulted once again during next CRCD meeting after re-catheterization with assessment of end-diastolic pressure in left ventricle and temporary occlusion of Blalock-Taussig.

REFERENCES

ESC guidelines on the management for diagnosis and treatment of pulmonary hypertension (2009).

ESC guidelines for the management of grown – up congenital heart disease (2010).