

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

This is the history of the patient after correction of tetralogy of Fallot. The Blalock - Taussig anastomosis was performed when he was 8, while total correction in 25 years of age (closure of the VSD, and dilatation of RVOT)

Currently the patient presents arrhythmias: episodes of atrial fibrillation / atrial flutter; drug-induced bradycardia. He also suffers from chronic hepatitis C. He was treated with IFN and ribavirin (intolerance of the drug). Currently, in the ultrasound - symptoms of cirrhosis of the liver. He presented also venous valves insufficiency and thrombocytopenia approx. 80-100 thousand cells/mm³. The NYHA class II / III, shortness of breath and a significant degree of weakness. The peripheral edema and varicose in both legs were observed. The Echo study revealed RVSP about 70 mmHg and a residual VSD shunt (left to right). In the cath. significantly increased late diastolic pressure in the left ventricle and increased pressure in the pulmonary artery with normal pulmonary vascular resistance. Qp / Qs was 1.1: 1. Treated: Eplerenone 25 mg 1-0-0, 1-1-0 Furosemide, Potassium 1-0-1, 23.75 mg metoprolol 1 / 2-0-0 enoxaparin 60 mg 1-0-1, Hepa - Merz 1 -1-1, Cyclo 3 fort 1-0-0, 1-0-0 Detramax, Pantoprazole 20 mg 1-0-0

DISCUSSION

Echocardiography and cardiac catheterization did not show how wide is the BT anastomosis . Based on own experience and the literature it is known that BT anastomosis may be responsible for pulmonary hypertension in some cases (1,2).

Yamaki analyzed 34 patients with TOF: 13 with the BT shunt and 21 without BT. He indicated that the cases with BT revealed thicker small pulmonary arteries than in the group

without BT (2).

The use of MR allows to assess the hemodynamic changes in patients with TOF after correction (3,4).

In our patient, the laboratory tests revealed the elevated level of BNP. It is a sensitive indicator of heart failure (5,6).

Lim et al. indicate that the development of pulmonary hypertension in patients with TOF may occur when aorto-pulmonary anastomosis (MPACAs) are present. In such cases, the beneficial effect can be achieved through the sildenafil administration. What more, sildenafil is well tolerated and the therapy leads to an improvement of saturation in those patients (7).

In this patient it seems reasonable complement of diagnostic tests. Probably the re-evaluation of cardiac catheterization to indicate the importance of BT anastomosis is needed. The patient still requires pharmacology treatment.

In the cases of arrhythmia, the ablation may be helpful (8).

As more and more patients are adults after total correction of TOF, such a problem will occurred more frequently in the future (9-11)]

EXPERT'S OPINION

[In my opinion the patient presents multi problem disorders, but in this moment the right and left ventricle failure as well as pulmonary artery hypertension dominates.

The prognosis worsens of hepatic cirrhosis and thrombocytopenia.

Small residual VSD does not matter, but it is not known what is the role of persistent Blalock-Taussig anastomosis.

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CONCLUSIONS

It seems reasonable complement of diagnostic tests. Re-evaluation of cardiac catheterization is needed to indicate the importance of BT anastomosis. The patient still requires pharmacology treatment.

REFERENCES

1. Swamy P, Bharadwaj A, Varadarajan P et al: Echocardiographic Evaluation of Tetralogy of Fallot. Echocardiography. 2014 May 29. doi: 10.1111/echo.12437. [Epub ahead of print]
2. Yamaki S: Pulmonary vascular disease in shunted and nonshunted patients with tetralogy of Fallot. Tohoku J Exp Med. 1990 Oct;162(2):109-19.
3. Yap J, Tan JL, Le TT et al: Magnetic resonance imaging predicts exercise capacity in adult operated tetralogy of Fallot: a retrospective study. BMC Cardiovasc Disord. 2014 Sep 23;14:122-4.

4. Rao UV, Vanajakshamma V, Rajasekhar D et al.: Magnetic resonance angiography vs. angiography in tetralogy of Fallot. Asian Cardiovasc Thorac Ann. 2013 Aug;21(4):418-25.
5. Kapoor PM, Subramanian A, Malik V et al: B-type natriuretic peptide as prognostic marker in tetralogy of Fallot surgery. Asian Cardiovasc Thorac Ann. 2014 May 13. pii: 0218492314534247. [Epub ahead of print]
6. Eindhoven JA, Menting ME, van den Bosch AE et al: Associations between N-terminal pro-B-type natriuretic peptide and cardiac function in adults with corrected tetralogy of Fallot. Int J Cardiol. 2014 Jul 1;174(3):550-6.
7. Lim ZS, Vettukattill JJ, Salmon AP, Veldtman GR: Sildenafil therapy in complex pulmonary atresia with pulmonary arterial hypertension. Int J. Cardiol. 2008 Oct 13;129(3):339-43..
8. Kapel GF, Reichlin T, Wijnmaalen AP et al: Left-sided ablation of ventricular tachycardia in adults with repaired tetralogy of fallot: a case series. Circ Arrhythm Electrophysiol. 2014 Oct;7(5):889-97.
9. Talwar S1, Meena A, Choudhary SK et al: Repair of tetralogy of fallot in or beyond the fourth decade of life. Congenit Heart Dis. 2014 Sep-Oct;9(5):424-32.
10. Martinez RM, Ringewald JM, Fontanet HL et al: Management of adults with Tetralogy of Fallot. Cardiol Young. 2013 doi: 10.1093/ejcts/ezt505. Epub 2013 Nov 1.
11. Starr JP: Tetralogy of Fallot: yesterday and today. World J Surg. 2010 Apr;34(4):658-68 |