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 54-year-old man with Tetralogy of Fallot underwent surgical correction at 30 years of age. The patient had an encephalitis in early childhood and right hemisphere damage in adultery. Now the patient presents the symptoms of easy fatique. On admission the patient's general condition was good. In physical examination the murmur in pulmonary valve pole – 4/6 Levine's scale was observed. During the hospitalization cardiac catheterization was performed with the following results: right atrium 16/13/12 mmHg, right ventricle 43/3/10 mmHg, pulmonary artery 43/9/14 mmHg, left ventricle 142/5/21 mmHg, aorta 147/23/77, PCWP 13/10/12 mmHg; Sat O2: right atrium 71.0 %, right ventricle 70.1%, pulmonary artery 72.4%, aorta 97.8%; cardiac index was 2.24 l/min/m2 and cardiac output 3.86 l/min. Laboratory studies showed: RBC – 5030000, WBC- 5230, Hg- 15.3 g/dl, INR- 1.11, ASPAT- 20 U/L, ALAT- 25 U/L, Creatinine – 73 umol/L; total cholesterol 5.35 mmol/l, LDL 3.86 mmol/l.

In ECG was sinus rhythm 60 bpm with RBBB and sings of right ventricle hypertrophy. Echocardiography showed enlarged right ventricle, hyperthophy of ventricle sep tal, paradoxical septal motion, preserved left ventricle ejection fraction, EF 56%, moderate pulmonary valve regurgitation, moderate/ sever aortic valve regurgitation, holo-diastolic flow in the descending aorta, end diastolic velocity 0,17 m/s, peak gradient across the right ventricular outflow tract 32/15 mmHg, turbulent flow in the pulmonary artery, mitral valve regurgitation on anterior leaflet, gradient RVOT prox 31 mmHg.

MR showed hyperthrophy of left ventricle and acinetic of segments of right ventricle anterior

wall.

DISCUSSION

Tetralogy of Fallot (ToF) is a cyanotic congenital heart disease. Prognosis after surgical correction is good. Long-term problems include arrhythmias, both supraventricular and ventricular, and residual hemodynamic problems, which are generally well tolerated for many years; these include residual ventricular septal defect and pulmonary stenosis but, most commonly, pulmonary regurgitation, with its consequent right ventricular dysfunction and tricuspid regurgitation. Although the most common reason for repeat surgery in the adult after TOF repair relates to problems in the right ventricular outflow tract, the aortic regurgitation is often forgotten. Aortic regurgitation is an acquired complication which earlier radical repair may prevent the complication.

We present a patient after ToF repair with aortic regurgitation.

According to ESC Guidelines the indications for intervention after repair of Fallot's syndrome include:

1. Aortic valve replacement in patients with severe aortic regurgitation with symptoms or signs of left ventricular dysfunction (I C)

2. Pulmonary valve replacement in symptomatic patients with severe pulmonary regurgitation and/or stenosis (right ventricular systolic pressure >60 mmHg, tricuspid regurgitation velocity >3.5m/s) (I C)

3. Pulmonary valve replacement in asymptomatic patients with severe pulmonary regurgitation and/or stenosis when at least one of the following criteria is present:

• Decrease in objective exercise capacity

• Progressive right ventricular dilation or systolic dysfunction

• Progressive tricuspid regurgitation (at least moderate)

• Right ventricular outflow tract obstruction with right ventricular systolic pressure >80 mmHg (tricuspid regurgitation velocity >4.3 m/s)

• Sustained atrial/ventricular arrhythmias (IIaC)

4. VSD closure in patients with residual VSD and significant left ventricular volume overload or if the patient is undergoing pulmonary valve surgery (IIaC)

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EXPERT'S OPINION

Aortic regurgitation together with mitral regurgitaiotn appear to be the most important issues of this case. Surgical procedure will be difficult in this patient. Therefore, careful assessment of both insuficiencesis required. Echocardiography shows moderate mitral regurgitation. Hemodynamic obtained on right heart catheterization are not indicative for surgery. Conservative treatment is indicated with regular check-ups. Close observation is recommended.

CONCLUSION

Conservative treatment and close follow-up is recommended.

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