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 a case of a 54-year-old man with tetralogy of Fallot who underwent surgical correction when he was 30 years of age.

The patient had an encephalitis in early childhood and right hemisphere damage persisted until adulthood.

The main current symptom is easy fatigue.

On admission the patient's general condition was good. Right heart catheterisation revealed decreased cardiac index and normal pulmonary pressure, and normal gradient through the right ventricular outflow tract. Echocardiography showed moderate pulmonary valve regurgitation, moderate-to-severe aortic valve regurgitation, and moderate mitral regurgitation. Cardiovascular magnetic resonance revealed hyperthrophy of left ventricle and akinetic segments of right ventricle anterior wall.

LITERATURE REVIEW

Tetralogy of Fallot (ToF) is a cyanotic congenital heart disease first described by Etienne-Pouis Arthur Fallot composed of subpulmonary infundibular stenosis, ventricular septal defect, rightward deviation of the aortic valve with biventricular origin of its leaflets, and right ventricular hypertrophy. The major site of obstruction of the right ventricular outflow tract is the infundibular muscle but frequently also the pulmonary valve is narrowed. Reparative surgery involves closing the VSD and relieving the right ventricular tract obstruction which may also involve (1) pulmonary valvotomy when the valve is bicuspid

and dysplastic, (2) resection of the infundibular muscle, (3) right ventricular obstruction tract patch without disruption of the integrity of pulmonary annulus, (4) transannular patch which is used when the valve annulus is restrictive, this kind of patch creates pulmonary regurgitation (5) pulmonary valve implantation, (6) extracardiac right ventricular - pulmonary artery conduit in patients with pulmonary atresia, (7) angioplasty or patch augmentation of stenosed central pulmonary arteries, (8) closure of concomitant patent foramen ovale or atrial septal defect (9).

EXPERT'S OPINION

The main goal of the reparative surgery is to avoid pulmonary regurgitation even at the expense of mild to moderate pulmonary stenosis. This is because pulmonary regurgitation has a detrimental effect on right ventricluar function, and is associated with the increased risk of arrhytmia and sudden cardiac death. Pulmonary valve implantation is recommended for severe pulmonary regurgitation and is associated with symptomatic improvement and improved right ventricular function. This procedure has a low operative risk. The timing of implantation of bioprothesis is influenced by its relatively short life span therefore some delay this operation. On the other hand too late operation may lead to an irreversible damage of the right ventricle. Current indications for pulmonary valve implantation in the presence of severe pulmonary rregurgitation late after tetralogy of Fallot include:

- one of the floowing criteria must be present: symptoms, atrial or ventricular arrhytmia, decrease in exercise tolerance, evidence of right ventricular dilation and decrease of right ventricular ejection fraction, new-onset tricuspid regurgitation, prolonged QRS $\geq$ 180 ms and evidence of its serial increase,

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- at least two of the following criteria must be present: right ventricular to left ventricular volume greater than 2:1, right ventricular end diastolic volume index of > 150 ml/m2, severe pulmonary regurgitation with any degree of residual stenosis, combined lesions that may not have been indications for intervention in isolation, contemplating pregnancy.

According to ESC Guidelines the indications for intervention after repair of tetralogy of Fallot 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)

CONCLUSION

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Patient requires regular follow-up with the evaluation of the exercise capacity (cardiopulmonary exercise test) and severity of valvular regurgitation as well the function and size of the right ventricle.

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