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

A 35 years old male with corrected tetralogy of Fallot (ToF) modo Waterstone (1977) and reoperation with total correction (1981) and surgical correction of coarctation of the aorta (CoA) with by-pass protesis implantation followed by stent implantation due to re-coarctation (1994, 1995, 2012) was admitted due to progressive limitation of exercise capacity. Additionally, he has double aortic arch with occluded posterior arch distal to left subclavian artery branch-of, arterial hypertension (ESC class II). He is in class II by New York Heart Association (NYHA). He has systolic-diastolic murmur over the heart audible during physical examination. No differences in blood pressures (BP) on extremities (upper left – 130/80 mmHg, upper right – 120/80 mmHg, lower left – 120/80 mmHg). ECG reveals sinus rhythm 70 bpm, right axis deviation, right bundle branch block (RBBB). Holter ecg monitoring revealed paroxysmal atrial fibrillation. Echocardiography showed enlargement of both right atrium (RA) and right ventricle (RV) and significant pulmonic valve regurgitation (PR). Right heart catheterization did not disclose signs of shunts or pathological gradients in the right heart or pulmonary arteries. No pulmonary stenosis was confirmed. The authors are hesitant regarding the treatment of PV regurgitation

DISCUSSION

Tetralogy of Fallot (ToF) is a cyanotic congenital heart disease. Surgical correction is a procedure of choice. It is usually performed in the infancy. The prognosis is favourable after the procedure. More than 90% survive to adulthood and live active, healthy, and productive lives. Sometimes reoperations are required.
Coarctation of the aorta (CoA) is a congenital narrowing of the lumen in a section of the aorta. The narrowing is most commonly in the upper thoracic aorta but can occur in the abdominal aorta. It is present at birth and males are more often affected than females. Clinical symptoms are variable and depend on the position, degree and extent of the narrowed segment of the aorta. Traditionally the treatment requires open heart surgery. Balloon

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angioplasty is an alternative option but recurrence, aneurysm and aortic dissection remained disadvantages of both treatments.

Combination of these two anomalies is exceptionally rare. Given the embryological theories for development of the CoA, one of which suggesting that decreased aortic blood flow due to left heart abnormalities leads to underdevelopment of the aortic isthmus and thus to CoA [1]. For this reason right heat obstructive anomalies, such as ToF with right-to-left shunting results in increased aortic blood flow, what, following the theory, protects against the development of CoA. The actual pathogenesis of the presence of both anomalies remains unknown. Nevertheless, there have been a few cases of ToF and CoA described so far [2,3,4].

EXPERT'S OPINION

Definite treatment of ToF often results in PR. The degree of insufficiency may vary depending on surgical approach and on the presence or absence of associated pulmonary artery stenosis. Chronic, long-standing severe PR leads to the enlargement of the right ventricle due to increased workload, what is than associated with increasing tricuspid insufficiency. All of these contributes to the RV enlargement and eventually to the RV failure. Therefore PR repair may be necessary to restore PV competence and improve RV function. Although optimal timing of PV replacement remains uncertain, this option should be considered in symptomatic patients including those with moderate or severe RV volume overload, RV dysfunction, tricuspid valve regurgitation, and clinical arrhythmias attributable to the right heart enlargement/dysfunction.

According to ESC Guidelines the ***indications*** for PV intervention after repair of Fallot's syndrome include [5]:

1. Pulmonary valve replacement in symptomatic patients with severe pulmonary regurgitation and/or stenosis (right ventricle systolic pressure >60 mmHg, tricuspid regurgitation velocity >3.5m/s) (I C)
2. 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 ventricle systolic pressure >80 mmHg (tricuspid regurgitation velocity >4.3 m/s)
   - Sustained atrial/ventricular arrhythmias (IIaC)

This patient experiences limitation of exercise capacity, enlargement of the RV and RA, elevation of the RA pressure and has developed paroxysmal atrial fibrillation. These are present in the setting of severe PR. Thereafter, PV replacement should be advised to this

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patient. Percutaneous PV implantation (PPVI) is an alternative option to surgical approach. If applicable from anatomical point of view, PPVI should be considered.

CONCLUSION

This patient has indications for PV repalecement. If applicable percutaneous PV implantation should be considered.

REFERENCES

1. Rudolph AM, Heymann MA, Spitznas U. Hemodynamic considerations in the development of narrowing of the aorta. Am J Cardiol 1972; 30: 514-525.
2. Yoshigi M, Momma K, Imai Y. Tetralogy of Fallot with coarctation of the aorta. Cardiol Young 1994; 4: 75-78.
3. Günthard J, Murdison KA, Wagner HR, et al. Tetralogy of Fallot and coarctation of the aorta: a rare combination and its clinical implications. Pediatr Cardiol 1992 ;13 :13-40.
4. Bullaboy CA, Derkac WM, Johnson DH, et al. Tetralogy of Fallot and Coarctation of the Aorta: Successful Repair in an Infant. The Annals of Thoracic Surgery 1984 ;38 :400-401.
5. Baumgartner H, Bonhoeffer P, De Groot NMS, et al. ESC Guidelines for the management of grown-up congenital heart disease. European Heart Journal (2010) 31, 2915–2957

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