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 medical records I have received for reviewing describe a 54-year-old patient, in whom TOF was surgically repaired 24 years earlier. The patient is relatively young; the surgeon performing the repair achieved a satisfactory result. No typical residual defects are seen, as it is common in post-TOF repair patients. The current clinical issue is practically limited to the aortic valve and thus, the defect should not pose a significant problem in case surgical treatment or any other intervention treatment is undertaken. The relatively young age of the patient definitely favors considering the cusp plasty with cusp margin elongation or – if the option is not feasible – the classic procedure of aortic valve replacement, optimally using a mechanical valve. Should the conditions of valve replacement be very difficult (due to calcifications), an attractive technique may be implantation of a sutureless valve. Interventional transvascular valve implantation is possible if the aortic valve annulus does not exceed 26 mm. The limitations of the method are associated with valve annulus distension, most commonly seen in this group of patients, what rules out transvascular valve implantation.

LITERATURE REVIEW

BACKGROUND

Tetralogy of Fallot (TOF) is a congenital heart defect that relatively commonly coexists with other anatomical and physiological malformations. The most frequently observed concomitant defects include coronary anomalies, anomalous pulmonary vascular system, CAVC, RAA, LSCV, etc. A significant additional strain on the patient suffering from this severe defect is produced by valve regurgitation, noted both in the right and left heart. A coexisting aortic valve anomaly may be congenital in character (in very rare instances) or acquired – for example as a consequence of past inflammatory processes, or else result from a

iatrogenic damage inflicted in the course of surgical correction of the primary defect. One should also bear in mind the fact that surgical correction may lead to disturbed dynamics of the aortic valve, e.g. due to pulling the valve annulus or suturing the aortic valve cusp. The reconstruction of the outflow tract from the left ventricle while patching the ventricular septal defect of the malalignment type at times changes the direction of blood flow, what many years later may result in accelerated degeneration and dysfunction of the valve.

Repair procedures of tetralogy of Fallot performed in childhood often require surgical or cardiological interventions many years later [1]. Most commonly, the reasons include secondary obstruction of the right ventricular outflow tract, residual defects and arrhythmias, but the left ventricular outflow tract – although rarely – may also be subject to necessary interventions [2]. The anatomy of tetralogy of Fallot is typically associated with distension of the aortic orifice [3,4]. The earlier the defect is repaired, the less severe damages develop that are related to overloading of the aortic valve [5]. However, distension of the aortic orifice (distension of the aortic valve annulus [8], dissecting aneurysm [7]) is observed even following tetralogy of Fallot repair performed in childhood.

Performing surgical repair is supported by the patients' lives being endangered by progressive left ventricular hypertrophy (or bi-ventricular hypertrophy), the risk of arrhythmias or sudden death [8].

EXPERT'S OPINION

The patient should be treated surgically. One should consider the aortic valvuloplasty (cusp elongation) if the anatomical conditions allow for performing the procedure. Aortic valvuloplasty offers an additional advantage consisting in avoiding fixing an artificial valve in the region of the patch closing the ventricular septal defect additionally complicated by coexisting calcifications. For this purpose, the patient should be subjected to a detailed assessment using transesophageal echocardiography. If valvuloplasty is not feasible, the patient qualifies for implantation of an artificial aortic valve.

Aortic valvuloplasty, if technically feasible, is a solution that is advantageous for the patient. The final decision should be arrived at intraoperatively. Implantation of an aortic valve, in spite of additional strains, should not, nevertheless, be an insurmountable technical barrier.

The decision to operate should not be delayed. Based on the clinical data (echocardiography), I assert that the patient requires urgent surgery.

CONCLUSION

The opinion addresses a 54-year-old patient who had a successful repair of tetralogy of Fallot when 30 years of age. His current results of examinations do not indicate the presence of typical residual defects that are commonly seen in similar patients; however, the patient presents with significant aortic regurgitation. The degree of aortic valve insufficiency obliterates the good effects of the surgical repair performed years ago. The patient undoubtedly qualifies for surgical treatment, which offers a perspective of a long life in complete or almost complete physical comfort.

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