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

An 18-year-old patient with history of BAV due to aortic stenosis (1997 r.), with significant aortic regurgitation, pulmonary hypertension (diagnosed in July 2012, sildenafil treatment was ineffective), was admitted due to NYHA II exertional dyspnea. Echocardiographic examination disclosed severe aortic and tricuspid regurgitations, enlarged right ventricle and both atria, preserved left ventricular ejection fraction, distended main pulmonary artery. Heart catheterization revealed decreased cardiac output, elevated pressure in right heart chambers and pulmonary arteries, increased pulmonary vascular resistance with poor response to nitroxide. During hospitalization the patient was also diagnosed with subclinical hyperthyroidism.

DISCUSSION

Predictors of mortality in patients undergoing BAV have been previously reported. In the largest series of patients undergoing BAV, multivariate analysis demonstrated baseline functional status, baseline cardiac output, renal function, cachexia, female sex, left ventricular systolic function, and mitral regurgitation as independent predictors of mortality. The present study detected New York Heart Association functional class IV as a predictor for mortality. This is not surprising because functional status is a well-known determinant of prognosis in patients with severe AS treated medically. Renal insufficiency was an independent predictor for mortality; in previous studies, renal failure was reported to be associated with worse outcomes in high-risk patients undergoing aortic valve replacement surgery. High pulmonary artery pressure in our cohort and in other series was associated with poor outcome as well. Long-term survival is dismal after BAV alone. Transcatheter or surgical aortic

valve replacement should be pursued because BAV as a bridge to transcatheter or surgical aortic valve replacement is feasible, safe, and associated with better outcome than BAV alone. This may be especially true in patients who develop restenosis after the first BAV because the second BAV is associated with a lesser increase in AVA. (1)

In symptomatic acute severe AR, urgent/emergent surgical intervention is indicated. In chronic severe AR, the goals of treatment are to prevent death, to diminish symptoms, to prevent the development of HF, and to avoid aortic complications in patients with aortic aneurysm. On the basis of robust observational evidence, recommended surgical indications are as follows:

Symptom onset is an indication for surgery in patients with severe AR. Surgery should also be performed in patients with LV dysfunction or marked LV dilatation after careful exclusion of other possible causes. Although, in these patients, postoperative outcome is worse than in those operated on earlier, an acceptable operative mortality, improvement of symptoms and acceptable longer-term survival can be obtained. Surgery is indicated in asymptomatic patients with severe AR and impaired LV function (EF 50%) and should be considered if LV end-diastolic diameter (LVEDD) is 70 mm or LVESD is 50 mm (or 25 mm/m BSA in patients with small body size), since the likelihood of developing irreversible myocardial dysfunction is high if intervention is delayed further, and postoperative results are excellent if surgery is performed without delay. Good imaging quality and data confirmation with repeated measurements are recommended before surgery in asymptomatic patients. A rapid worsening of ventricular parameters on serial testing is another reason to consider surgery. The rationale for surgery in patients with ascending aortic and root dilatation has been best defined in Marfan patients. In borderline cases, the individual and family history, the patient's age, and the anticipated risk of the procedure should be taken into consideration. In patients with Marfan syndrome, surgery should be performed with a lesser degree of dilatation (≥50 mm). In previous guidelines, surgery was considered when aortic diameter was 45 mm. The rationale for this aggressive approach is not justified by clinical evidence in all patients. However, in the presence of risk factors (family history of dissection, size increase 2 mm/year in repeated examinations using the same technique and confirmed by another technique; severe AR; desire to become pregnant), surgery should be considered for a root diameter ≥45 mm. With an aorta diameter of 40–45 mm, previous aortic growth and family history of dissection are important factors which would indicate advising against pregnancy. Patients with Marfanoid manifestations due to connective tissue disease, without complete Marfan criteria, should be treated as Marfan patients. In individuals with a bicuspid aortic valve, the decision to consider surgery in aortic diameters ≥50 mm should be based on patient age, body size, comorbidities, type of surgery, and the presence of additional risk factors (family history, systemic hypertension, coarctation of the aorta, or increase in aortic diameter 2 mm/year in repeated examinations, using the same technique and confirmed by another technique). In other circumstances, aortic root dilatation ≥55 mm indicates that surgery should be performed, irrespective of the degree of AR. For patients who have an indication for surgery on the aortic valve, lower thresholds can be used for

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concomitant aortic replacement (45mm) depending on age, BSA, aetiology of valvular disease, presence of a bicuspid aortic valve, and intraoperative shape and thickness of the ascending aorta. Lower thresholds of aortic diameters may also be considered in low-risk patients, if valve repair is likely and performed in an experienced centre with high repair rates. The choice of the surgical procedure is adapted to the experience of the team, the presence of a root aneurysm, characteristics of the leaflets, life expectancy, and desired anticoagulation status. Vasodilators and inotropic agents may be used for short-term therapy to improve the condition of patients with severe HF before proceeding with aortic valve surgery. In individuals with chronic severe AR and HF, vasodilators (angiotensin-converting enzyme (ACE) inhibitors or angiotensin receptor blockers (ARBs)) are useful in the presence of hypertension, when surgery is contraindicated, or LV dysfunction persists postoperatively. A positive effect of these agents, or dihydropyridine calcium channel blockers, in asymptomatic patients without hypertension in order to delay surgery is unproven. (2)

EXPERT'S OPINION

Despite the underlying causes in symptomatic acute severe AR, urgent/emergent surgical intervention is indicated. Timing of intervention and type of procedure should be carefully discussed with the patient because of his young age. By the time of procedure pharmacotherapy with vasodilators, inotropic agents and ACE –inhibitors should be considered.

REFERENCES

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