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Case report

SURGICAL TREATMENT OF THE RARE CONGENITAL HEART MALFORMATION: QUADRICUSPID AORTIC VALVE

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Abstract

Quadricuspid aortic valve is a rare heart defect, with case frequency of 0.008% to 1.46%. In most cases, this failure is found accidentally during surgery or autopsy. The failure often progresses with the development of a valvular insufficiency that can manifest itself in adulthood and may require surgical treatment.

Quadricuspid aortic valve is a rare congenital cardiac malformation, usually found as an isolated anomaly, but it may be associated with coronary artery anomalies, ventricular septal defects, patent arterial duct and others. In article we report a 39-year-old female patient who was referred to us for the surgical treatment of the aortic valve insufficiency. This anomaly is rarely diagnosed as the majority of patients do not have any clinical signs of the anomaly. Clinical case underlines the importance of a use of the diagnostic transesophageal echocardiography.

Aortic valve was excised and mechanical double-hinged prosthesis "St. Jude Medical №21" was implanted; the defect of the interatrial septum was sutured. The postoperative period was smooth and uneventful, and the patient was discharged in a satisfactory condition on the 8th day after the operation.

In the presented case, quadricuspid aortic valve was not diagnosed, due to the fact that not all patients in the pre-operative period undergo transesophageal echocardiography. It has been recently proved that real-time three-dimensional 3D transesophageal echocardiography is the most effective method of making correct diagnosis in cases of regurgitation on the aortic valve, especially in the case of quadricuspid aortic valve.

Replacement of the aortic valve with quadricuspid aortic valve is a method of choosing a traditional treatment for a patient with aortic insufficiency. The immediate results are positive; however, further monitoring of the patient should be continued to assess the long-term outcome.

Keywords: *congenital quadricuspid aortic valve, transesophageal echocardiography.*

Introduction

Quadricuspid aortic valve (QAV) is a rare heart defect with case frequency of 0.008% to 1.46% [*Balington J., 1862; Hayakawa M. et al., 2014; Miyata Y. et al., 2014*]. In most cases, this failure is accidently found at the time of surgery or autopsy. The defect often progresses with the development of a valvular insufficiency that can manifest itself in adulthood and may require surgical treatment.

QAV found in the process of autopsy was first described by *Balington* in 1862 [*Balington J., 1862*]. *Simonds* reported 2 cases per 25666 autopsies with a frequency of 0.008%, and *Hurwitz and Roberts* about 2 cases per 6000 studies with a frequency of 0.033% [*Simonds J.P. 1923*; *Hurwitz L.E. et al., 1973*].

Tutarel et al. reported that the functional status of QAV was regurgitant in 74.7%, combined stenosis and regurgitation in 8.4%, stenotic - in 0.7%, and normally functioning - in 16.2% [*Tutarel O., 2004*]. *Yotsumoto et al*. reported that, among 616 patients who underwent an aortic valve operation, 9 (1.46%) patients had a QAV, all with significant aortic regurgitation except one with combined aortic stenosis and mild aortic regurgitation [*Yotsu-*

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moto G. et al., 2003]. Quadricuspid aortic valve was previously diagnosed only during surgery or autopsy, nowadays, more and more cases are diagnosed before surgery (0.013-0.043%) as a result of advances in echocardiography, and especially with the advent of transesophageal echocardiography.

According to Hurwitz and Roberts' anatomical classification, quadricuspid aortic valve is divided into the following 7 anatomical variations of quadricuspid valve, the most common consisting of 3 equal-sized and 1 smaller cusp. [*Hurwitz L.E. et al., 1973*]

Nakamura et al. have proposed the following more simplified classification of quadricuspid valve it into 4 variations, where type I: supernumerary valve between the left coronary cusp (LCC) and right coronary cusp (RCC); type II: supernumerary valve between the RCC and noncoronary cusp (NCC); type III: supernumerary valve between the LCC and NCC; and type IV: supernumerary valve indistinguishable because of NCC division into 2 equal parts. (Fig.1) [*Nakamura Y. et al., 2001*]:

According to many authors, the surgical indications for QAV are severe aortic regurgitation, severe aortic stenosis, or dysfunctional QAV associated with other lesions, such as occlusion of the left coronary ostium [*Olson L.J. et al., 1984; Idrees J.J. et al., 2015; Savino K. et al., 2015; Alpat S. et al., 2016; Shi-Min Yuan et al., 2016; Tsang M.Y. et al., 2016; Yuan S.M., 2016 Yotsumoto G. et al., 2003*]. Respectively the overall survival rate of QAV patients was 89.9% and 84.9% at 5- and 10-year follow-up [*Olson L.J. et al., 1984; Idrees J.J. et al., 2015; Savino K. et al., 2015; Alpat S. et al., 2016; Shi-Min Yuan et al., 2016; Tsang M.Y. et al., 2016; Yuan S.M., 2016 Yotsumoto G. et al., 2003*].

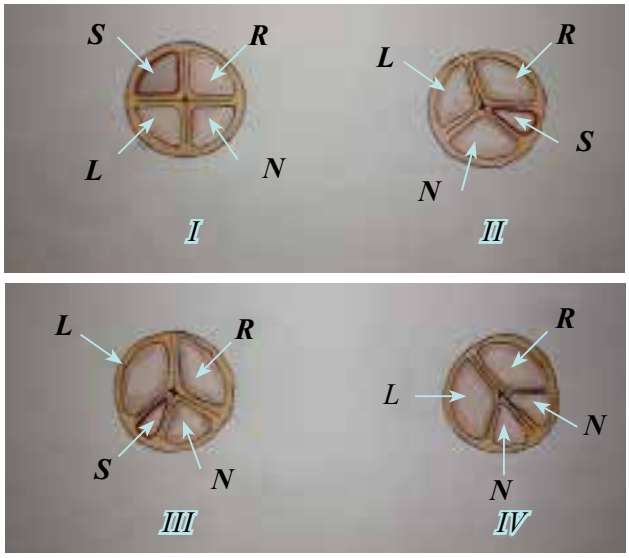

Figure 1. *Four anatomical variations in quadricuspid valves according to Nakamura Y. et al.: L -Left coronary cusp; R - right coronary cusp; N -noncoronary cusp; S - supernumerary cusp.* [*Nakamura Y. et al., 2001*]

Clinical case

A 39-year-old patient was admitted to join-stock company "National scientific center of surgery after A.N. Syzganov" for surgical treatment. Rheumatic aortic malformation was for the first time diagnosed by a cardiologist 3 months before hospitalization. The patient was sent to our hospital for the diagnosis confirmation and choice of further treatment tactics.

Split-sternum echocardiography findings were as follows: the diameter of aorta in the ascending section was 4.4 *cm*, sinus - 3.4 *cm*, sinotubular junction - 3.2 *cm*; the diameter of the ring was 2.0 *cm*, sealed tricuspid valves, fibrotic changes, limited movement of valves; vena contracta – 0.6 *cm*. The left coronary cusp was moderately connected through commissure, regurgitation of 2-3 degrees. The left atrium was 3.1 *cm*. The left ventricle: EDS (end diastolic size) – 6.2 *cm*, ESS (end systolic size) – 4.1 *cm*, EDV (end diastolic volume) - 198 *ml*, ESV (end systolic volume) -72 *ml*, IV (impact volume) - 126 *ml*, EF (ejection fraction) - 63%; mitral valve: the valves were sealed, mobile, regurgitation 0-1 degree. Pulmonary artery without features. The tricuspid valve: the valves are thin, mobile, regurgitating 0-1 degree. interatrial septum: defect was present, the size of the defect is 0.6-0.7 *cm,* the direction of discharge was from the left to the right. The average pressure in the right ventricle was 20 *mm Hg*. Contractility of the left ventricle myocardium was satisfactory. The conclusion: aortic vice, insufficiency of 2-3 degrees; secondary atrial septal defect.

Chest X-ray revealed that the roots were structural, the heart was without features, the aortas were not altered. ECG data: sinus rhythm. Heart rate of 62 beats per minute. Normal position of electrical axis of the heart.

Protocol operation. The operation was performed under conditions of artificial circulation and occlusion of the aorta. After longitudinal median sternotomy, the device of artificial circulation through the aorta and the eye of the right auricle

was connected. After antegrade cardioplegia, transverse aortotomy and aortic valve revision were performed. Unlike preoperative echocardiography, the aortic valve turned out to be four-cuspid with fibrous thickening of the valves and marginal calcification. The valve had the same all four valves, type A according to the anatomical classification of Hurwitz and Roberts [*Hurwitz L.E. et al., 1973*]. An additional flap was located between the right and left coronary valves. No anomalies of the coronary arteries were found. Aortic valve was excised and mechanical double-hinged prosthesis "St. Jude Medical №21" was implanted; the defect of the interatrial septum was sutured. The postoperative period was uneventful and smooth, and the patient was discharged in a satisfactory condition on the 8th day after the operation. Echocardiography revealed that the prosthesis was satisfactorily functioning, the size of the left ventricle and the function were preserved.

Discussion

Aortic insufficiency, if it is not corrected, plays a significant role in the development of heart failure and adversely affects the natural survival of patients. Correction of aortic valve insufficiency allows to improve the survival rates of the patients in the long-term period.

QAV is an extremely rare heart failure. According to autopsy data, the incidence of this disorder varies between 0.008% and 0.033%, whereas according to echocardiography, the frequency is 0.043% [*Olson L.J. et al., 1984; Yotsumoto G. et al., 2003; Savino K. et al., 2015; Alpat S. et al., 2016; Shi-Min Yuan et al., 2016; Tsang M.Y. et al., 2016; Yuan S.M., 2016; Das* A. *et al., 2018*]. Among patients who have undergone prosthetic replacement of the aortic valve, the frequency of the defect varies from 0.55% to 1,46%. Embryologically, the semilunar valves occur from the mesenchymal lining in the trunk of the aorta and the pulmonary artery. The abnormal leaf develops as a result of either an aberrant fusion of the aorto-pulmonary septum or an abnormal proliferation of mesenchymal cells in the main trunk. The development of the valves of the aortic valve occurs immediately after the development of the coronary artery insertion from the sinuses of Valsalva. Thus, it is quite possible that the anomalies in these two zones are embryologically interrelated. QAV usually develops as an isolated heart defect, but can also be combined with other vices, such as the open arterial duct, ventricular septal defect VSD, pulmonary valve stenosis, pseudo rheological fibro-muscular stenosis, and coronary artery anomalies. Anomaly of development of coronary arteries occurs in approximately 30% of patients with QAV. In our case, there were no abnormalities of coronary artery abnormalities. From the point of view of many surgeons, it is important to notice any displacement of the coronary artery mouth in order to prevent obstruction of the orifice during replacement of the valves or its plastics. In contrast to the quadricuspid valve of the pulmonary artery, there is a tendency for progression of regurgitation in patients with QAV, due to progressive fibrosis of the valves and progressive insufficient coaptation of valve flaps with age. Replacement of the aortic valve with QAV is the method of choice in patients with aortic regurgitation, there are only a few reports on the use of aortic valve reconstruction technique (tricuspidalization or bicuspidalization) with QAV. As previously noted, care must be taken to prevent coronary artery obstruction or unintentional damage during replacement or valve plastics. The risk of complete AV block is also high after a valve replacement, because the additional valve is usually located between the right and non-coronary valves, above the membranous part of the interventricular septum [*Pirundini P.A. et al., 2006*].

Conclusion

In the presented case, QAV was not diagnosed, due to the fact that not all patients in the pre-operative period undergo transesophageal echocardiography. It has been recently proved that real-time three-dimensional 3D transesophageal echocardiography is the most effective method of setting the correct diagnosis in cases of regurgitation on the aortic valve, especially in the case of QAV.

Replacement of the aortic valve with QAV is a method of choice of traditional treatment for a patient with aortic insufficiency. The immediate results are positive; however, further monitoring of the patient should be continued to assess the long-term outcome.

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