Conservative approach to mitral valve replacement in hypertrophic cardiomyopathy with systolic anterior motion – a case report

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Kardiochirurgia i Torakochirurgia Polska 2015; 12 (4): 345-347

Abstract
The authors report the case of a 60-year-old patient with hypertrophic cardiomyopathy (HCM), systolic anterior motion (SAM), and high gradient in the left ventricular outflow tract (LVOT) who underwent surgical treatment. During the surgery, myomectomy of the septum was performed using the Morrow method: despite the persisting SAM and increased LVOT gradients, the mitral valve was not replaced. The case study presents a conservative approach to mitral valve replacement during HCM surgery.

Key words: systolic anterior motion, SAM.

Introduction
Hypertrophic cardiomyopathy (HCM) is a hereditary disease of the myocardium; its first descriptions were published by Russell Brock in 1957 and Donald Tear in 1958 [1, 2]. Its definition includes left ventricular hypertrophy that is not caused by volume overload (which occurs, e.g., in the course of valvular diseases, arterial hypertension, and congenital heart defects) [3]. In its most common form, the muscle hypertrophy is asymmetric and primarily involves the interventricular septum. Left ventricular outflow tract obstruction (LVOTO) at rest is observed in approximately 25% of patients, but almost 70% of patients can be diagnosed with so-called latent LVOTO [4]. Left ventricular outflow tract obstruction is caused by systolic anterior motion (SAM) of the mitral valve and the consequent contact between the valve and the interventricular septum during systole. As a result, cardiac output decreases significantly, and left ventricular filling pressure rises; in time, progressive hypertrophy occurs and is followed by myocardial ischemia which manifests as chest pain, dyspnea, and syncope. The first symptom may also come in the form of sudden cardiac death [5].

The authors present the case of a 60-year-old patient suffering from left ventricular outflow tract obstruction with systolic anterior motion and a discussion of the problems associated with the therapeutic decisions.

Material and methods
The 60-year-old male patient diagnosed with hypertrophic cardiomyopathy was admitted to the Department of Cardiovascular Surgery and Transplantology, JP II Hospital in Krakow in order to undergo surgical treatment of LVOTO. Transthoracic echocardiography (TTE) performed before admission revealed massive hypertrophy of the basal and middle segments of the interventricular septum (up to 30 mm) as well as thickening of the remaining cardiac walls. The measured resting gradient in the left ventricular outflow tract (LVOT) was 140 mmHg; after the...
performance of a Valsalva maneuver, it rose to 180 mmHg. The patient had been suffering from anginal ailments (CCS class II) for the previous two years, and his family medical history included sudden cardiac death. Transesophageal echocardiography confirmed the presence of a very high LVOT gradient as well as abnormal motion of the anterior mitral valve cusp (SAM). Additionally, the examination demonstrated 1st degree mitral insufficiency (lateral jet). The patient was qualified for surgical treatment by the heart team.

**Results**

The procedure was performed using median sternotomy, extracorporeal circulation, and general hypothermia (28°C). The muscle of the interventricular septum was resected via aortotomy using Morrow's method: a muscle fragment approximately 2 x 1 cm in size and 5 mm in depth was excised 1.5 cm below the aortic annulus. Intraoperative TEE revealed a drop in the LVOT gradient to 90 mmHg and persisting SAM. The aorta was reopened, and an additional fragment of the septal muscle was resected (additional 1 cm in the direction of the left ventricular apex and additional 5 mm in depth), resulting in the formation of a “trough” in the ventricular septum (3 x 1 cm in size and 1 cm in depth). The subsequent intraoperative TEE revealed that the gradient decreased further (down to max. 70 mmHg), and, despite the persisting SAM, the procedure was ended. In the early postoperative period, pressor agents were discontinued, and fluid supply was increased. On the second postoperative day, transthoracic echocardiography was performed, revealing the maximum gradient through LVOT to be 50 mmHg and demonstrating that SAM was still present. The patient was discharged from the intensive care unit on the 3rd postoperative day. On the postoperative ward, treatment with β-blockers and antiarrhythmic agents was implemented. After the patient’s heart rate slowed down and stabilized, another echographic examination was performed: the maximum gradient through the left ventricular outflow tract was 17 mmHg, and the abnormal motion of the anterior mitral valve cusp (SAM) was no longer present. The patient was discharged from the clinic on the 8th postoperative day in good general condition, without complaints.

**Discussion**

The mechanism of the systolic motion of the anterior mitral valve cusp (SAM) in HCM patients results from anterior displacement of the mitral valve’s papillary muscles and the Venturi effect caused by intense blood flow through the narrow outlet of the left cardiac ventricle. Both these mechanisms result in the development of forces pulling on the mitral valve cusp in the direction of the interventricular septum [6, 7]. Additionally, anomalous papillary muscle insertion directly into the anterior mitral valve cusp can be observed in some patients [8]. The abnormal motion of the anterior mitral valve cusp is accompanied by mitral insufficiency, which is visualized by echocardiographic examination as a jet that is directed posteriorly or laterally [9]. Mitral insufficiency in HCM can also be caused by abnormalities in the internal structure of the mitral valve, as can be inferred from insufficiency jets directed centrally or anteriorly.

Approximately 5-30% of HCM patients require surgical treatment of LVOTO and SAM [10]. For over 40 years, the gold standard of surgical management has been septal myomectomy using the Morrow method; it consists in the excision of a fragment of the interventricular septum from approximately 2 cm below the aortic annulus in the direction of the left ventricular apex (approximately 3-4 cm in length and approximately 1 cm in width), thus widening the outflow tract and stopping the forces causing SAM [11]. Some authors suggest performing a more extensive resection of the septal muscle (7-8 cm) [12, 13] and partially freeing the insertion of the papillary muscles from the left ventricular wall [13]. In the case of concomitant significant mitral insufficiency associated with anomalous valve structure, it becomes necessary to repair or replace the valve. The efficacy of septal myomectomy is assessed intraoperatively using transesophageal echocardiography (TEE) in order to confirm the reduction of LVOT gradients and the lack of SAM. There are no unequivocal management guidelines for cases in which intraoperative examination reveals the persistence of SAM; the guidelines also do not specify when the decision to replace the mitral valve should be taken. In the vast majority of cases, the decision to replace the mitral valve is made intraoperatively by the surgeon, based on their experience and considering the valve anatomy. According to the literature, in most cases in which three attempts to stop SAM had failed, as confirmed by intraoperative echo, a decision was made to replace the mitral valve.

In the present case, a more conservative approach was employed. Despite the performance of two septal muscle resections and the persistence of abnormal mitral leaflet motion demonstrated by intraoperative evaluation, a decision was made not to replace the mitral valve, which was based primarily on the amount of LVOT gradient reduction. As can be surmised from this case, intraoperative echocardiography does not always reflect the actual (target) postoperative gradients, which can be properly assessed only after fluid replacement and implementation of appropriate pharmacological treatment of the underlying condition, i.e., hypertrophic cardiomyopathy. In cases in which SAM persists, mitral valve replacement can always be performed at a later time as the ultimate solution.

**Conclusions**

A conservative approach should always be considered in cases in which myomectomy must be performed in SAM patients.

The target postoperative gradients can be properly assessed only after fluid replacement and implementation of appropriate pharmacological treatment. It appears that therapeutic decisions should be based on the amount of gradient reduction during the Morrow procedure, rather than on absolute gradient values.
Disclosure
Authors report no conflict of interest.

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