THE DIAGNOSTIC AND TREATMENT CHARACTERISTICS OF A RARE MORPHOLOGICAL TYPE OF SUBAORTAL STENOSIS — DOME-SHAPED MEMBRANE

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The aim of the investigation was to analyze the features of surgical interventions, correction technique, immediate and long-term results of the treatment of a rare morphological type of subaortal stenosis — dome-shaped membrane taking into consideration differential diagnosis with other congenital anomalies causing left ventricular outflow obstruction.

Materials and Methods. Two medical cases were under study: a 20-year-old male patient and a 4-year-old girl operated in Nizhny Novgorod Specialized Cardiological Clinical Hospital. The operations were performed according to standard practice using transsternal median approach. Both patients underwent normothermic perfusion with aortic occlusion under pharmaco-hypothermic cardioplegia.

Results. Echocardiography was found to be the most informative diagnostic technique of any anatomical variant of subaortal stenosis, since it enables to make a diagnosis and determine a morphological obstruction type on the way "left ventricle-ascending aorta". When determining the indications for a surgical correction of the studied anomaly, one should carry out a differential diagnosis of a dome-shaped morphological type of subaortal membrane with characteristic deformity of the anterior mitral leaflet as one of the causes of subaortal obstruction. The surgery technique for a dome-shaped membrane consists in circular resection of fibrous mass obstructing the left ventricular outflow.

Key words: subaortal stenosis; subaortal membrane; mitral valve.

Subaortic congenital stenosis is characterized by a wide variety of anatomical variants, which differ in anatomical substrate, length, involvement of mitral and aortic valves [1–4]. These variants are represented by four basic morphological types: fibrous, fibro-muscular, muscular, dome-shaped membrane. The most uncommon is discrete subaortic stenosis represented by dome-shaped membrane (3.7% cases among all subaortic stenoses), and the most frequent is subaortic obstruction in the form of a short segment, the length of which is less than a third of aortal diameter [5]. The major treatment problems are caused by such rare forms of subaortic obstruction, as subvalvular pathology of mitral valve consisting in the presence of additional structures or fixation anomaly of chords and papillary muscles in the exit pathway of the left ventricle (mitral dome-shaped deformity) [6–8]. Such forms of subaortic stenosis are frequently associated with other intracardiac defects and can be disguised by them [9–11].

The aim of the investigation was to analyze the features of surgical interventions, correction technique, immediate and long-term results of the treatment of a rare morphological type of subaortic stenosis — dome-shaped membrane taking into consideration differential diagnosis with other congenital anomalies causing left ventricular outflow obstruction.

Materials and Methods. Over the last 10 years 2 patients: a 20-year-old man (patient L.) and a 4-year-old girl (patient T.) were operated (2005 and 2009) for congenital subaortic stenosis due to a rare type of dome-shaped membrane in the 1st Cardiac Surgery Department of Specialized Cardiological Clinical Hospital, (SCCH, Russia). Along with clinical examination, both patients underwent electrocardiography, chest roentgenography and transthoracic echocardiography. The operations were performed according to standard practice using transsternal median approach. There was used normothermic perfusion with aortic occlusion under pharmaco-hypothermic cardioplegia. Aortal valve and subaortic structures were accessed through transverse aortotomy. The patients were long-term followed up (patient L. — 6 years after the operation, and patient T. — 2 years after the operation).

Case history. In 1991, at the age of 7 patient L. was operated for aortic coarctation. He underwent indirect oartic isthmoplasty. The examination in 2002 revealed subaortic stenosis with moderately impaired circulatory dynamics. Further subaortic stenosis progression was observed. In 2005 the patient underwent a planned surgery.

A 4-year-old patient T. was first diagnosed congenital heart disease at birth. There was no circulatory

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decompensation. The child was examined in SCCH in December 2006, the diagnosis "subaortic stenosis, bicuspid aortic valve" being made. Operative therapy was not suggested due to moderate hemodynamic impairments. Further subaortic stenosis progression was observed. In 2009 she was admitted for a planned operation.

Anamnesis vitae. Patient L.: unburdened hereditary background; previous diseases: influenza, chickenpox. There were no viral hepatitis, tuberculosis, infectious diseases. No drug allergies were found. No blood transfusions were performed.

Patient T.: unburdened heredity; a child from the first physiological pregnancy, born in time, abdominal delivery (poor uterine contraction strength); birth weight — 3190 g; artificial feeding. The growth and development of the child were according to age. At the age of a year her body weight was 9000 g. Previous diseases: acute respiratory diseases. There were no viral hepatitis, tuberculosis, infectious diseases. No drug allergies were found. There were no blood transfusions.

Objective status. Patient L.: satisfactory condition, clear consciousness, active condition. Skin and visible mucosa had physiologic color. Peripheral lymph nodes were not enlarged. There was vesicular respiration, no rales. Respiration rate: 17 per minute. Rhythmic heart sounds. Heart rate (HR) was 82 beats per minute. Arterial pressure — 110/70 mm Hg. Systolic murmur was heart over the aorta. The abdomen was soft and painless. The liver was not enlarged. There was no edema. Passage of urine and bowel movement were normal. Pulsation of femoral arteries was clear.

Patient T.: satisfactory condition, clear consciousness, active condition. Skin and visible mucosa had physiologic color. Peripheral lymph nodes were not enlarged. There was vesicular respiration, no rales. Respiration rate: 25 per minute. Rhythmic heart sounds. HR was 120 beats per minute. Heart boundaries were not enlarged. There was systolic murmur, the epicenter being in the 2 intercostal space right from the breastbone. The abdomen was soft and painless. The liver was not enlarged. There was no edema. Passage of urine and bowel movement were normal. Pulsation of femoral arteries was clear.

Clinical data. Patient L. Electrocardiography: respiratory arrhythmia, HR — 55–73 per min. Levocardiogram. Left ventricular hypertrophy with systolic overload. Incomplete blockage of the anterior branch of the left crus of atrioventricular bundle. Chest roentgenography: clear pulmonary fields, the lung pattern — intense vascular component, cardiothoracic index — 50%, Moore index — 25%. Echocardioscopy: left ventricular concentric hypertrophy. Aortic valve ring diameter was 21 mm, bicuspid valve, with fibrous cusps. Eccentric subvalvular membrane of round shape with systolic pressure differential (PD) being 66 mm Hg.

Patient T. Electrocardiography: sinus rhythm, HR — 96 per min. Levocardiogram. Left ventricular hypertrophy. Chest roentgenography: moderately enlarged shadow of the heart in diameter, elongated prominence of the pulmonary artery, extended left ventricular arch, aortic configuration of the heart shadow, cardiothoracic index — 57%, Moore index — 37%. Uniformly dilated vascular fascicle, mainly due to thymus shadow. Air visible pulmonary fields, visible lung pattern, and moderately dilated vascular component. Echocardioscopy: left ventricular concentric hypertrophy in its exit pathway, 4 mm below non-coronary cusp an additional movable round-shaped mass was located bulging into systole through aortic hiatus forming turbulent flow. PD 66 mm Hg. Aortic valve cusp fibrosis, tricuspid valve. No regurgitation into exit pathway of the LV was observed.

Surgical indications. Indication for operation in both cases was subaortic obstruction with PD "the left ventricle–ascending aorta" over 50 mm Hg.

Operative technique. Both operations were performed under artificial circulation, pharmaco-hypothermic cardioplegia. Access to the heart was made — through median sternotomy; access to aortic valve and subvalvular structures — through transverse aortotomy (Fig. 1).

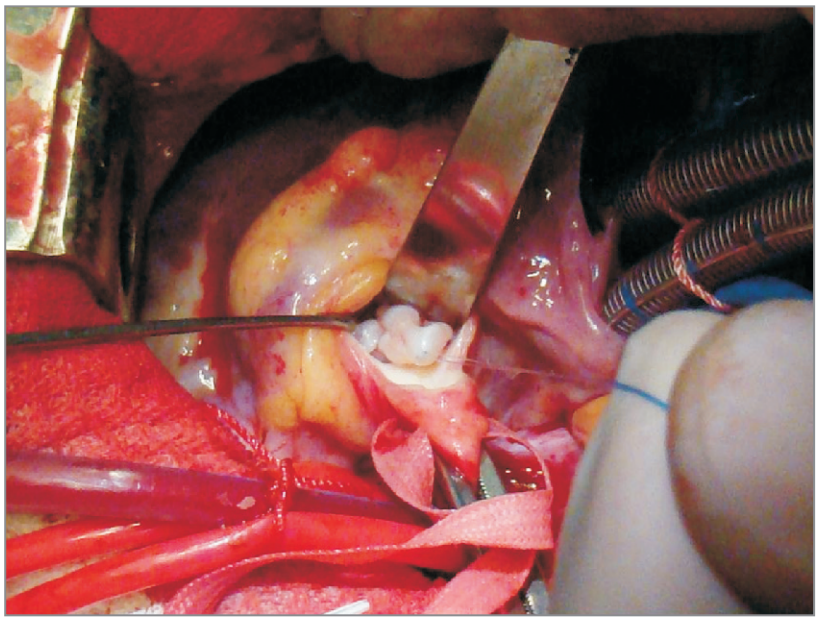

Fig. 1. Dome-shaped membrane

The technique of the surgery consisted in circular subaortic membranectomy, the membrane in those cases appearing as a thin fibrous mass, unconnected to aortic valve cusps and being like a dome-shaped bulging, looking like distorted anterior mitral leaflet (Fig. 2). It determined clear intraoperative differentiation of valvular structures and fibrous membrane, which had to be resected. Aortic cross-clamping time was 74 min in the first case and 42 min — in the second.

Results and Discussion. In general, postoperative period in both cases was uneventful. Both patients had moderate cardiovascular insufficiency arrested within two postoperative days. The patients were extubated on day 2 after the operation

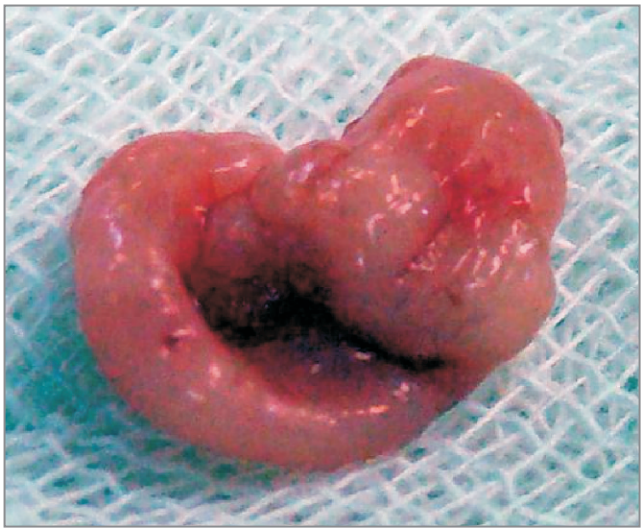

Fig. 2. Dissected membrane

and warded on day 3. On discharge, according to review echocardiography findings, residual PD at the exit pathway of the left ventricle was 10 mm Hg in patient L. and 5 mm Hg in patient T. There was no aortic valve insufficiency. Patient L. on day 7 after the operation had moderate pericardial effusions estimated as postcardiac injury syndrome manifestations. Pericarditis signs were arrested by the 10th postoperative day against the background of non-steroidal anti-inflammatory and diuretic treatment. The patients were discharged in satisfactory condition and referred to a cardiologist for outpatient supervision according to the place of residence on day 11 and 9, respectively.

Long-term results of both patients were studied: 6 years later in patient L. and 2 years — in patient T. At the moment of the last examination in July 2011 the patients aged 26 и 6 years, respectively. The result of the surgery in both cases was considered good. Physical examination revealed no events of cardiovascular insufficiency. The patients had no subjective complaints, and well tolerated a moderate exercise load. In 2011, standard diagnostic methods revealed the improvement in the form of left ventricular hypertrophy reduction (See the Table).

Systolic pressure differential of the pathway "the left ventricle–aorta" in patient L. was 21 mm Hg, in patient T. — 14 mm Hg. No aortic valve insufficiency was observed.

Summarizing, it is worth noting that the surgeries for congenital subaortic stenosis due to a rare type of dome-shaped membrane is related to a number of additional technical challenges consisting in the necessity of clear differentiation of membrane with characteristic "dome-

The dynamics of left ventricular wall thickness values, systolic/diastolic, mm

Patients	Left ventricle posterior wall, mm		Interventricular septum, mm	
	before operation	after operation	before operation	after operation
Patient L.	22/13	18/13	23/15	19/14
Patient T.	10/7	9/6	11/8	10/7

shaped" mitral valve deformity, which has a similar clinical picture of subaortic obstruction, but its correction requiring quite different surgical approach.

Conclusion. In diagnosing and determining the indications for a surgical correction of a rare subaortic stenosis — dome-shaped membrane — it is necessary to carry out a differential diagnosis with characteristic deformity of the anterior mitral leaflet as one of the causes of subaortic obstruction. The surgery technique for dome-shaped membrane consists in circular resection of a fibrous mass obstructing the left ventricular exit pathway.

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