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**THE EXPERIENCE OF SURGICAL TREATMENT OF SHONE'S ANOMALY**

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In this research the experience of operative treatment of Shone’s anomaly in the period from 2004 to 2015 is presented. It is shown that despite the need of reinterversion, the prognosis in this population is favourable.

**Key words:** *Shone’s anomaly, parachute-like mitral valve, congenital cardiac insufficiency***.**

In 1963 John D. Shone et al. first described the intricate complex of anomalies, characterized by left heart obstruction at different levels. This syndrome includes supravalvular mitral ring, parachute-like mitral valve, subaortic stenosis, and coarctation of the aorta. Shone’s anomaly is a quite rare anomaly. There may be a full form of the complex, comprising all four components, nevertheless the incomplete forms including two of three signs are also described and occur more frequently [1]. In modern literature, Shone’s anomaly is mentioned by many authors in those cases where mitral stenosis associated with one of the elements of the left heart outflow obstruction is present [2].

Understanding of mitral valve morphology is essential for determining the approach and prognosis. Pathology of mitral valve (MV) includes supravalvular mitral ring, parachute-like mitral valve, and classic congenital mitral stenosis. Supravalvular mitral ring is a crest or membrane along the line of the valve annulus. The membrane is formed at the atrial surface of mitral valve and is often fused with valve anulus, that impairs leaflet opening and results in valve stenosis. In case of parachute-like MV all the chordae are attached to a single papillary muscle. There might be two papillary muscles, but one of them would then be hypoplastic. At that, chords are, as a rule, shortened and thickened, interchordial space fusing is observed [3, 4]. As a result, infundibular obstruction is formed. In classic mitral stenosis leaflets are domed and thickened, MV aperture is narrowed drastically. Pathology of the left ventricular outflow tract may include subaortic stenosis, aortic valve stenosis [2, 5].

**Objective** is to analyse our own experience of operative treatment of patients with Shone’s anomaly.

**Materials and methods**. We have conducted a post-hoc analysis of operative treatment of patients with Shone’s anomaly over the period of eleven years. From 2004 to 2015 in State Institution “Scientific and Practical Medical Centre of Pediatric Cardiology and Cardiac Surgery of MoH of Ukraine” 29 patients with Shone’s anomaly were hospitalized, among them were 13 boys and 16 girls. In our observation there were 11 patients with a complete form of the anomaly. In most patients with incomplete form (n=18) concurrent pathology in a form of patent ductus arteriosus, or ventricular or atrial septal defect was detected. The incomplete form was presented as follows: 3 patients had coarctation of the aorta, aortic valve stenosis, MV stenosis due to valve dysplasia, interchordial space fusing; 2 patients had coarctation of the aorta and supravalvular mitral ring; 1 patient had coarctation of the aorta, MV stenosis with parachute-like MV; 1 patient had coarctation of the aorta and MV stenosis due to valve dysplasia and 4 patients had subaortic stenosis, coarctation of the aorta and classic mitral stenosis.

**Results and discussions**. At the time of the first intervention the average age of the patients was 94±21.6 days (from 1 to 180 days). 15 patients were at the age of less than 10 days. On the first stage 17 endovascular interventions were performed (15 interventions for coarctation of the aorta and 2 interventions for both coarctation of the aorta and aortic stenosis). One child’s death occurred after endovascular intervention. Cause of lethal outcome was heart failure associated with multiple congenital malformations. Surgical interventions were performed into 12 patients: aortic coarctation repair – 4 patients, repair of coarctation and stenosis of pulmonary artery – 7 patients, and one single-step correction of malformation. The single-step correction had lethal outcome due to the complications – development of a complete atrioventricular block and chylothorax, low cardiac output syndrome was observed. After repair of aortic coarctation and stenosis of pulmonary artery, the one child’s death occurred. The causes of death were pneumonia and sepsis.

Repeated surgery was performed in 23 patients. The interval between the first and the second surgeries averaged 16.6 mon. (from 2 to 69 mon.). At the time of the repeated surgery the average age was 19±16.2 mon. (from 2 to 72 mon.). It was performed 15 interventions on MV (6 repairs of supravalvular mitral ring and 9 MV plasties, during which papillotomy, commissurotomy and secondary chorde exsicion were conducted), 12 surgeries for the left ventricular outflow tract obstruction (LVOT) repair, which included 2 Ross-Konno procedures, 4 plasties of aortic valve. 27 surgeries were therefore performed in 23 patients.

The third intervention was performed in 7 patients. At the time of the third surgery the average age was 72±35.03 mon. (from 5 to 108 mon.). The interval between the second and the third interventions averaged 61.8 mon. (from 2 to 90 mon.). The reasons for the third intervention performance were 2 MV replacements, 1 procedure on pulmonary artery conduit replacement following Ross-Konno procedure, 1 endovascular intervention due to X-ray endovascular dilation of pulmonary artery stenotic conduit following Ross-Konno procedure, 3 repeated MV plasties and 3 repeated repairs of subaortic stenosis. 10 intervention were therefore conducted. One child required the fourth surgery 67 months after the last operative treatment in the age of 6 years due to the pulmonary artery conduit replacement following Ross-Konno procedure.

As can be seen from the above, the approach to surgical correction of the left-heart obstruction depended on its morphological traits. For instance, coarctation of the aorta was detected in 28 patients (96 %), in 15 patients among them was performed X-ray endovascular dilation of aortic coarctation, at that 5 (33 %) patients required the following repeated intervention on aortic arch. Surgical repair of aortic coarctation was performed in 12 (45 %) patients simultaneously with pulmonary artery narrowing in 7 of them. The latter was performed due to the ventricular septal defects. In one patient stenting of aortic arch was further performed.

Left ventricular outflow tract obstruction was documented in 26 (89 %) patients, of whom 12 (46 %) patients had subaortic stenosis, 7 (27 %) patients had obstruction at the valve level, and 7 (27 %) patients had combined valvular and subvalvular stenosis. Despite the fact that bicuspid aortic valve is not included in this pathology description, it was detected in all (100 %) the patients. Subaortic stenosis repair was performed in all 12 patients, in 2 (16 %) of whom Ross-Konno procedure was performed. For one patient, after Ross-Konno procedure, a favourable result has been achieved, while the other patient still has a mild aortic insufficiency. In remote period two (16 %) patients required repeated intervention due to LVOT obstruction and in two (16 %) patients a mild obstruction, which does not require any correction, is detected. One (8 %) patient, after LVOT obstruction repair, is followed up due to mild aortic insufficiency and one (8 %) patient – due to mild combined aortic valve failure with no obvious domination.

In four (57 %) patients with combined valvular and subvalvular stenosis aortic valve plasty was performed, meaning commissurotomy and subvalvular obstruction repair. At a later stage one (25 %) patient required repeated plasty and, further, aortic valve replacement due to pronounced failure with artificial cardiac pacemaker implantation. In three (75 %) patients a mild aortic stenosis was observed with gradient of up to maximum 35 mm Hg. Two (28 %) patients has been followed up with mild combined valvular and subvalvular stenosis without correction.

Balloon angioplasty of aortic valve was performed in 2 (28 %) patients. In remote period in one of them moderate combined aortic insufficiency with dominating stenosis.

Mitral valve anomaly took place in all 29 (100 %) patients. At that, parachute-like MV is detected in 7 (24 %) patients, subvalvular mitral ring – in 8 (28 %) patients, while in 14 (48 %) patients obliteration of interchordial spaces and MV ring hypoplasia were diagnosticated . 9 MV plasties were performed which goals were commissurotomy, papillotomy, and secondary MV chorde exsicion. In two (22 %) patients MV replacement was further performed, in one of whom re-replacement was done. Repeated plasty was performed in 2 (22 %) patients. In remote period in all the patient a favourable result is indicated. Subvalvular MV has been repaired in 6 (75 %) patients. Two (25 %) patients has been followed up in the dynamic with a mild mitral stenosis. In remote  [postoperative period](http://www.multitran.ru/c/m.exe?t=4798217_1_2&s1=%EF%EE%F1%EB%E5%EE%EF%E5%F0%E0%F6%E8%EE%ED%ED%FB%E9%20%EF%E5%F0%E8%EE%E4) 2 (34 %) patients have a mild combined mitral insufficiency, 2 (34 %) have mild mitral restenosis and 1 (32 %) patients has been followed up with a moderate restenosis. In two (25 %) patients residual mild combined mitral insufficiency has been detected.

In one (3 %) child with Shone’s anomaly a single-stage definitive repair at the age of 7 days was performed with lethal outcome. The surgery was complicated by complete atrioventricular block and chylotorax, low cardiac output syndrome was observed. One another child’s death occurred after repair of aortic coarctation and stenosis of pulmonary artery. The causes of death were pneumonia and sepsis. The third child’s death occurred after balloon dilation of critical aortic stenosis and aortic coarctation. The child had multiple congenital malformations. As can be seen from the above, three children has died in total in early postoperative period, lethality is 10 %. There have been no lethal outcomes in remote period.

**Conclusions:**

Shone’s anomaly is a complicated congenial left heart pathology, with is associated with unfavourable natural course and variety of surgical correction methods. The approach of surgical treatment in our clinic is a staged repair of critical obstruction. This approach is supplemented by repeated reinterventions. Nevertheless, the long-term survival up to 7 years is 90 %. For wide implementation of this approach in a clinical practice, more observations and a long-term follow-up in remote period are required.

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