**Experience of Hypoplastic Left Heart Syndrome Surgical Treatment**

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The basis of observation is experience and theoretical argumentation of surgical treatment of hypoplastic left heart syndrome (HLHS). Hypoplastic left heart can be varied in anatomical characteristics depending on the underdevelopment of certain morphological structures of the left heart. This pathology occurs with a frequency 4% to 8% of all congenital heart disease. Annualy 200 children are born with such disease in Ukraine. If not prevented physiological closure of open ductus arteriosus by prostaglandin infusion, develop cardiogenic shock and child dies. Curative treatment is phased surgical correction.

**Key words:** congenital disease, hypoplastic left heart syndrome, Norwood I procedure.

Hypoplastic Left Heart Syndrome (HLHS ) is one of the most difficult congenital heart disease and Great Vessels, and can be divided into four categories independing anatomical characteristics of aortic valve and mitral valve:

* HLHS with Aortic and Mitral Stenosis
* HLHS with Aortic and Mitral Atresia
* HLHS with Aortic Atresia and Mitral Stenosis
* HLHS with Aortic Stenosis and Mitral Atresia

Hypoplastic Left Heart Hemodynamic is because oxygenated blood, that returns to the left atrium from the lungs, can not pass to hypoplastic left ventricle. Instead, Blood through the atrial septal enters into the right heart, where mixes with deoxygenated blood. Mixed blood goes from the right ventricle and through the pulmonary artery enters into the lungs, and also through patent ductus arteriosus enters into Systemic Circulation, which accept blood only through Right-Left Shunt. That is why the prognosis for life at birth depends on the preservation of patent ductus arteriosus.

**Abstract:** theoretically justify surgical treatment of hypoplastic left heart syndrome.

**Materials and Methods.** During the period from July 2010 to April 2014 in Amosov National Institute of Cardiovascular Surgery NAMS of Ukraine were operated 49 patients with HLHS. Patients were 32 males (65,3%) and 17 females (34,7%). Middle patient’s age was 8±2,2 days (from 6 to 11 days). Average patient’s weight was 3,5±0,6 kg (from 2,4 to 4 kg). 30 patients (61,2%) have congenital disease which was diagnosed prenatal and confirmed at birth by echocardiography. Preoperative preparation was performed 7-10 days and was directed at balancing systemic and pulmonary vascular resistance in compliance with blood oxygen saturation within 80%. In necessary, conducted correction of metabolic acidosis and administered inotropic drugs.

Recognized and accepted treatment is currently the phased method, created by W. Norwood.

Norwood procedure provides:

* Creation of a free and continuous access to the systemic circulation
* Ensure stabile and controlled source of pulmonary blood circulation
* Creation of wide connection on the atrial level
* Adequate and unobstructed coronary circulation.

Modified Norwood procedure first proposed japanese cardiac surgeon *Shunji Sano.*

In our institute was developed own algorithm, which allow to choose the optimal way to surgery for patients with HLHS.

In 28 (57.1%) patients in the first stage was used Norwood procedure. 14 patients (28,6%) were operated the hybrid operation, which included an open ductus arteriosus stenting and bilateral narrowing of the pulmonary arteries. In 9 (32.1%) cases surgery was performed Norwood I procedure in modified Sano and in 19 (67.8%) cases used technique of R. Mee using shunt Blalock-Taussig ( 3,5mm). Surgical treatment was performed using Heart-Lung Machine (Bypass).

Protection of myocardium is antegrade cold crystalloid cardioplegia. During the main stage applies circulatory arrest and deep hypothermia (child body temperature decreases to 16 ° C), with a complete circulatory arrest should not exceed 40 - 45 minutes to prevent irreversible changes in the central nervous system. For effective hemostasis using recombinant factor VIIa clotting. In the postoperative period all patients had peritoneal dialysis. Take into account, that 2 patients two patients in the presence of left ventricular hypoplasia, of the aortic and mitral valves, ascending aorta and aortic arch were developed normally, was undergone Damus-Kay-Stensеl operation. Five patients underwent reconstruction of the aortic arch with bypass and antegrade cerebral perfusion. In this group of patients left ventricle, aortic and mitral valves developed normally, but was hypoplastic aortic arch. Of the five patients in the early postoperative period died 1 patient (20.0%), 4 patients (80,0%) was discharged to outpatient treatment in satisfactory condition.

**Results.** Of the 28 patients who had Norwood I procedure survived 6 patients (21.4%). The second stage correction of defects (bidirectional cavopulmonary anastomosis) spent 5 patients, of which 4 patients (80,0%) have a favorable outcome. The average duration of bypass during Norwood I procedure was 230 ± 43 minutes, duration of circulatory arrest 39 ± 8 minutes. Practice has shown, for successful treatment HLHS required professionalism, coordinated work at all stages, from maternity home (obstetrician, neonatologist), transportation in cardiac surgery department (brigade reanimobile), preoperative preparation (resuscitationist), surgery (surgeon, anesthesiologist, perfusionist), postoperative treatment and care (resuscitators, nurses).

The organization of screening pregnant women in optimal gestational age (up to 22 weeks, according to the order of Ministry of Health of Ukraine) for further management planning of pregnancy (interruption or prolongation). Analysis and evaluation of phasing of clinical and instrumental data allows to set correct diagnosis. Accurate differential diagnosis of defects should be conducted in a specialized department using echocardiography and other techniques, immediately after birth.

All newborns should be immediately hospitalized in the Neonatal Intensive Care Unit (NICU). To ensure:

* Central venous access;
* Infusion of prostaglandin E1 (PGE1; initial dose 0,05-0,1 mcg / kg / min intravenous);
* Tracheal intubation and begin artificial pulmonary ventilation, in necessary;
* Correction of metabolic acidosis;
* Newborn in critical condition with cardiogenic shock may require the appointment of inotropic drugs and diuretics to improve heart function and control blood volume.

Further newborns need gradual correction, after which the right ventricle begins to function as a systemic.

Correction of HLHS performed in 3 stages:

* Norwood procedure performed in firsts 14 days;

Pulmonary trunk separated, sealed distal part by autopericardial patch and ligature patent ductus arteriosus. Then make right shunt by Blalock-Taussig or forming a channel between the right ventricle and pulmonary artery (modified Sano). Atrial septal defect increase – atrioseptectomy, and proximal pulmonary artery and hypoplastic aorta connected with allograft aortic or pulmonary artery to form a new aorta (neoaorta).

* Bidirectional cavopulmonary anastomosis in age 4-6 months (Norwood II);
* Total cavopulmonary anastomosis in age after 2 years (Norwood III).

In some centers, heart transplantation is the method of treatment; at the same time prostaglandin E1 infusion should continue until will not be know about the availability of donor hearts. Also is limited availability of donor hearts; approximately 20% of newborns die waiting for a donor heart. 5-year survival after heart transplantation and correction after multistage about the same. After heart transplantation should be used Immunosuppressants. These medications make patients more susceptible to infections and over 5 years of cause pathological changes in coronary artery graft in over 50% of patients. The only known way to treat coronary artery disease is the re-transplant graft.

**Discussion.** Three-stage treatment became the basis for surgical treatment HLHS. Complications after Norwood procedure are common, carry a significant risk of mortality, they are associated with features of preoperative preparation of patients and type of hypoplastic left heart. Implementation of these operations requires careful preparation and coordinated work of all services that provide diagnostic, anesthesia, surgery and resuscitation stages of work. In addition, the importance of concentration of patients with this pathology in one center, which as experience is gained steadily improved outcomes.

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