UDK 616.132-007.271:616.832.21]-089

TREATMENT EXPERIENCE OF CRITICAL CONGENITAL HEART DISEASES AND CRITICAL CONDITIONS CAUSED BY THEM AS ELEMENT OF URGENT OR EMERGENCY CARDIOSURGERY

Siromaha S.O., Rudenko K.V., Zalevskyi V.P., Truba Ya.P., Prokopovych L. M., Fylypchuk V.V., Lazoryshynez V.V., Knyshov G.V.

M.M. Amosov National institute of Cardiovascular Surgery of the National Academy of Medical Sciences of Ukraine, Kyiv

We present generalized treatment experience of 152 patients with critical congenital heart diseases and critical conditions in children in the Institute for the last 2 years (2012-2013).

***Key words**: critical congenital heart defects, critical conditions, urgent cardiosurgery.*

Congenital heart diseases (CHD) occupy the 3rd-4th place among all developmental pathologies, after congenital locomotor system and CNS pathologies. CHDs frequency composes from 8 to 12 cases for 1000 live-birth infants. During early neonatal period without surgical intervention die 35-50% of children with complex CHD, up to 1 month of life – 51.7%, and up to 1 year – 91% of children. As part of children mortality structure CHD makes for 8.2 to 16.6% in various years.

Critical CHDs – are heart development abnormalities that do not allow blood ejection with sufficient pressure to sustain life and oxygen saturation; in urgent surgery absence they lead to patient death during the first days of life. Critical conditions of patients with congenital cardiovascular diseases are caused by the following principal pathological syndromes: hypoxemia, cardiac

insufficiency, life-threatening heart rhythm disturbances, or their combination. Arterial hypoxemia syndrome may manifest already during the first hours after a child birth. As the rule, cyanosis has central character, and is present also on mucous membranes. Arterial hypoxemia in CHD is associated with right-left venous blood shunting and its influx into aorta; decreased pulmonary blood flow and decreased return of arterial blood to heart; and systemic circulation and pulmonary circulation division. In order to characterize arterial hypoxemia its level should be assessed (for the majority of CHDs hypoxemia is diagnosed as pO2 level less than 80 mm Hg, with blood oxygen saturation less than 85%); its severity (acidosis degree) should be measured, as well as vital organs secondary dysfunction, because all of the above determines therapeutic measures complex. Another wide-spread and threatening syndrome in CHD infants – cardiac insufficiency (CI) – is the condition, when heart can not secure circulation adequate for body needs. Principal cardiac insufficiency symptoms during neonatal period are: tachycardia, apnoea, congestion signs in pulmonary circulation (lung rattlings) or systemic circulation (liver increase, oedemas); in other words, cardiac insufficiency rapidly develops congestive character. All this is accompanied by problems during child feeding (poor sucking, apnoea during feeding, etc) [1].

Critical CHDs compose 1.2 – 2 cases for every 1000 live births. Thus, according to our calculations each year in Ukraine 750-800 children with CHDs are born. Notwithstanding almost 100% pregnancies coverage with Echo-CG (96.16%), prenatal diagnosis rate remains low [2]. The same situation is observed in other countries. For example, in the USA 97-99% of pregnant undergoes EchoCG; however, the rate of prenatal CHD diagnosis is only 28-68.3% [3-5]. Thus, only in California state each year die up to 30 new-borns with undiagnosed CHD both during pre- and post-natal period. As the rule these are children with hypoplastic left-heart syndrome, or coarctation of aorta [6].

Patients with critical aortal stenosis, critical coarctation of aorta, completely interrupted aortic arch, pulmonary artery critical stenosis, pulmonary

atresia, transposition of great arteries with intact IVS (intra-ventricular septum), total anomalous pulmonary veins drainage, hypoplastic left-heart syndrome, severe forms of Ebstein disease, and common truncus are included in tehe group of article. [7]

Publication goal was to study treatment experience of critical CHD patients and improvement of surgical correction immediate results in such patients.

Materials for of the study were critical CHD patients, who received treatment in M.M.Amosov National institute of Cardiovascular Surgery of the National Academy of Medical Sciences of Ukraine in 2012-2013. Apart from that, the study group included patients, who were in critical condition during hospital admission – children with congenital complete atrioventricular block, and infective endocarditis (Fig.1). The criteria for urgent or emergency surgical intervention in patients with congenital AV block were the following: heart rate resistance during exercise (less than 60 beats per minute), substitutive ventricular arrhythmia, and fixated «pauses» longer than 5 sec. during daily ECG monitoring, intervals ratio RR≥3 pp. Also as the above patients cohort may be classified patients with iatrogenic diseases of heart and magisterial vessels (iatrogenic complete AV block, heart chambers perforation during intervention procedures, heart tamponade after surgeries, etc).

Such patients demand urgent (during the first 24 hours after admission to specialized in-patient clinic) or emergency cardiosurgical help (such help should be provided as soon as possible after patient admission or after clinical diagnosis establishment, as a rule during the first few hours). Critical patients group was 10.3% of general patients number (n=1473), who received help in 2012-2013.

Table 1. General patients characteristics (n=152).

CHD	**Number**	**Up to 1 month of life**	**Up to 1 year of life**	**Older than 1 year**
Hypoplastic left-heart syndrome	29	29	-	-
Transposition of great arteries (TGA)	20	19	1	-
Anomalous pulmonary veins drainage	4	2	2	-
Critical coarctation of aorta	20	15	3	2

Critical pulmonary artery stenosis	16	6	5	5
Critical aortic stenosis	11	4	2	5
Interrupted aortic arch	12	12	-	-
Infective endocarditis	2			2
Congenital AV block	20	1	3	16
Common truncus	1		1	
Hypoplastic left-heart syndrome	5	4	1	
Iatrogenic complete AV block	4	-	-	4
Hypertrophic cardiomyopathy, exudative pericarditis	1	1	-	-
Fallot tetralogy, with esophagoplasty	1	1	-	-
High pulmonary artery hypertension 100%	4	1	3	-
Complete A-V communication	**2**	1	1	-
Total	152	96	22	34

In the study group both male (n=86), and female (n=66) patients were included. Patients' age varied from 1 day to 17 years; average age was 9.6 years.

Surgery group included 125 patients, who received 138 interventions and surgical procedures (Figure 2).

Table 2. Surgical procedures and interventions in 125 patients.

	Endovascular n =40	Palliative n=11	Radical n=55	Hybrid surgeries n= 9	Artificial pacemakers implantations n= 23
Hypoplastic left-heart syndrome (n=24)	4		11	9	
Transposition of great arteries (TGA) (n=18)	11	1	12		

Anomalous pulmonary veins drainage (n=4)			4		
Critical coarctation of aorta (n=18)	5		17		
Critical pulmonary artery stenosis, pulmonary atresia (n=15)	9	7	2		
Critical aortic stenosis (n=10)	9		1		
Interrupted aortic arch (n=5)			5		
Hypoplastic left-heart syndrome (n=5)	2	3			
Infective endocarditis (n=2)			2		
AV- block (n=23)					23
Common truncus (n=1)			1		

Intervention procedures included Rachkind procedure (n=17), pulmonary artery valve dilatation (n=9), aortal valve dilatation (n=9), dilatation of aortal coarctation zone (n=5), and aortal coarctation stenting (n=2). Palliative interventions included modified Blelok anastomosis (n=9), pulmonary artery narrowing (n=2), and atrial septal fenestration (n=1).

27 patients did not receive interventions or surgeries due to their very severe condition, caused by late hospital admission or accompanying diseases, which were contraindications to surgery. (Figure 3).

Table 3. Non-operated patients (n=27)

Diagnosis	Number	From 0 to 1 months	From 1 month to 1 year	Older than 1 year	Average weight, kg
Hypoplastic left-heart syndrome	5	5			3.42
Transposition of great arteries (TGA)	2	1	1		3.05
High pulmonary artery hypertension 100%	4	1	3		1.750
Critical coarctation of aorta	2			2	18.50
Hypertrophic cardiomyopathy	1	1			2.70
Aortal stenosis	1	1			3.50
Aortal arch interruption	6	6			1.95
Complete A-V комн.	2	1	1		3.60
Congenital AV block	1	1			2.49
Fallot tetralogy, with esophagoplasty	1	1			3.00
Hypoplastic right-heart syndrome	1	1			3.00
Pulmonary atresia –I type	1	1			3.20
Total	27	20	5	2	1.86

Time from patient admission (or after final diagnosis establishing) till intervention depended completely on patient condition on admission, need for additional examinations and complex measures, aimed at patient condition stabilization, and it was on average 1.4 days.

Results. Hospital mortality in studied group was 33.5% (n=51). Mortality in non-operated patients group was 92.6% (n=25). Post-operative mortality among patients with critical CHDs was 20.8% (n=26). The highest post-operative mortality presented the group of hypoplastic left-heart syndrome (66%,

n=19). The mortality causes were the following: brain oedema, anastomosis dysfunction, stent thrombosis, acute cardiac insufficiency, respiratory insufficiency, intra-operative bleeding, sepsis, tracheal mucosal detachment, pulmonary artery thromboembolia (PATE), and pulmonary vessels sclerosis.

Non-lethal post-operative complications were noted in 18 patients, and were the following: cardiac insufficiency, sepsis, respiratory insufficiency, lymphorrhea, myocardial insufficiency, phrenic nerve paralysis, and encephalopathy.

At the same time during the period mentioned in M.M.Amosov National institute of Cardiovascular Surgery 1321 patients with CHD were treated, who were not in critical condition, with general hospital mortality 2.6% (n=35). This patient category (n=964) received 1108 surgeries with post-operative mortality 0.8% (n=8).

Discussion. Among CHD patients there is large number of persons, demanding urgent or emergency cardiosurgical help. Such help postponement leads to irreversible changes, which may result in death or severe disability. As the rule, CHD causes ductus-dependent defects, or defects with pronounced obstruction of outflow tracts from left (right) heart chambers, and decompensated congenital abnormalities of cardiac conduction system. In addition, these are patients with acquired critical conditions due to cardiac insufficiency progression, and iatrogenic life-threatening heart and magisterial vessels disturbances. Quality medical service to such patients depends on many factors, the principal factor is timely intervention or surgery. Other factors are less critical, however, they play important role in reaching of principal goal – new-born life and health preservation on the background of life-threatening condition. Such factors are – quality and timeliness of prenatal diagnostics, patient transporting to cardiosurgical clinic, agreed work of medical personnel, multidisciplinary team (primary care clinician, gynaecologist, neonatologist, transportation brigade, and medical collective of specialized cardiosurgical in-patient clinic). This is because specialists from Amosov National institute of

Cardiovascular Surgery during the last 3 years actively collaborate with primary care colleagues, specialists from Institute of Paediatrics, Obstetrics and Gynaecology of National Academy of Medical Sciences of Ukraine aiming at better CHD prenatal diagnostics, timely delivery of such patients to the Institute and providing them with whole range of treatment procedures.

Providing with medical help of critical CHD patients, or children in critical condition is integral part of urgent or emergency cardiosurgical care, which during the recent several years is actively developed in M.M.Amosov National institute of Cardiovascular Surgery and in other cardiosurgical centres, which are part of Association of Cardiovascular Surgeons of Ukraine.

Conclusions:

1. During the recent years in Ukraine is formed system for urgent and emergency cardiovascular care, which allows significantly increasing medical care quality to patients with critical heart defects through standardizations of "patient routes", and scope of diagnostic and treatment procedures.
2. Surgical treatment of children with critical CHD or conditions is associated with high post-operation mortality (20.8%), which considerably differs from the results of surgical treatment for planned patients (0.8%).
3. Hospital mortality among non-operated patients with critical CHDs is 92.6%.
4. The basic constituents for successful surgical correction and good late fate are – timely and professional work of multi-disciplinary specialist team, as well as development of algorithms and protocols of surgical help to patients with critical CHDs.

Literature

1. *Руденко Н.М.* Лікувальна тактика при критичних вроджених вадах серця у немовлят // Хірургія дитячого віку 2012; 3: 12-18.
2. Стан здоров'я жіночого населення в Україні за 2012 рік, МОЗ, «Центр медичної статистики МОЗ України», Київ, 2012.

3. *Friedberg M K et al* Prenatal detection of congenital heart disease // J Pediatr., 2009;155(1).
4. *Priya Jegatheesan, MD et al* Oxygen Saturation Nomogram in Newborns Screenedfor Critical Congenital Heart Disease // PEDIATRICS, 2013, Volume 131, Number 6.
5. *Landis BJ et al.* Prenatal diagnosis of congenital heart disease and birth outcomes // PediatrCardiol. 2013 Mar;34(3).
6. *Chang RK, Gurvitz M, Rodriguez S.A.* Missed diagnosis of critical congenital heart disease// Arch PediatrAdolesc Med. 2008 Oct;162(10):969-74.
7. *Talner CN* Report of the New England Regional Infant Cardiac Program, by Donald C. Fyler, MD, Pediatrics, 1980;65(suppl):375-461 //Pediatrics.1998;102(pt 2):258-259.

ДОСВІД ЛІКУВАННЯ КРИТИЧНИХ ВРОДЖЕНИХ ВАД СЕРЦЯ ТА СПРИЧИНЕНИХ НИМИ КРИТИЧНИХ СТАНІВ ЯК ЕЛЕМЕНТ НЕВІДКЛАДНОЇ АБО ЕКСТРЕНОЇ КАРДІОХІРУРГІЇ

Сіромаха С.О., Руденко К.В., Залевський В.П., Труба Я.П.,
Прокопович Л.М., Филипчук В.В., Лазоришинець В.В., Книшов Г.В.

Узагальнено досвід лікування в НІССХ ім. М.М. Амосова НАМН 152 пацієнтів із критичними вродженими вадами серця та критичними станами у дітей за останні два роки (2012–2013 рр.).

Ключові слова: *критичні вроджені вади серця, критичні стани, екстрена кардіохірургія*

ОПЫТ ЛЕЧЕНИЯ КРИТИЧЕСКИХ ВРОЖДЕННЫХ ПОРОКОВ СЕРДЦА И ВЫЗВАННЫХ ИМИ КРИТИЧЕСКИХ СОСТОЯНИЙ КАК ЭЛЕМЕНТ НЕОТЛОЖНОЙ ИЛИ ЭКСТРЕННОЙ КАРДИОХИРУРГИИ

Сиромаха С.О., Руденко К.В., Залевский В.П., Труба Я.П.,
Прокопович Л.М., Филипчук В.В., Лазоришинец В.В., Кнышов Г.В.

Обобщен опыт лечения в НИССХ им. Н.М. Амосова НАМН 152 пациентов с критическими врожденными пороками сердца и критическими состояниями у детей за последние два года (2012–2013 гг.).

Ключевые слова: *критические врожденные пороки сердца, критические состояния, экстренная кардиохирургия*