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Epidemiology and Dynamics of Congenital Heart Diseases at the Newborns in the Zhambyl Region, Kazakhstan

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Abstract: An study is considered the analysis of frequency of Congenital Heart Diseases (CHD) at the newborns that were on treatment in the regional perinatal center of the Zhambyl Region for 2 years (2014-2015). Frequency of all heart diseases on the average has 9-1000, counting on children population. In the structure of pathology there are prevailed: ventricular septal defect, atrial septal defect, patent ductus arteriosus and pulmonary artery stenosis. By last years the quantity of the non correlated valvular defects are increased considerably. It is important to take into account this circumstance in clinical practice. There are reflected the features of the natural history of congenital heart diseases with a high lethality at children, especially in the 1st year of life.

Key words: Congenital heart diseases, newborns, prevalence, structure, defect

INTRODUCTION

Epidemiology of Congenital Heart Diseases (CHD): Prevalence of CHD at newborns in the different countries of the world including in Kazakhstan.

Basis strategy of the cardiac surgery help to patients with the Congenital Heart Diseases (CHD) is studying of a prevalence and structure of this pathology. According to the opinion of foreign researchers (Knowles *et al*., 2005) and Russian researchers (Bokeria *et al*., 2007a), there is observed the tendency to increase of frequency and detectability of CHD in the developed countries. Frequency of CHD detection is varied over a wide range 2. 4-4.15 on 1000 of live-born (Sharykin, 2009). According to EUROCAT institute, average population prevalence of CHD in countries of Western Europe is averaged as 8.0 on 1000 children's population (Dolk *et al*., 2011). According to statistical WHO data, annually there are born in the countries of the world about 5-6% of children with malformations, at the same time in half of cases, it is the lethal and severe heart diseases, requiring serious surgical correction (Lazareva, 2007).

According to clinic-epidemiological research of the Canadian researchers, the population prevalence of CHD in Quebec (Canada) was made 11.89 on 1000 children's population (Marelli *et al*., 2007). Congenital malformations come out on top in structure of perinatal and infant mortality in the countries of Europe and North America (Zhuchenko *et al*., 2006). Annually about 30-35 thousand children with congenital heart disease are born in the USA.

CHD and hereditary diseases win first place in structure of the reasons of infant mortality in the countries with the high level of medical care at low indicators of infant mortality (6.7-8.5%) and it is not due to true increase of their frequency and in connection with decline in mortality from other pathology (Barashnev *et al*., 2004).

According to the international data, 40% of children of total need expeditious surgical correction of heart disease within the 1st year of life. Mortality is presented about 29% of children at the natural history of congenital heart disease by the end of the 1st week and by the end of the 1st month of 42%, 1st year of 87% (Bankl, 2007).

Annually, there are born about 10,000 children with CHD in Russia from which 30-50% with critical heart diseases at the neonatal period. As a rule, failure to assist by the specialized medical care at such conditions during the 1st hours, days or weeks of life was led to a lethal outcome. The lethality at such CHD is extremely high: 29% of newborns are died by the end of the 1st week, 42% by the 1st month, 87% of children by the 1st year of

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life. Meanwhile, 98% of children with CHD, surgically operated in early periods after the birth, live full-fledged life (Grishina and Rakh, 2010).

In Kazakhstan, the structure of CHD child mortality has one of the first places. Every year the percent of detectability and birth rate of children with this pathology are increased. In the Republic annually about 3000 children with CHD are born from them 80% die before they reach 1 year in the 1st weeks of life to 20% in the 1st month to 27%. In the age structure of mortality from congenital heart anomalies and the great vessels are made the children of the first year of life (91%), among them more than a half are made the children of the neonatal period (the first 28 days of life).

Indicators of mortality and disability in connection with various congenital malformations of heart: Recession of decrease in indicators of infant mortality, growth of indicators of child disability, high level of prevalence of congenital heart diseases and unsatisfactory efficiency of delivery of qualified medical care for children at such conditions put questions of improvement of system organization of the qualified medical care in a rank of urgent problems of the state and society. Despite ongoing efforts, growth of detectability of congenital heart diseases is registered.

According to literature, CHD are the reasons, leading to an early and heavy disability and define development of the disability approximately in 7 children from 1000 (Zhilyuk *et al*., 2011). It was revealed in other researches that prevalence of child disability because of CHD was made 3.5 for 10,000 of child population; the analysis of structure of CHD among handicapped children was shown approximately equal shares of isolated (48.6%), combined (46.3%) and the smaller number associated (5.1%) of CHD.

Among CHD the considerable share is made by Ventricular Septal Defect (VSD) 22.9%, Tetralogy of Fallot 14.2% and Atrial Septal Defect (ASD) 10.6%. The greatest share of handicapped children is the share of age groups 3-6 and 11-14 years (32.9 and 26.1%, respectively). CHD is diagnosed at half of handicapped children in the neonatal period 46.06% at every tenth who are elder than 3 years 11.0%. Surgical treatment was received by 56.2% of handicapped children (Chernova, 2005). It is grown the level of primary disablement in connection with CHD at children of early age (Albitsky *et al*., 2010).

In general and owing to congenital heart diseases in particular, the sufficient number of scientific works is devoted to studying of a problem of child disability. At the same time as there were shown the observations, the list of CHD obligatory registration is not quite reflected the true prevalence of these or those nosological forms that leads to underestimation of the separate CHD (Askarova *et al*., 2014). Now the special importance is gained by questions of studying of regional prevalence and medical and social aspects of disability of children with cardiovascular pathology which have to be the cornerstone of planning of priority for this territorial subject of the federation on the actions for improvement of the health status of the pediatric population. At the same time in regions it is insufficiently carried out the researches devoted to search of ways of optimization of medical care to handicapped children (Zielinskaya and Ter, 2008).

Fetus CHD is remained an actual problem of perinatology, taking the second place in the structure of perinatal and infant mortality. Among the reasons of neonatal incidence there are made about 30% of the share of CHD and chromosomal abnormalities.

It is known that at 40-50% of children with congenital malformations there can be kept a life at early diagnosis and surgical correction of CHD at fetus and the newborn during the first hours of life.

It is carried out the fetus echocardiography to all women from the 18th-22nd week of pregnancy for an exception of cardiac pathology in the USA and the European countries (Norway, France, Germany, etc.). Such approach was promoted that in Western Europe the level of prenatal diagnostics of CHD at a dispensary stage reaches 48%. Unfortunately, prenatal diagnostics in our country is not developed practically, especially in remote regions.

Fetus heart structure can be identified intravaginal on the 11-14th week of a gestation. Despite some disagreements of rather optimum term of studying of fetus heart (in France the 20-22nd week in Norway the 18th in Germany and the USA the 18-20th week of a gestation), researchers in the most countries were come to the conclusion that it is optimum to test cardiovascular system of the fetus on the 18-20th week of a gestation at the screening level.

During intrauterine growth, the frequency of the heart defects is higher as there are considered the fetuses which were aborted. The CHD of cardiovascular system are connected with changes at an embryonal development, the deviation from normal development at an early stage of an embryogenesis. The atypical forms of cardiovascular circulation, arising because of anatomical defect can be significantly influenced structural and functional development of a fetus. Besides, life after the delivery is had a noticeable impact on the clinical manifestations of the concrete isolated defects, so congenital defects of cardiovascular system are the most

frequent reason of deaths among children which are also considered as one of the main problems of public health around the world (Bernier *et al*., 2010).

Modern cardiac surgery and early diagnosis and correction of CHD: Early diagnosis of CHD at a fetus or during the asymptomatic period immediately after birth will reduce risk of clinical deterioration with the timely beginning of the appropriate treatment of a disease. Technical progress in diagnostics within the last two decades, especially in experienced hands was helped to the experts to detect the CHD. Screening of the cardiovascular system of fetus, the fetus echocardiography, for the purpose of early identification of CHD and other alternative methods of diagnostics are allowed to make the choice about an abortion in case of difficult defects.

Diagnostics of fetus CHD helps to reveal a great number of the combined noncardiac and chromosomal abnormality (Verspyck *et al*., 1999). It is more and more leading clinics as well as abroad, consider fetal echocardiography as an integral part of a neonatology, child cardiology and cardiosurgery. Numerous foreign publications demonstrate that the method of complex echocardiography allows to diagnose precisely up to 97% of CHD at a fetus. In foreign clinics there are developed the surgical interventions in the antenatal period that is the future of a cardiosurgery (Graf *et al*., 2000). CHD detection before the birth of the child gives the chance and time for the organization of obstetrics in specialized maternity hospitals and transportations to the cardiac surgery clinic. Introduction in work of cardiological clinic of a telemedicine expands the contingent of the pregnant women who need such help. Especially, it is important for children with "critical valvular defects" of the period of newborns. If these children are born in the regions where there is no cardiac surgery clinic, they die during the first days and even hours of life.

Prenatal diagnostics of fetus CHD plays a large role in the organization of conducting pregnancy as it is detected the forecast for pregnancy prolongation, the birthplace of the child, a possibility of surgical correction. At detection of surgical pathology of a fetus, further tactics needs to be defined together with the surgeon neonatologist (Nemilova, 1998).

Now in the world practice the number of early operations at newborns is made from 40-90%. Delivery of health care on the cardiac surgery to newborns and children of the first year of life with CHD in the countries of Europe and USA has the beginning from 60th years of the last century in Russia from 80th years. Timely diagnostics and surgical treatment at early age allow keeping lives of 1000 children with CHD that not only positively influences a demographic situation in the republic but also allows reducing costs of a social and medical support of this category of patients. Thanks to the timely taken by the Government of the Republic of Kazakhstan of the sectoral programs on development of cardiology and cardiac surgery care for 2007-2009 there is created the effective cardiological and cardiac surgery service in the country. It is increased the bedspace and there are improved the material and technical resources of the organizations of health care, giving cardiac surgery care to children.

Experience of the world and domestic practice is shown that for development and realization of such task it is necessary the creation of the adequate medical and organizational and methodological base, providing successful functioning of the treatment quality control system (Little and Whipple, 1996). There is a wish to emphasize as well that the international experience of introduction of industrial methods of the organization of medical process is shown substantial increase of efficiency of activity of clinic and treatment results without significant increase in financial expenses (Habib *et al*., 1997).

There is no doubt that the relevance and demand of the cardiac surgery care for children has primary priority. The problem of decrease in a lethality and improvement of the forecast of newborns life with heart and vascular malformation was always the most actual and difficult solved task in a child cardiosurgery and it remains so today (Bokeria *et al*., 2007b).

Problems of the organization of the cardiac surgery help to newborns and children of the first year of life with heart malformations: Important problem of the present stage of development of child cardiology and child cardiosurgery is the organization of medical care at congenital malformations of the cardiovascular system. Today there are no accurate criteria between norm, pathology and congenital features of a structure of cardiovascular system.

Today around the world the surgery of congenital heart diseases and blood vessels achieves gradually more and more favorable results: modern medical technologies allow to provide medical assistance at diseases which seemed absolutely inaccessible 10 years ago, there are increased the number of the patients who had timely cardiac surgery care and thanks to it, considerably survived "critical age of the natural history". In general on the Republic it is noted increase in number of cardiosurgery clinics and it is made specialized medical care more available. Besides, the maintenance of

cardiosurgery clinics is very expensive action for any state. Therefore, it is impossible to rely only on intuition at the solution of the problems, demanding huge expenses and including a large number of participants of the process from whom depends on the ultimate result. For the organization of optimum medical care it is necessary to use the formalized technologies of decision-making (the system analysis, algorithms of actions). The concept of the organization of optimum medical care had been formulated by WHO experts: "it is obligatory carrying out of all actions (according to standards) which are safe and accepted in terms of the spent means in this society and their influence on mortality, incidence, disability". Standards according to Donabedian triad are divided into 3 groups: the standard on a resource, the standard on process, the standard on result (Nazarenko and Polubentseva, 2000). In order to define the standard on a resource, it is necessary to know about need for different types of cardiac surgery interventions. For detection of need, it is necessary to have information about birth rate, prevalence of each type of CHD, expediency and a possibility of surgical correction.

It has been noted that at stratification of the research results on prevalence of CHD on the basis of the dagree of influence of the revealed anatomic anomalies of a formation of heart on haemodynamics, the prevalence of heavy CHD (Group 1) and CHD with average degree of intensity of haemodynamics changes (Group 2), according to different researches, practically didn't differ. Significant differences in researches were revealed at studying of CHD (Group 3).

1st group: Heavy abnormalities of haemodynamics are shown at the birth of the child or in the first day. Prevalence 2.5-3 to1000 live born babies. D-transposition of the main arteries, Fallot's tetrad, a hypoplasia of the right parts of heart (an atresia of the tricuspid valve, pulmonary valve, Ebstein's anomaly), a hypoplasia of the left parts of heart (an atresia of the aortic valve, an atresia of the mitral valve), single ventricle, a double origin from the right ventricle, a total anomalous pulmonary veins drainage, a critical pulmonary artery stenosis, Truncus arteriosus, the open atrioventricular canal, a big patent ductus arteriosus, ventricular septal defect, a critical aortic stenosis, a critical coarctation of aorta.

2nd group: Severity of abnormalities of haemodynamics depends on degree of manifestation of defect and indications for treatment require expert assessment. There is prevalence 3-1000 live born babies. Moderate or severe aortic stenosis, moderate pulmonary artery stenosis, noncritical coarctation of aorta big atrial septal defect, complex ventricular septal defects.

3rd group: Symptoms aren't shown. Low atrial septal defect, low ventricular septal defect, moderate pulmonary artery stenosis, bicuspid arterial valve, low patent ductus arteriosus. Thus, in recent years the growth of child incidence with cardiovascular diseases is probably substantially caused by wide use of modern methods of research and absence of accurate criteria between norm, pathology and congenital features of the formation, about which there is confirmed the data submitted by O.V. Sharapova.

It seems reasonable for rational use of health care resources and the organization of the optimum help to children with CHD that perinatal echocardiography has to be executed without fail to the pregnant women having risk factors of the child birth with CHD (women with existence of CHD, diabetes, elder than 40 years, etc.). The optimum term of detection of CHD at fetus during perinatal echocardiography it is considered the term from 18-20 weeks of pregnancy. The organization of the phased help is possible in the presence of the following conditions:

- Provision by the gynecologists of female consultations an information about risk factors of formation of CHD at a fetus
- Regulations of the direction of the pregnant woman on examination to the relevant medical institution (in which there are an ultrasonic equipment with the "fetal cardium" program and an expert with the corresponding qualification)
- Carrying out at the detection of CHD 1 and 2 of group, the combined CHD or the combined extracardiac pathology of the council of experts with the obstetrician-gynecologist, a neonatologist, the cardiosurgeon and parents of future child for the purpose of an assessment of expediency of maintenance of pregnancy and elaboration of further tactics of conducting pregnancy
- Regulations at maintenance of pregnancy of the procedure for tracking a pregnant woman (the choice of time of hospitalization in the perinatal center and a specialized maternity hospital, a planned attachment to neonatal cardiac surgery office according to opportunities of clinic and geographical availability to the pregnant woman)

Thus, it is necessary the development and creation of the adequate medical and organizational recommendation for process optimization the organization of medical care to the newborn and also creation of the methodological base providing successful functioning of the quality control system.

Objective of this research was studying of epidemiology of congenital malformations of the newborn in the Zhambyl Region.

MATERIALS AND METHODS

In the capacity of material was the retrospective analysi of 176 newborns histories with various CHD of the Regional Perinatal Center, Zhambyl Region for 2014-2015. CHD were registered according to the nomenclature headings Q20-Q28 "congenital anomalies of the blood circulatory system" of the 17 class of "congenital anomalies (malformations), deformations and chromosomal abnormalities" of the international statistical classification of the diseases and problems connected with health (the 10th revision). In the capacity of primary documentation there were used: histories of diseases of the children with malformations who were on treatment in the regional perinatal center of the Zhambyl Region. Frequency of congenital heart disease was calculated as ratio of number of all cases of defect to total number of live-born. For statistical development there were registered only those cases of malformations at children, whose parents constantly live at the territory of the Zhambyl Region. At the detection of the frequencies of separate nosological forms of congenital anomaly, the received relation was multiplied on 1000, i.e., the frequency of separate types of defects was calculated on 1000 births.

The obtained data were processed by means of the Statistica 5.5 program (StatSoft Inc. USA). Quantitative signs are presented as the arithmetic mean value ± a standard deviation, their comparison is executed with use of t-criterion of student. Comparison of frequency signs was carried out by means of criterion χ^2. As statistically significant there were considered the distinctions at $p<0.05$.

RESULTS AND DISCUSSION

The carried-out analysis was allowed to reveal that in 2014 from 7695 babies with heart anomalies there was found 84 children (1.1%) from them 31 boys, 51 girls. In 2015, there were registered 92 children (1.2%) with CHD on 7748 who were born and from them 48 boys, 44 girls (Table 1).

For 2 years period (2014-2015) on the Zhambyl Region 67 children (from them there were died 20) received the highly specialized medical care for newborns and infants with CHD in the 4 Child cardiac surgery centers (Astana, Almaty). The mortality rate was 29.8% (Table 2 and 3).

Among all CHD there were diagnosed more often the Ventricular Septal Defect (VSD) 4.9 (counting on 1000 child population); Patent Ductus Arteriosus (PDA) 2.4; Atrial Septal Defect (ASD) 0.7 and a pulmonary artery stenosis -0.6. There were detected less often transposition of great vessels, Tetralogy of Fallot and malformations of system of peripheric vessel. The combined CHD were made 23.85% from number of all heart diseases. In 2014, there were died from CHD 7 children in 2015-13. The lethality indicator was raised from 8.3-14.1%. But at the same time we would like to note that the child death was

Table 1: Prevalence and a lethality of phenotypical variants of CHD at children of the Zhambyl Region

	The absolute number of detected (structure (%))		The absolute number of dead (Lethality (%))	
Phenotypic variants of CHD	2014	2015	2014	2015
Congenital Ventricular Septal Defect (VSD)	43 (51.2)	32 (34.8)	-	2 (6.2)
Congenital Patent Ductus Arteriosus (PDA)	14 (16.7)	23 (25.0)	2 (14.2)	3 (13.0)
Congenital Atrial Septal Defect (ASD)	3 (3.6)	2 (2.2)	-	-
Congenital defect atrioventricular septum	1 (1.2)	4 (4.3)	-	3 (75.0)
Congenital atresia of the pulmonary valve	4 (4.8)	6 (6.5)	3 (75.0)	1 (16.6)
Other congenital malformations of great arteries (d-Transposition of the great arteries, d-TGA)	3 (3.6)	7 (7.6)	-	-
Tetralogy Of Fallot (TOF)	3 (3.6)	4 (4.3)	-	-
Congenital aortal valve stenosis	2 (2.4)	3 (3.3)	-	1 (33.3)
Syndrome of the left-sided hypoplasia of heart	2 (2.4)	3 (3.3)	-	1 (33.3)
Other congenital malformations of cardiac chambers and cardiac muscular fiber (single ventricle)	2 (2.4)	-	-	-
Congenital duplication of the right ventricle outlet	1 (1.2)	2 (2.2)	1 (100.0)	-
Syndrome of the right-sided heart hypoplasia	1 (1.2)	2 (2.2)	-	-
Common arterial trunk	1 (1.2)	-	-	-
Other congenital malformations of the aorta (aortic arch septum)	1 (1.2)	-	-	-
Total Anomalous Pulmonary Venous Drainage (TAPVD)	1 (1.2)	2 (2.2)	-	1 (50.0)
Ebstein's anomaly	-	1 (1.1)	-	1 (100.0)
Congenital aortic coarctation	2 (2.4)	1 (1.1)	1 (50.0)	-
Total	84 (100)	92 (100)	7 (8.3)	13 (14.1)

Table 2: Number of children who received highly specialized medical care in Almaty and mortality from CHD

Name, medical organizations	Period (according to the years)	The absolute number of children with congenital heart disease	Dead
RSE "Scientific	2014	11	4
Center of Pediatrics and	2015	24	6
Pediatric Surgery"			
SMGE "Center for and	2014	10	3
Perinatology Pediatric	2014	14	4
Cardiac Surgery"			

Table 3: Number of children who received highly specialized medical care in Astana and mortality from CHD

Name, medical organizations	Period (according to the years)	The absolute number of children with congenital heart disease	Dead
AJSC "National Science	2014	2	1
Cardiac Surgery Center"	2015	3	1
RSE "National Scientific	2014	1	-
Medical Center"	2015	2	1

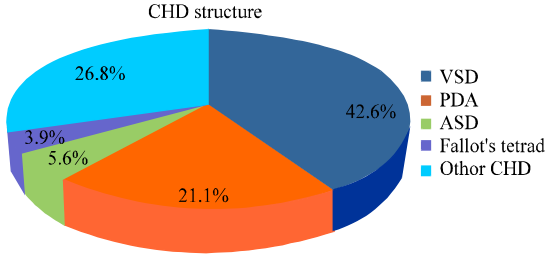

Fig. 1: Structure of CHD at newborns and 1st year of life children (2014-2015) of the Zhambyl Region

connected more often with the combined complex heart diseases or with the complications, arisen at the moment of surgical correction or at the postoperative period (Fig. 1).

CONCLUSION

Thus, during 2014-2015 it is noted the tendency to increase in birth of children with congenital heart diseases. At the present stage, features of CHD histories are growth of their frequency, increase of the specific weight of the complex and combined heart diseases. CHD natural clinical course is characterized by a high lethality of children, especially on the 1st year of life. Timely detection of CHD with the subsequent correction at early age will be promoted to decrease in death rate and an disability among children. Unfortunately, many CHD are not diagnosed on time or there are revealed when it is impossible to cure children.

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