

Optimization of medical and surgical care for congenital heart defects in children

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ABSTRACT

In a review of the literature discussed the organization of medical care for children with congenital heart disease; especially natural course of the disease; quality of life and social aspects; rehabilitation of patients. According to published research articles and monographs on the problems of congenital heart defects in children, it concluded that, despite the high achievement of modern medicine in this area, are still not fully explored optimistsatsiya medical care with congenital heart disease and as a consequence, improve the quality and duration of their life.

An article 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 South Region Kazakhstan for 5 years (2020-2024). Frequency of all heart diseases on the average has 9,1 to 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 first year of life.

Keyword:

children; congenital heart disease; optimization of medical care.

INTRODUCTION

Congenital heart defects (CHD) are the most common congenital birth defects affecting 1–2% of all live births globally with an estimated incidence of 8–10/1000 live births⁽¹⁻³⁾. There are regional differences in prevalence and incidence due to genomic, clinical and environmental factors ^(4,5). CHD is defined by structural and functional malformations of the heart which if not managed through appropriate interventions affects quality of life of the individual and potential premature death. Early detection, diagnosis and prompt interventions result in significantly decreased morbidity and mortality⁽⁶⁻⁷⁾. The estimated incidence of CHD is similar across all countries globally [1, 8]; however, CHD data for African countries is sparse and needed⁴. CHD malformations occur as single lesions or in combination with other heart defects ⁹. Commonly diagnosed CHD lesions isolated or single lesions include atrial septal defects (ASD), ventricular septal defects (VSD), and pulmonary stenosis (PS). Complex or combination lesions include atrioventricular septal defects (AVSD), tetralogy of Fallot (TOF), and transposition of the great arteries (TGA) ⁽⁹⁻¹¹⁾. TOF and TGA are the two most common cyanotic CHD conditions that result in oxygen saturation below 90%. Majority of septal defects

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such as VSD, ASD and AVSD are classified as acyanotic CHD lesions with oxygen saturation normally above 95%^(3,12,15).

Congenital heart defects (CHD) are an important problem in pediatrics and cardiac surgery due to their high prevalence and the need for early surgical correction due to significant health disorders and disability in children^(8,16). According to various authors, congenital heart defects are found in 0.7-1.7% of newborns. According to WHO, congenital heart disease occurs in 1% of newborns, regardless of the level of medicine in the country^(17,18).

The organization of medical care for the population with congenital malformations of the cardiovascular system is an important task of public health. Protecting the health of the child population is one of the conditions for the sustainable development of the future of Kazakhstan.

In Kazakhstan, congenital heart defects (CHD) remain one of the leading causes of infant and child mortality. Each year, more children are diagnosed with these conditions and more infants with CHD are surviving birth, which noticeably influences the overall health profile of the population. This makes the importance of pediatric cardiac surgery undeniable.

Reducing mortality and improving the long-term outlook for newborns with heart and vascular abnormalities has long been one of the most challenging and urgent tasks in pediatric cardiac care — and it continues to be a critical priority today.

MATERIALS AND METHODS

To study the contribution of CHD to perinatal and early childhood mortality, disability, and improvements in specialized medical care.

Characteristics of research materials

The study was performed in accordance with the standards of Good Clinical Practice and the principles of the Helsinki Declaration. The protocol of the study was confirmed by the ethics committee of the South Kazakhstan medical academy. Written information consent about the treatment procedure and possible complications was received from the parents of all study participants.

This research work was carried out in the «Shymkent City Children's Hospital» and «Regional Children's Hospital» in Shymkent, Regional Perinatal Center,

“Scientific center of Pediatrics and Pediatric Surgery” in Almaty and “Perinatology and pediatric cardiac surgery”.

The object of clinical research: In the South region for 2020-2024, children under 1 year old with congenital heart defects and for the first time in their lives were formed.

The course of the analysis of the prevalence of certain nosological types of congenital heart malformations was recorded in accordance with the positions of the International classification of Diseases (ICD) and the nomenclature for health (10th revision) recommended by WHO Q20-Q28 “Congenital malformations of the circulatory system” of the XVII class “congenital anomalies [malformations], deformities and chromosomal anomalies”.

Ethical clearance

This study was conducted in accordance with ethical standards. Ethical approval was obtained from the appropriate institutional review board, and informed consent was secured from all participants prior to data collection.

Results and discussion

Today, the effective organization of care for patients with cardiovascular diseases is hindered by the lack of continuity in providing care at different stages of its implementation, and the variety of management approaches for children. There are very few studies in the domestic and foreign literature assessing the need of the population of children under 1 year of age for certain types of cardiac surgery. The results of treatment in regional high-tech medical institutions depend on the organization of medical care for children under 1 year of age with CHD in the field. Modern diagnostic methods make it possible to detect in fetuses and newborns those congenital malformations and abnormalities of the heart and blood vessels that previously remained undiagnosed and, as a result, not operated on. As experience gained, treatment tactics and methods improved, and strict indications for performing a particular type of surgery were determined.

Checklist method

In order to identify risk factors causing CHD, in 2020-2024, a questionnaire survey of parents of healthy and young children with CHD, consisting of 24 special requests, was conducted in the South regional Perinatal Center. This test questionnaire was prepared by Vasiliev V. V., 2018; Tikhonova O. S., 2020 and at the suggestion of the co-authors (Table.1) [18].

Table 1-The main risk factors causing CHD

Risk factors	Gradiations	diagnostic coefficient	information coefficient
1	2	3	4
Medical and biological			
The presence of infectious infections in parents	yes	5	0,40
	no	-2	0,24
The presence of intrauterine infections in the mother: cytomegalovirus, ureaplasmosis, viral hepatitis B, chlamydia, syphilis	yes	5	0,36
	no	-2	0,35
Pregnancy:			
1-st	yes	0	0,01
2-st	yes	-1	0,05
3-st	yes	1	0,02
4-st and higher	yes	5	0,26
The presence of a medical abortion in the mother's anamnesis	yes	4	0,38
	no	-1	0,08
The presence of anemia in the mother during pregnancy	yes	3	0,37
	no	-2	0,25
Exacerbation of chronic diseases during pregnancy in the mother	yes	3	0,26
	no	-1	0,13
Social and hygienic			
Mother's smoking	yes	4	0,29
	no	-1	0,05
Father's smoking	yes	4	0,29
	no	-1	0,05
Mother's alcoholism	yes	3	0,50
	no	-2	0,45
Father's alcoholism	yes	3	0,50
	no	-2	0,45
Demographic			
Mother's age			
≥ 18	yes	3	0,26
≤ 35	yes	-1	0,13
Incomplete family	yes	9	0,61
	no	-1	0,04
Mother's education:			
Average	yes	3	0,08
Special professional	yes	3	0,50
Higher	yes	-3	0,55
Father's education:			
Average	yes	1	0,01
Special professional	yes	3	0,50
Higher	yes	-5	0,82

The questionnaire card consists of 3 sections, taking into account the causes of the main risk factors for CHD. The results of multifactorial studies are presented in a special forecast table – according to 14 main features. To conduct surveys of parents, gradational signs are recorded in accordance with the diagnostic coefficients of each trait, after which it is necessary to add all the signs (Table 1). With the sum of the coefficients +13 or higher, the forecast of the birth rate of newborn CHD is 95%. The questionnaire for parents, consisting of

3 parts, indicated by 14 main features, requests were filled out voluntarily. Standard methods were used to calculate statistical indicators. According to the results of the study, groups were formed and a mutual correlation was revealed.

The carried-out analysis was allowed to reveal that in 2018 from 7695 babies with heart anomalies there was found 84 children (1,1%), from them 31 boys, 51 girls. In 2019 there were registered 92 children (1,2%) with CHD on 7748 who were born, and from them 48 boys, 44 girls.

Table 2. Prevalence and of phenotypical variants of CHD at children of the South Region Kazakhstan

Phenotypic variants of CHD	The absolute number of detected				
	2020	2021	2022	2023	2024
Congenital ventricular septal defect (VSD)	43 51,2%	32 34,8%	38 40,9%	39 44,8%	31 45,6%
Congenital patent ductus arteriosus (PDA)	14 16,7%	23 25,0%	26 28,1%	22 25,3%	17 25%
Congenital atrial septal defect (ASD)	3 3,6%	2 2,2%	2 2,1%	3 3,5%	3 4,4%
Congenital defect atrioventricular septum	1 1,2%	4 4,3%	5 5,4%	1 1,2%	2 2,9%
Congenital atresia of the pulmonary valve	4 4,8%	6 6,5%	1 1,1%	4 4,6%	3 4,4%
Other congenital malformations of great arteries (d-Transposition of the great arteries, d-TGA)	3 3,6%	7 7,6%	3 3,2%	6 6,9%	4 5,9%
Tetralogy of Fallot (TOF)	3 3,6%	4 4,3%	6 6,4%	3 3,4%	1 1,5%
Congenital aortal valve stenosis	2 2,4%	3 3,3%	2 2,1%	4 4,6%	2 2,9%
Syndrome of the left-sided hypoplasia of heart	2 2,4%	3 3,3%	1 1,1%	-	-
Other congenital malformations of cardiac chambers and cardiac muscular fiber (single ventricle)	2 2,4%	-	3 3,2%	1 1,2%	1 1,5%

Phenotypic variants of CHD	The absolute number of detected				
	2020	2021	2022	2023	2024
Congenital duplication of the right ventricle outlet	1 1,2%	2 2,2%	-	-	-
Syndrome of the right-sided heart hypoplasia	1 1,2%	2 2,2%	1 1,1%	-	1 1,5%
Common arterial trunk	1 1,2%	-	2 2,1%	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 1,2%	1 1,5%
Ebstein's anomaly	-	1 1,1%	2 2,1%	-	-
Congenital aortic coarctation	2 2,4%	1 1,1%	1 1,1%	2 2,3%	2 2,9%
Total	84 100%	92 100%	93 100%	87 100%	68 100%

Table 3. Lethality of phenotypical variants of CHD at children of the Turkestan region

Phenotypic variants of CHD	The absolute number of dead Lethality (%)				
	2020	2021	2022	2023	2024
Congenital ventricular septal defect (VSD)	-	2 6,2	1 16,6	-	3 9,6
Congenital patent ductus arteriosus (PDA)	2 14,2	3 13,0	1 3,8	1 4,5	2 11,7
Congenital atrial septal defect (ASD)	-	-	1 50,0	-	-
Congenital defect atrioventricular septum	-	3 75,0	-	3 75,0	-

Phenotypic variants of CHD	The absolute number of dead Lethality (%)				
	2020	2021	2022	2023	2024
Congenital atresia of the pulmonary valve	3 75,0	1 16,6	-	1 16,6	2 75,0
Other congenital malformations of great arteries (d-Transposition of the great arteries, d-TGA)	-	-	1 33,3	-	-
Tetralogy of Fallot (TOF)	-	-	2 33,3	-	-
Congenital aortal valve stenosis	-	1 33,3	-	1 33,3	1 50
Syndrome of the left-sided hypoplasia of heart	-	1 33,3	1 100	1 33,3	-
Other congenital malformations of cardiac chambers and cardiac muscular fiber (single ventricle)	-	-	1 33,3	-	-
Congenital duplication of the right ventricle outlet	1 100,0	-	-	-	1 100,0
Syndrome of the right-sided heart hypoplasia	-	-	1 16,6	-	-
Common arterial trunk	-	-	-	-	-
Other congenital malformations of the aorta (aortic arch septum)	-	-	1 50,0	-	-
Total anomalous pulmonary venous drainage (TAPVD)	-	1 50,0	-	1 50,0	-
Ebstein's anomaly	-	1 100,0	-	1 100,0	-
Congenital aortic coarctation	1 50,0	-	-	-	1 50,0
Total	7 8,3%	13 14,1%	10 10,7%	16 18,2%	12 17,6%

For two year period (2020-2024) on the South Region kazakhstan 132 children (from them there were died - 58) 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 4. 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 Center of Pediatrics and Pediatric Surgery"	2020	11	4
	2021	24	6
	2022	21	5
	2023	13	2
	2024	10	2
SMGE "Center for Perinatology and Pediatric Cardiac Surgery"	2020	10	3
	2021	14	4
	2022	8	1
	2023	12	2
	2024	9	1

Table 5. 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
"National Science Cardiac Surgery Center"	2020	2	1
	2021	3	1
	2022	4	-
	2023	2	-
	2024	2	-
"National Scientific Medical Center"	2020	1	-
	2021	2	1
	2022	2	-
	2023	5	-
	2024	3	-

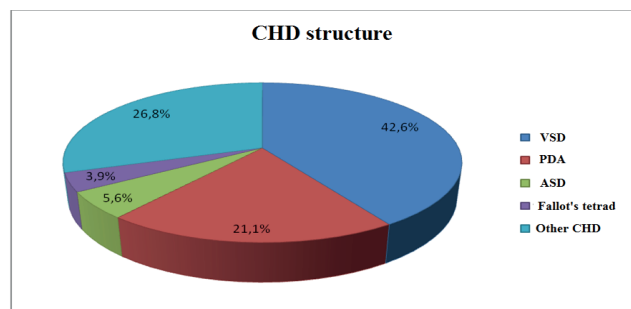


Figure 1 - Structure of CHD at newborns and 1st year of life children (2020-2024) of the South Region Kazakhstan

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 2018 there were died from CHD 7 children, in 2022 – 12. The lethality indicator was raised from 8,3% to 18,2%. But, at the same time we would like to note that the child death was 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.

In Kazakhstan, CHD occupies one of the first places in the structure of child mortality. The percentage of detectability and fertility of children with this pathology is increasing every year. About 3,000 children with CHD are born in the Republic every year, 80% of them die before the age of one, up to 20% in the first weeks of life, and up to 27% in the first month. In the age structure of mortality from congenital anomalies of the heart and great vessels - 91% are children of the first year of life, among them more than half are children of the neonatal period (the first 28 days of life).

The slowdown in the rate of decline in infant mortality rates, the increase in child disability rates, the high prevalence of congenital heart defects and the unsatisfactory effectiveness of providing qualified care to children in these conditions raises the issues of improving the system of organization of qualified medical care to the rank of urgent tasks of the state and society. Despite the efforts being made, there is an increase in the detection of congenital heart defects. The

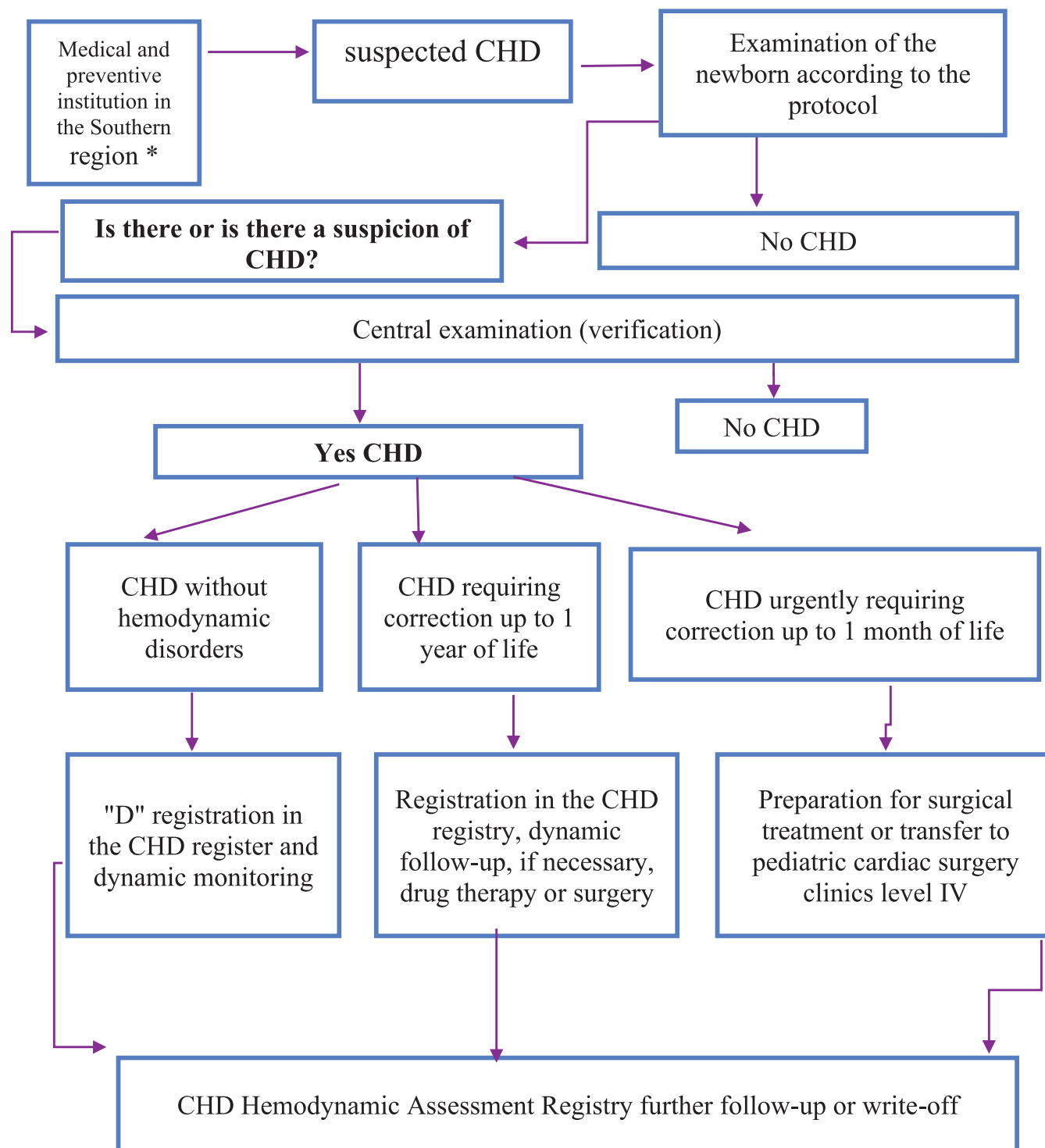


Figure 2. Algorithm of periodic medical (surgical) care for the detection of CHD at the antenatal and postnatal stages

relevance and relevance of cardiac surgery in children is beyond any doubt. The problem of reducing mortality and improving the prognosis of newborns with heart and vascular malformations has always been the most urgent and difficult to solve in pediatric cardiac surgery and remains so today [19].

The most important task of the current stage of development of pediatric cardiology and pediatric cardiac surgery is the organization of medical care for congenital malformations of the cardiovascular system. More than 90 nosological forms of congenital heart defects (CHD) are known. Modern medical technologies make it possible to provide assistance for diseases that seemed inaccessible 10 years ago, and the number of patients who have received timely cardiac surgery is increasing and, thanks to this, have survived the “critical age of the natural course of the disease.”

In general, there is an increase in the number of cardiac surgery clinics in the Republic of Kazakhstan, which makes specialized medical care more accessible. In addition, the range of services provided by cardiac surgery clinics is a very expensive undertaking for any state. Therefore, it is aimed at solving tasks that require high costs and involve a large number of participants in order to achieve the final result. To organize optimal medical care, it is necessary to use formal decision-making technologies (system analysis, algorithms of actions).

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state. Therefore, it is aimed at solving tasks that require high costs and involve a large number of participants in order to achieve the final result.

CONCLUSION

Thus, during 2020 - 2024 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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Authors's contribution

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