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The results of a three-year research program on assessment of primary diagnostic issue of congenital heart defects in children <1 year and the availability of cardiac surgical care in the Kyrgyz Republic

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Abstract

In this article, the results of 3-year research program on primary diagnostics of congenital heart defects in children < 1 year of age are presented.

Key words: congenital heart disease, echocardiography, diagnosis, cardiac surgical care

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Introduction

The congenital heart diseases (CHD) are defined as proposed by Mitchell et al: "a gross structural abnormality of the heart or intrathoracic great vessels that is actually or potentially of functional significance" (1). The frequency of CHD at birth (sometimes referred to as the prevalence per childbirth) depends on how the population is studied (2). According to the World Health Organization (WHO), 1.35 million newborns with CHD are born each year (3).

When examining newborns using the echocardiography, small defects (small defects of the interventricular and interatrial septa, small sized patent ductus arteriosus (PDA)) predominate. With the inclusion of all forms of CHD into study, the prevalence of CHD, according to Hoffman and Kaplan (4) can reach 75 cases per 1000 live births. Performing an echocardiogram in all newborns revealed that about 5% have small ventricular septal defects, most of which spontaneously close for up to a year. Vast majority of newborns have delayed closure of the PDA (4). In 90% of cases CHDs are found in general populations without any particular risk factor, and only in 10% of cases CHD is associated with certain risk factors (5).

Early detection of CHD and timely cardiac surgery is essential for prognosis in children with heart defects. The total mortality from CHD without intervention is sufficiently high. Usually, in most low- and middle-income countries, the diagnosis of CHD is considerably delayed (6). The average age of the primary diagnosis establishment can vary from the first day of life to almost 80 years (7).

A previous study of CHD in Kyrgyz Republic territory showed that only 14% of children had been diagnosed before discharge from the maternity hospital, and 86% of patients had a delayed diagnosis (8).

The aim of study is evaluation of the scope of problem and gaining of novel statistical data on prevalence and detectability of CHD in various regions of Kyrgyz Republic

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Objectives:

1. To assess the significance of contribution of various risk factors in complex CHD formation in intrauterine period
2. To evaluate the prevalence of CHD in newborns of Kyrgyz Republic
3. To determine the causes of disability and early mortality in adults with CHD.

Methods

The study was conducted in all territories of the Kyrgyz Republic (KR), in the high-altitude, mid-altitude and low-altitude regions, including the most remote areas (Table 1). The researchers organized more than 60 on-site evaluation of the local population.

The population of the country, according to the National Statistics Committee, is 6.4 million citizens. The average annual birth rate in the country over the past three years is 158.160 thousand annually.

Investigation was achieved by 2D transthoracic echocardiography assessment (US scanners of General Electric Vivid E9, Phillips CX – 50 (portable)), prenatal screening was conducted by convex transducer and fetal echo program. Included groups were: newborns of 0-28 days, children under 1 year, pregnant women with more than 20th week gestation.

Table 1. Regions, districts of Kyrgyz Republic where study was conducted (geographical altitude is shown)

	Regions of Kyrgyz Republic	Districts	Altitude
2016			
1	Chuy region	Kemin district: Orphanage District hospital, Orlovka village	Mid-altitude
2	Issyk-Kul region	Jeti-Oguz district	High-altitude
		Ak-Suu district, Ak-Chiy djayloo	High-altitude
		Karakol city	Mid-altitude
3	Talas region	Regional maternity hospital and pediatric unit of Talas Regional Hospital	Mid-altitude
		Talas district, maternity hospital	Mid-altitude
		Talas district, Karaoy djayloo	High-altitude
4	Batken region	Kyzyl-Kiya city	Low-altitude
5	Djalal-Abad region	Aksy district	Mid-altitude
		Ala-Buka district	High-altitude
6	Bishkek city	City perinatal center	Mid-altitude
		City maternity hospital №2, №4, №6, Chuy regional maternity hospital.	Mid-altitude
		Baby house	Mid-altitude
2017			
7	Chuy region	Moskva district Regional hospital maternity unit	Mid-altitude
		Tokmok city	Mid-altitude
		Issyk-Ata district Regional hospital maternity unit	Mid-altitude
		Djayil district Regional hospital maternity unit	Mid-altitude
		Sokuluk district Regional hospital maternity unit	Mid-altitude
		Kemin district Regional hospital maternity unit	Mid-altitude
8	Naryn region	Naryn city	High-altitude

		Ak-Talaa district Regional hospital maternity unit	High-altitude
		At-Bashy district Regional hospital maternity unit	High-altitude
9	Batken region, Leylek district	Isfana city	Low-altitude
		Dostuk village	Low-altitude
		Kulundu village	Low-altitude
		Katyran village	Mid-altitude
10	Batken region, Batken district	Batken city	Mid-altitude
		Kyzyl-Kiya city	Mid-altitude
		Suluktu city	Mid-altitude
11	Batken region, Kadamjay district	Kadamjay city	Mid-altitude
12	Osh region	Osh city, regional unified territorial hospital	Mid-altitude
		Osh city, city perinatal center	Mid-altitude
		Alay region	High-altitude
		Gulcho village	High-altitude
13	Bishkek city	City perinatal center City maternity hospital №2, №4, №6, Chuy maternity hospital National Institute of maternal and pediatric care City pediatric clinical hospital № 3	Mid-altitude
2018			
14	Chuy region	Chuy regional maternity hospital	Mid-altitude
		Tokmok city Republic orphanage disability center	Mid-altitude
		Moskva district Regional hospital maternity unit	Mid-altitude
		Belovodskoye village Belovodskoe psychoneurological house Belovodskoye baby house	Mid-altitude
15	Naryn region	Naryn city	High-altitude
16	Djalal-Abad region	Toguz-Toro district, Kazarman village	Mid-altitude
17	Batken region, Leylek district	Isfana city	Low-altitude
		Suluktu city	Low-altitude
		Kulundu village	Low-altitude
18	Batken region, Batken district	Batken regional hospital, maternity unit	Mid-altitude
		Zardaly village	High-altitude
19	Bishkek	Republic specialized center of rehabilitation of pediatric patients with central nervous system disorder City perinatal center City maternity hospital №2, №4, №6, Chuy maternity hospital National Institute of maternal and pediatric care City pediatric clinical hospital № 3	Mid-altitude

Results

The research study was conducted by two sections: prenatal diagnostics and workup of newborns and children under 1 year

I. Prenatal diagnostics: Prenatal echocardiography is the most valuable method for the diagnosis of fetal heart disease. As a non-invasive and innocuous procedure, it allows examiner to fully evaluate cardiac structures, assess intracardiac hemodynamics in the prenatal period and makes it available for dynamic control during pregnancy. All these qualities determine the priority of this method in the prenatal diagnosis of fetal congenital heart disease.

A standard fetal cardiac scan is carried out at 18-22 weeks of gestation in accordance with the protocol of the reference centers (5).

We started examinations of pregnant women from the 21st week of gestation, which was explained by the first visits of women to our center.

Table 2. Number of conducted examinations

		Retrospectively 2010 - 2015	2016	2017	2018	Total
1.	Fetal echo in SRIHSOT	850	79	57	62	198
2.	Fetal echo in regions		34	63	39	136
	Total		113	120	101	334
	Retrospectively	850				

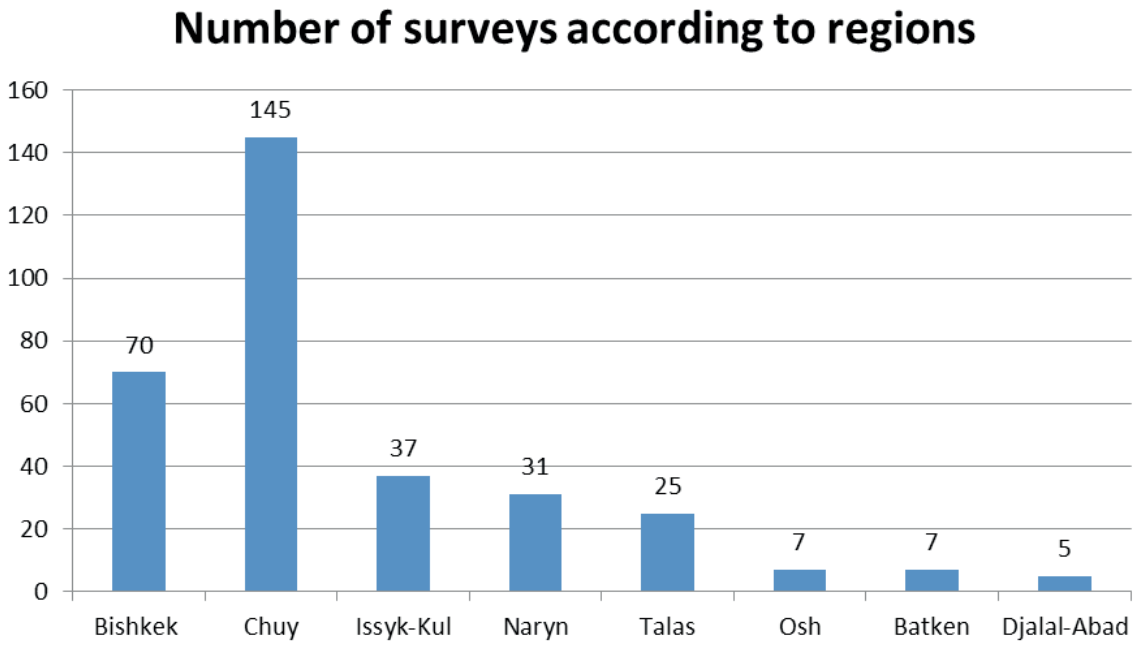

Figure 1. Number of fetal examinations by regions of Kyrgyz Republic

Table 3. Detectability of CHD by fetal echocardiography

	Years	Investigated	Detected	%
1	2016	113	14	12.39%
2	2017	120	13	10.84%
3	2018	101	8	7.92%
	Total:	334	35	10.48%

CHD – congenital heart disease

Table 4. Detected CHD forms by fetal echocardiography

	CHD forms		Outcome
2016	Atrio-ventricular septal defect, complete type	2	
	Single ventricle	1	
	Ventricular septal defect	5	
	Ventricular septal defect (muscular)	3	
	ASD in IAS aneurysm	2	
	Atrial septal defect	1	
2017			
	Atrio-ventricular septal defect, complete type	4	
	Ventricular septal defect	2	
	Single case –ASD in IAS aneurysm	1	
	Hypoplastic left heart syndrome	1	
	Total anomalous pulmonary venous connection with tricuspid anomaly	1	
	Tricuspid atresia	2	
	Hypoplastic right heart syndrome	2	
2018			
	Aortic dextraposition. VSD. Hypoplastic pulmonary artery.	1	Ds has been confirmed, patient is on follow-up
	PA stenosis (valvular), ASD	1	Ds has been confirmed, lethal outcome on 10th day
	Ventricular septal defect (muscular)	1	-
	Persistent truncus arteriosus, Double-chamber heart.	1	-
	Ventricular septal defect	1	-
	Atrio-ventricular septal defect, complete type	1	Did not born
	Atrio-ventricular septal defect, complete type	1	?
	Total anomalous pulmonary venous connection with tricuspid anomaly, TR moderately severe	1	Ds has been confirmed, lethal outcome on 3rd day

ASD – atrial septal defect, CHD – congenital heart disease, IAS – interatrial septum, TR – tricuspid regurgitation, VSD – ventricular septal defect

Table 5. Examination of the risk groups

	Risk factors of CHD	Risk groups	Detected CHD	%
1	Existence of CHD in mother	58	10	17.24%
2	Prior pregnancy CHD	10	4	40%
3	Arrhythmias in mother	3	1	33.34%
4	Chronic rheumatic heart disease	1	-	-
5	Somatic disease in mother (SLE)	1	-	-

CHD – congenital heart disease, SLE – systemic lupus erythematosus

II. Primary workup of CHD in newborns and children under 1 year:

The study included newborns and children under 1 year old in the investigated area. The presence of a structural anomaly of the heart was regarded as the presence of CHD. Patients with congenital rhythm disturbances, cardiomyopathy, myocarditis, additional persistent vena cava, patent foramen ovale, acquired valvular heart disease were excluded.

A total of 4177 patients were examined, 523 had a primary diagnosis of congenital heart disease, which was 12.52%.

Table 6. Results of primary examination of children

	Study	2016	2017	2018	Total	Total number of investigated children
1	Echo in SRIHSOT					4177
2	Echo in regions	531	579	784	1894	

Table 7. Detectability of CHD

	0 – 1 month	1 month – 1 year	Total
Examined	998	3179	4177
Detected CHD	240	283	523
Detectability%	24.12%	8.93%	12.52%

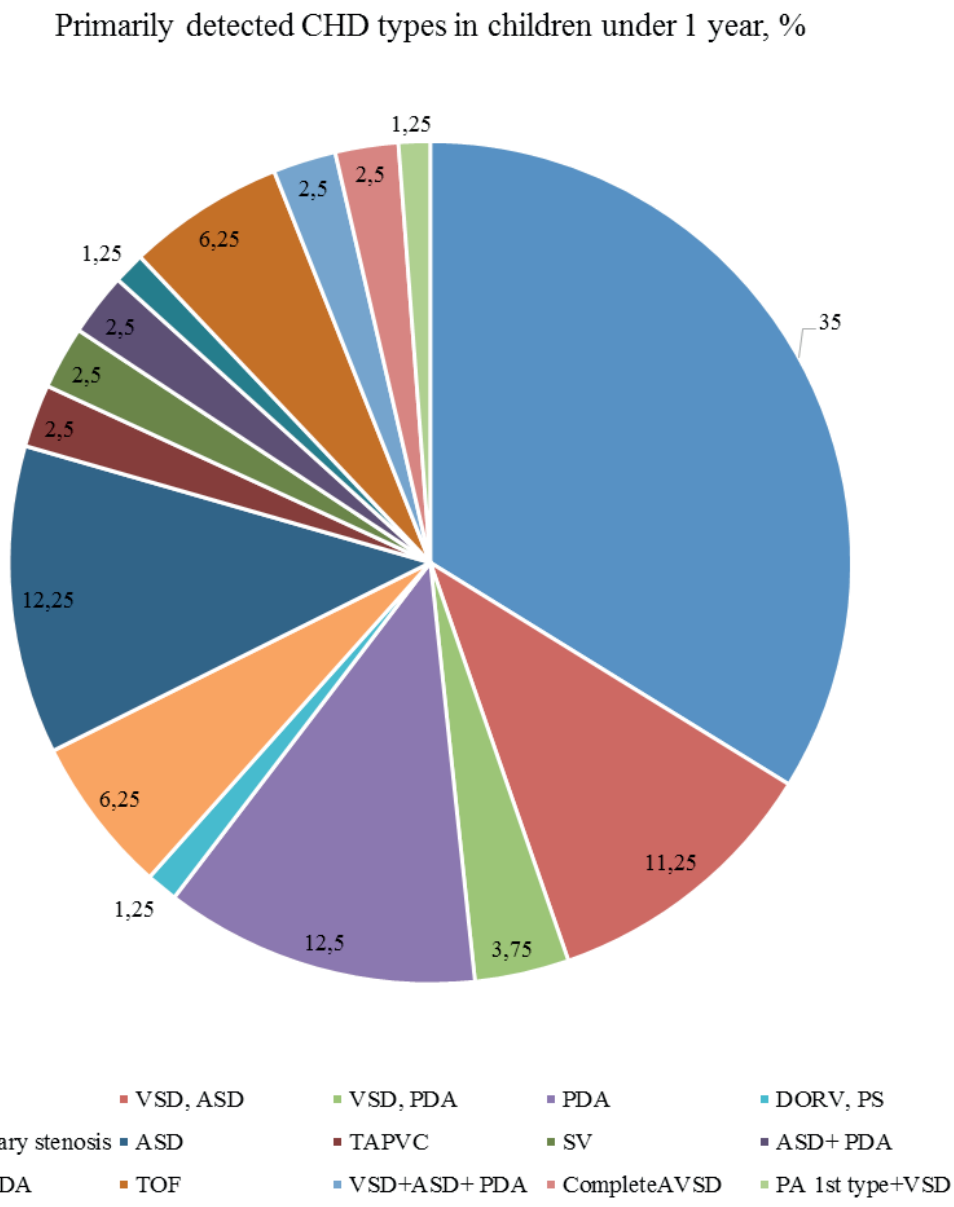

Figure 2. CHD forms detected in primary examination of children <1 year old

ASD – atrial septal defect, AVSD – atrioventricular septal defect, CHD – congenital heart disease, DORV – double-outlet right ventricle, PA-pulmonary atresia, PDA – patent dusctus arteriosus, PS – pulmonary stenosis, SV – single ventricle, TAPVC – total anomalous pulmonary vein connection, TOF – tetralogy of Fallot, VSD – ventricular septal defect

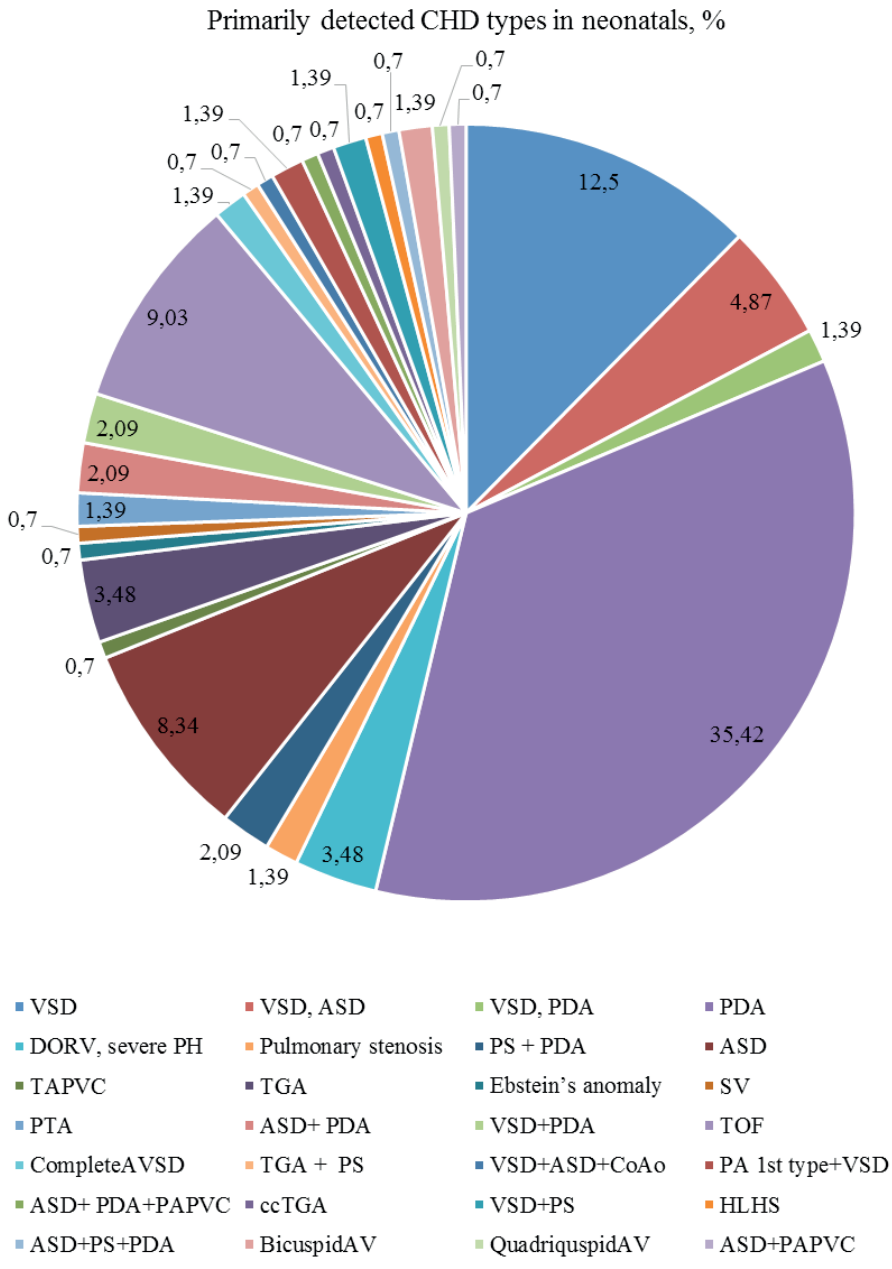

Figure 3. CHD forms detected in primary examination of newborns (0-28 days)

ASD – atrial septal defect, AV – aortic valve, AVSD – atrioventricular septal defect, CHD – congenital heart disease, CoAo – coarctation of aorta, DORV – double-outlet right ventricle, PA- pulmonary atresia, PAPVC – partial anomalous pulmonary vein connection, PDA – patent dusctus arteriosus, PH – pulmonary hypertension, PS – pulmonary stenosis, SV – single ventricle, TAPVC – total anomalous pulmonary vein connection, TGA – transposition of great arteries, TOF – tetralogy of Fallot, VSD – ventricular septal defect

1. In newborns group cyanotic defects rate was 26.38%, non-cyanotic defects – 73.61%.
2. In the group of children 1 month-1 year cyanotic defects 14.70%, non-cyanotic defects – 85.29%

Discussion

According to European authors, only 49% of pregnancies in Europe are planned. On the territory of our country, the exact data of the planned pregnancies is unknown, but according to a preliminary survey of the women examined, only 5% of the pregnancies were planned. The program was implemented with a focus on women with CHD in history. Only 21% of the examined pregnant women were at risk of having children with CHD.

The accessibility of the study is the highest in the Chuy region and the capital of the country - Bishkek, that is, in areas located in close proximity to the central institution, and it is probably higher due to the internal migration, because the registration address during the survey have not been supplied.

The influence of high-altitude has not been revealed as a causative factor, which corresponds to the international data obtained in different years and in different altitudes (9-12).

Based on the obtained results, the most common forms of CHD (in children under 1 year old and newborn babies) in our country are "acyanotic" heart defects, such as the patent ductus arteriosus (12.5% and 35.42%), atrial septal defect (11.25% and 8.34%), ventricular septal defect (32% and 12%) and their combinations. On the other hand, assessment of newborns and the children under 1 year reveals the high prevalence of critical and complex forms of CHD requiring well-qualified care and timely surgical treatment. The proportion of detection of cyanotic defects is significantly reduced with age, which also correlates with international data that more than half of children born with cyanotic CHD do not live up to 1 year.

The main causes of disability and early mortality in vast majority of patients with CHD in remote regions of Kyrgyz Republic are late detection, lack of accessible specialized medical care, and low awareness of the parents/relatives of the patient about the importance of timely surgical treatment.

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Conflict of interest: None to declare

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