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CLINICAL EVALUATION OF CONGENITAL HEART DISEASE IN CHILDREN IN TUZLA CANTON AREA

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Abstract

The purpose of this study has been evaluation our possibilities of diagnostic and treatment children with congenital heart disease. We were analyzing medical documentation from 352 children with discovered congenital heart disease in period of six years in Tuzla Canton area. disease. The average age at diagnosis was $2,15 \pm 2,28$ years. During first cardiac examination 51,98% children presented symptoms of cardiac disease. Growth retardation in postnatal period is noticed in 13,35%. Electrocardiographic changes are registered in 47,76%, while changed chest radiogram in 53,85% of patients. Medicament treatment was needed in 20,74% and cardiovascular surgery procedure in 40,60%. Urgent surgery treatment was needed in 62 patients with critical heart disease. Cardiovascular surgery was made in 23,29% of patients, and average age was $4,81 \pm 3,23$ years. Mortality of children with congenital heart disease is relatively high and it is 19,60%, the biggest number of them (95,65%) died before and only 4,35% after cardiosurgical treatment. Average age of children who died was $0,51 \pm 0,59$ year. Considering clinical indicators of congenital heart disease in Tuzla Canton area, it is evidenced that this children health problem, which due difficult clinical picture, big involvement in infant mortality, growth retardation with progression in chronic cardiopathy, require urgent measures with main goal on advanced organizing of it's diagnostic and treatment.

Key words: *congenital heart disease, children, Tuzla Canton area.*

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Introduction

Congenital heart diseases (CHD) present heterogeneous group of morphological and functional changes of heart and big blood vessels. With its high prevalence, from 5 to 10 cases in 1000 live newborns children, they are on the second place among all congenital anomalies. Clinical demonstration of CHD depends on morphological complexity of disease, or on level of hemodynamical disorders and age of children, as well as combination with other diseases and disorders. For adequate medical treatment of CHD is necessary timely diagnostic, or early definition of morphological and hemodynamical disorders on heart. According to information from Program of Pediatric cardiology from New England (1), excluding premature with open patent ductus arteriosus in 10 children with CHD, three are requesting cardio surgical treatment in its early childhood, and five are requesting some of therapeutic procedures during childhood.

Prevalence of CHD in Tuzla Canton area is 6,12 in 1000 of live born, and 1,46 in 1000 live born have critical disease of heart, or anomaly which vitally endangers its health during first days of life (2).

Course and prognosis of diseases depend on time of diagnosing, type of anomaly, degree of hemodynamical disorders and development level of pediatric cardiology and cardiosurgery. Considering variations of clinical picture, course of disease could be followed in wide diapason, from heavy anomaly with fatal end in first hours or days of life up to those, which have spontaneous recovery. According to Brodeu (3) (1989) 25% of not treated children with heart disease die in first month of their life, and 25% until the end of first year of life, and 12% until age of 15.

Mortality rate is significantly different in relation to type of anomaly and level of socio-economical development of the region in which anomaly has been discovered. In our country due impossibility to offer early cardiac surgical therapy, mortality is almost hundred percentage in children with transposition of great artery, hypoplastic heart left syndrome, atresion of the tricuspid or pulmonary valve and other complex anomaly.

With long-term following it has been registered that children with CHD in Tuzla Canton area present significant part of pathology of children, and that its treatment is not in accordance to recent modern accomplishments in field of pediatric cardiology and cardiac surgery, and in accordance to above mentioned this problem could be considered as open one. That is a reason why this research has as a main goal analyzing of diagnostic and treatment possibilities of these anomalies and gained results are used as basic for resolving of above mentioned problem.

Material and methodes

Source of information was register at Department of cardiology at Pediatric Clinic in Tuzla, which is in advanced conceived to collect relevant information on CHD. By research has been included medical documentation of 352 patients both sex, age up to 14 years, in which have been discovered congenital heart disease in period from 1.1. 1994. to 31.12. 1999. In research were included all patients in which have been determined CHD with echocardiography, catheterization or autopsy. In research were not included children with bicuspidal aortal valve without aortal stenosis, premature children with patent ductus arteriosus which closed until third months of life, as well as children with mitral valve prolapse which is not part of Marfan syndrome.

The method of work was retrospective analyze of medical documentation from Department of Cardiology at the Pediatric Clinic in Tuzla. There were analyzed anamnesis, clinical manifestation, diagnostically procedures (electrocardiography, chest radiografy and echocardiography), usage of therapeutically measures and results of disease in children with proved CHD. In patients who died due CHD it was analyzed autopsy findings if was done.

Results

Age when CHD was diagnosed is shown in Table 1. CHD in fetal age has not been discovered, but there were 188 diagnosed cases in newborn and infant age, from which 61 or 32,45% were in year of 1999. Average value of life age in which CHD was diagnostic was $2,15 \pm 2,28$ year.

Table 1. *Age when congenital heart disease was diagnosed*

Year	CHD (n)	Age when congenital heart disease was diagnosed					
		Praenatal	Newborn	Infant	2-7 year	Scool age	X*
1994	48	-	7	10	29	2	2.10
1995	54	-	8	22	20	4	2.32
1996	54	-	13	23	13	5	1.94
1997	53	-	5	24	17	7	2.68
1998	58	-	8	17	21	12	3.22
1999	85	-	21	40	22	2	0.99
Total	352	-	62	136	122	32	2.15

*Average age when CHD was discovered.

By analyzing reason for cardiac examination, we have found that from total 352 children with congenital heart disease, 225 or

63,92% were referral on examination due changed auscultatory findings on heart, and 110 or 31,25% due occurrence of cardiovascular disorders. Reason for cardiac examination in 10 or 2,84% of patients was some of risk factor in anamnesis, in 4 or 1,14% changed chest radiograph, and in 3 or 0,85% pathological electrocardiographically finding.

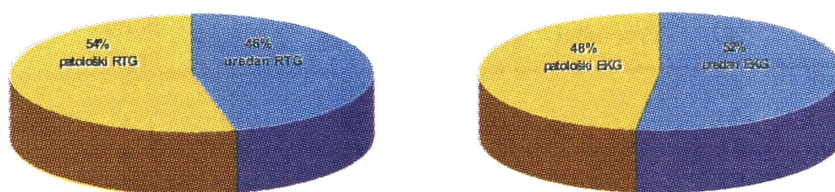
According to analyze of history of disease, we have found cardiovascular disorders in 183 or 51,987% of patients. In more than half or 98 patients, it was about multiply symptomatology. The most frequent symptoms were fatigue (47 or 25,68%) and cyanosis (39 or 21,30%). Information on frequent respiratory infections was found in 19, and growth retardation in 17 patients.

Analyzing anthropometrics parameters during first cardiac examination it has been found that from totally 352 children with CHD, 47 of them or 13,35% have had values of body weight bellow third percentile for age, and in 28 or 7,95% value of body height were below third percentile for age.

Arterial blood pressure during first cardiac examination has been measured in 288 patients. From that in 11 patients (3,82%) values were according to nomogram for blood pressure (4) above 90th percentile. In 6 patients from those 11, ere noticed coarctation of the aorta.

Chest radiograph has been done in 325 patients. Finding was regular in 150 examiners or (46,15%) and pathological in 175 or 53,85% of patients. Electrocardiogram has been done in 335 patients in which 175 or 52,24% was regular, and in 160 or 47,76% pathological (figure 1).

Figure 1. *Chest radiography and electrocardiography in children with congenital heart disease*



Final estimation of morphological and hemodynamical changes is made on basis of echocardiographical examination, by which is, using sequenced segmental analyses in 5 examiners was registered situs inversus, in one right atrial isomerism and in the others 364 situs solitus. In

level of artioventricular connection, by echocardiographcal examination in all examiners was found biventricular harmonious connection. By analyze of ventricul-arterial connection in 13 patients was registered discordant, in 3 patients double outlet by right ventricle and in others concordant connection.

While estimating about needs for further treatment during first examination we have found that 73 (20,74%) patients have needed medicament therapy. Cardiotonic and diuretics were for 41 patients, only diuretics for 3 patients, propranolol for 11 and prostaglandin E11 for 18 patients. From totally 352 patients with CHD, 188 requested only control examination, 61 invasive diagnostic and 157 or 44,60% cardiosurgical treatment. From those patients, 62 were critical CHD with heavy hemodynamical disorders, which requested urgent cardiosurgical intervention. Among them 41 (66,13%) patients were in infant age.

Table 2. *Course of disease in children with congenital heart disease*

Congenital heart disease	Course				Died
	n	Spontaneous recovery	Need operation	Done operation	
Ventrikular septal defect	140	13	5	20	20
Atrijal septal defect	53	-	8	18	3
Patent ductus arteriosus	26	7	4	6	-
Pulmonary stenosis	24	-	1	3	-
Atrioventricular canal	23	-	7	3	13
Tetralogia of Fallot	23	-	1	16	6
Aortic stenosis	22	-	2	6	3
Transposition of great artery	12	-	1	3	8
Coarctatio of the aorta	9	-	-	4	1
Tricuspid atresia	7	-	-	2	7
Hypoplasia of the left ventricle	4	-	-	-	4
Total anomalous pulmonary vein returns	2	-	-	-	2
Fibroclastosis	2	-	-	-	2
Truncus arteriosus	1	-	-	1	-
Epstain' s disease	1	-	-	-	-
Other congenital heart disease	3	-	-	-	-
Total		20	29	82	69

By analyzing implementation of requested therapy we have found that medicament treatment was implemented in accordance to recommendation, except in cases of prostaglandin E1 which was needed in 18 patients but was used only in one patient. Catheterisation of heart

as a diagnostic procedure, independently on surgical intervention, has been done only in 5 examiners, and in preoperative preparation in 68 examiners.

Course of disease according to type of anomaly is shown in table 2. It is noticeable that from 20 patients in which were registered spontaneous recovery, 13 have had ventricular septal defect (10 muscular and 2 perimembranous defect). In two patients after complete invasive diagnostic surgical treatment was contraindicated. In group of patients who are waiting for surgical treatment, in two of them (one with atrioventricular septal defect and one with transposition of great artery) have been done palliative operation.

Cardiosurgical procedure has been done in 82 patients. From them 15 patients have been operated in Sarajevo, and the others were operated in cardiosurgical centers outside of Bosnia and Herzegovina. Two children died in early postoperative period, and one six months after the surgery. In 12 examiners were registered post operative complications: in five temporary disorder of heart rhythm, in four postpericardiotomy pericardial effusion and in three disorder in using type complete atrioventricular block. In patients with complete atrioventricular block, in the same cardio surgical centers where have been done correction CHD it has been done implantation of pace-maker as well.

There were no surgical procedures in infant age among patients. The highest number of patients (50 or 60,97%) had surgical procedure in preschool age, and less number in school age (20 or 24,90%) and infant age (12 or 14,63). The youngest patient had surgical procedure in age of 0,17 and the oldest in age of 15,9 years. The average age in which children have had surgical procedure was $4,81 \pm 3,23$ years with standard deviation 3,95.

From total 352 patients with CHD 69 or 19,6% died. Those 66 died earlier, and 3 after surgical procedure. In neonatal age 32 (8 in first day of life) patients, 27 infants, and 10 died until age of 5 year. Average age of children who died was $0,51 \pm 0,59$. Comparative review of patients who had surgical procedure and those who died in period of examination is shown in figure 2. Number of died was from 5 in 1995 to 17 in 1996, even although in the last three years was equal. Number of those with surgical procedure had shown increase from 4 in 1994 to 27 in 1999.

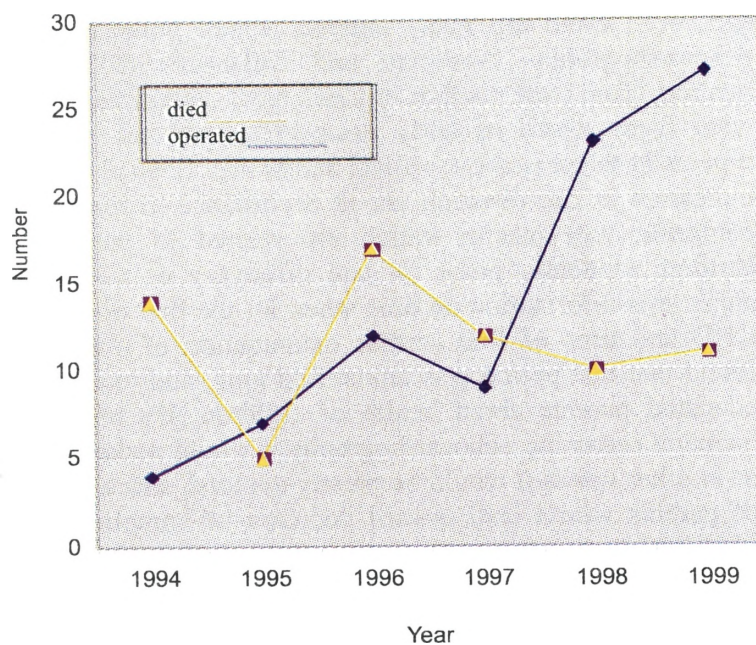


Figure 2. *Comparative review of patients who had surgical procedure and those who died*

Discussion

Diagnostic possibilities for discovering of CHD in Tuzla Canton area until 1992 were very limited, thus children with suspicious on this disease were refereed to different centers out Bosnia and Herzegovina, in order to do diagnostic and eventual treatment. In such conditions great number of CHD, especially those critical, stayed unknown. All these as result had impossibility of its precise registration, and that is a reason that information about epidemiological and clinical indicators practically never existed.

When conditions for non invasive diagnostic of CHD at the Pediatric Clinic in Tuzla and organizing of health care for children in Tuzla Canton area were provided in 1994, it was possible existing of an diagnostic center for this pathology. This has opened possibilities for further continuing determination and following of patients with CHD. Results of diagnostic center during its first six years of existing we have evaluated in our research, what is also presents first step in forming register of CHD in this region.

According to our results there were no CHD diagnosed in fetal age, even there are many studies, which point on success of fetal echocardiography. Andersen and colleagues (5), by applying this method, from total number of CHD have determined 5,5% intrauterine, what from aspect of early discovering presents significant success, especially when is about critical anomalies. Even although, some of age indicators in our research are in accordance to research in developed countries, still results, which are related to preschool and school children evidently point on late discovery of CHD. By analyze of anamnesis information on time when for the first time was suspected on CHD and time of first cardiac examination of our patients, we have found that that period was unjustified long, and mostly as result of non educated parents about health of children. By introducing fetal and neonatal screening echocardiography it would make this period shorter, or in other words it would be mostly avoided, and with health education of parents would lead toward decrease of complication of CHD and neonatal mortality.

Reason for first cardiac examination we have analyzed from anamnesis survey, and gave answer on question: "Why this child was send for cardiac examination?" Received results points that the most frequent reason for cardiac examination were deranged auscultation finding on heart and presence of cardiovascular disorders, while less seldom reason were presence of some potential risk factor and changed electrocardiographically finding. Great frequency of changed auscultatory findings on heart, as one of the reason for cardiac examination is expected since, that is consequence of hemodynamical disorders, presented in all types of CHD. Results on higher frequency of cardiac vascular disorders, as reason for cardiac examination in our research, shows that diagnosis of CHD was relatively late.

Frequency of growth retardation in our patients, which is defined by anthropometrics measurement of body mass and weight, has tendency of increasing in relation on fetal growth. Results point that stoppage in body mass is bigger in relation to stoppage in body height, what is in accordance to results from literature (6,7). Those mentioned authors have proved that level of stoppage in growing depends on type of anomaly, and they are more expressed in category of cyanotic anomaly. Some results (6,8) point that the main cause of stoppage in growing of children with CHD is hypoxemia, which occurred as consequence of hemodynamical disorders. Namely, they have proved that first step are control of growing and development of children with CHD, what would be basis for further studies.

Values of arterial blood pressure were increased in, 82% of patients. All examiners with coarctation of the aorta have had higher blood pressure, what indicates that routine measure of arterial blood

pressure in primary health care is successful method for detection of coarctation of the aorta.

Electrocardiogram (ECG) was changed in almost half of patients. Swenson and colleagues (9) found changes on ECG in 8% patients, to who after auscultation was excluded possibility of existing of CHD. To those patients have been done echocardiography and has been determined atrial septal defect. Those results point that even with advanced and modern and powerful diagnostic procedures, ECG still remains unchangeable diagnostic method in evaluation of children with CHD.

Changed chest radiogram we have found in almost half of patients, what is significantly higher in relation to 28% what is found in literature (9). Chest radiogram still remains as important tool in evaluation and control of children with CHD. Opinions about its routine usage are divided. Namely, from aspect of expenses, chest radiogram is recommended in routine use for all patients, but in trying to avoid eventual harmful effect of radiation, it has been recommended only while indicating cases, or there where is expected its help in determining correct diagnosis.

In estimation of needs for further treatment during first examination we have found that 20,74% of patients have requested medicament therapy, from what 24,66% cases needed usage of prostaglandin E1. Indication for cardiosurgical procedure during first examination was determined in 44,6% patients, what is in accordance to data from literature (3). Because critical CHD urgent surgical intervention was needed in 17,89% of patients what is less in relation to data from literature (10,11). Less number of children with CHD does not mean that they are seldom in our region. We could explain that as possibility that certain number of children with critical CHD died immediately after birth, before cardiac test was performed and which parents never approved autopsy.

By analyzing recommended therapy we have found that prostaglandin E1 was used only in one case from those 18 that were necessary. Similar information we have not found in literature, what was expecting, because it is about medicament palliative treatment, what is part of routine therapy, especially in centers such as ours, without possibilities for urgent catheterisation and cardiosurgical therapy. Results Dinarević and colleagues (12) point on successful usage of prostaglandin in neonatal cardiology in our country.

Spontaneous recovery was registered in 14,29% of our patients with ventricular septal defect. According to literature (1) it was registered in 15% to 50% of children with ventricular septal defect. Such great variation depends mostly on diagnostically possibilities. In region where is possible detection of small muscular ventricular septal

defect, frequency of spontaneous recovery is higher, and there where CHD was diagnosed in older age there is higher number of children with this anomaly will stayed as unknown.

According to our results, it is necessary to upgrade early diagnostic of CHD in our country. Lateness in diagnostic is expressed with information that in our 2 patients with CHD was discovered late, when is already developed heavy lung hypertension where surgical intervention was contraindicated.

From those 150 patients who needed cardiac surgical intervention, the same was done only in 51,57%. With critical anomaly were 63 patients, and surgical intervention have been done in only 3 or 4,76% cases. Fact that there were no surgeries in newborn age is very disturbing. According to results of Bosi and colleagues (13) in newborn age have been 12% of surgical interventions among all diagnosed CHD, from what only in one third part have been done palliative procedure.

Our research which is related on treatment of patients with CHD, started from usage of prostaglandin E1, need for urgent or diagnostic catheterisation, early palliative procedure and finally corrective cardiac surgical procedure point on its insufficiency. Justifying with high price of treatment is also unsaid, when we have fact that many other undeveloped countries, by investing money in development of cardiac surgery more and more reaching world standards for advanced treatment of CHD. Also, it is necessary to make relation between cost of treatment and advantages which will reach in that case as: decrease of mortality of children, better quality of life, and less number of chronic ill persons in older age. Garson and colleagues (14) analyzing cost of treatment of CHD from birth to age of 21 in the USA have found that cost is in average 59,877US\$, what makes it on 5th place in relation to some other chronic diseases (hemophilia, cystic fibrosis, malignant diseases, muscular dystrophy). All this point on necessity of producing systematic and unique program of medical treatment of children with CHD, starting with control of epidemiological parameters, permanent monitoring, diagnostically possibilities up to providing necessary therapeutic measures. By individual consideration of justification and cost it is evident that many needed measures could fit into current financial situation in our country.

Results on mortality of patients with CHD show that the biggest number of them died in age of newborn, what is in accordance to other studies (13,15). Comparative review of performed cardiac surgical procedures and mortality in accordance to age, point that the highest mortality is in age group for which cardiac surgery in our region is the least available.

Considering epidemiological and clinical indicators of CHD in Tuzla Canton area it is evident that this is a public health problem,

whose resolving requests coordinated work of all segment of public health care of children as well as engage of broader social community. Our research, which presents beginning of total following and control of children with congenital heart disease, gave great benefit on focusing problem related to its diagnostic and treatment in our region.

Apstrakt

Cilj istraživanja bio je procjena mogućnosti dijagnostike i liječenja urođenih anomalija srca (UAS) u našim uslovima. Analizirana je medicinska dokumentacija od 352 djece sa ovim oboljenjem u šestogodišnjem periodu na području Tuzlanskog kantona. Srednja vrijednost životne dobi, u kojoj je UAS dijagnostikovana, bila je $2,15 \pm 2,28$ godina. Tokom prvog kardiološkog pregleda, 51,98% djece je imalo prisutne simptome bolesti srca. Zastoj u tjelesnoj masi i tjelesnom rastu u postnatalnom dobu nađen je u 13,35%. Elektrokardiografske promjene su registrovane u 47,76% a izmijenjen radiogram srca i pluća u 53,85% ispitanika. Medikamentozno liječenje je bilo potrebno u 20,74%, a kardiohirurški zahvat u 40,60% ispitanika. Hitno kardiohirurško zbrinjavanje je bilo neophodno u 62 ispitanika sa kritičnom UAS. Kardiohirurški zahvat je urađen u 23,29% ispitanika, a prosječna dob u kojoj je urađen iznosila je $4,81 \pm 3,23$. Smrtnost djece sa UAS je relativno visoka i iznosi 19,60%. Najveći broj njih (95,65%) umrlo je prije, a samo 4,35% nakon kardiohirurškog zahvata. Prosječna dob u kojoj su djeca umirala iznosila je $0,51 \pm 0,59$ godina. Analiza kliničkih pokazatelja UAS u djece na području Tuzlanskog kantona ukazuje na to da se radi o zdravstvenoj problematici djece, koja zbog teške kliničke slike, velikog učestća u perinatalnoj i dojenačkoj smrtnosti, usporenja rasta i razvoja s progresijom u hroničnu kardiopatiju, zahtijeva hitne mjere savremenog organizovanja njihove dijagnostike i liječenja.

Ključne riječi: *urođene anomalije srca, djeca, Tuzlanski kanton.*

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