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The Incidence of Congenital Heart Disease: Previous Findings and Perspectives

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SUMMARY
Congenital heart defects (CHD) are the most common of all congenital anomalies, and represent a significant global health problem. Involvement of medical professionals of different profiles has led to drastic changes in survival and quality of life of children with CHD. The motivation for the implementation of the first large population studies on this subject was not only to obtain answers to the question on the level of incidence of CHD, but the harmonization of criteria and protocols for monitoring and treatment of certain defects as well as the planning of medical staff dealing with children with CHD. Data on the incidence varies from 4-10/1000 live births. Fetal echocardiography can have potential impact on decrease of CHD incidence. The increase in incidence may be due to the possibility that children with CHD will grow up and have offsprings. Owing to the progress that has been made, an increasing number of patients experiences adulthood, creating an entirely new and growing population of patients: patients with "adult" CHD. Survivors suffer morbidity resulting from their circulatory abnormalities as well as from medical and surgical therapies they have been subjected to. Application of the achievements of human genome projects will in time lead to drastic changes in the approach to the patients with CHD. Until the time when it is possible, the goal will be further improvement of the existing system of service: networking in a unique, multicenter clinical registry of patients with CHD, as well as upgrading of technical and non-technical conditions for the treatment of patients with CHD. We are in an unprecedented time of change, but are actually at the end of the beginning of making pediatric cardiac care a highly reliable institution.
Keywords: congenital heart disease; incidence; echocardiography

INTRODUCTION

Congenital heart defects (CHD) are the most common of all congenital anomalies (CHD accounts for 28% of congenital anomalies) [1], and represent a significant global health problem. Heart morphogenesis is a complex process whose disturbance can produce a range of CHD from harmless to fatal ones. Interest in their detection and treatment has evolved from an initial position of several hundred years ago when they were considered the result of unfortunate mistakes of nature, mysterious accidents, a natural development with a short history and lethal consequences [2].

CHD is the leading and most common cause of death in the first year of life compared with other birth defects [3]. Data about the incidence varies from 4-10/1000 live births. If two less complex and most often CHD (bicuspid aortic valve and prolapse of the mitral valve) were taken into account, the total number of CHD would rise to 2% of live births. Although the percentage of CHD incidence does not look impressive, do not lose sight of the fact that the treatment and diagnosis of CHD is very expensive, in some cases, CHD requires lifelong follow-up of patients, and that a number of CHD to some extent may lead to restrictions of physical abilities and disabilities.

Consolidated enthusiasm of specialists in various medical and public services throughout the world has led to a drastic change in survival and quality of life of children with CHD. If 89.3% of children with transposition of the great arteries died in the first year of life only 50 years ago [4], 85% of children with the same CHD would survive 20 years after surgery [5]. In that way, small "blue children" will get a better quality, and even completely normal life. Pediatric Cardiology and Cardiosurgery, whose principal object is dealing with CHD, have become a leader in quality of improvement and patient safety. These medical disciplines are credited for initiating the systemic change in the organizational culture of health, successful cohesion of recognized individuals of different areas of medicine, and a high level of adoption of new knowledge and the implementation of technological innovations [6].

Data of early studies on CHD were discoveries that had been made *post mortem*. William Harvey published *De Motu Cordis* in 1628 [7]. Niels Stenson [8] described with an amazing precision hemodynamic consequences of the anatomical malformations known as Tetralogia Fallot in 1671. From that time until the early 20th century, studies were individual and unorganized with medical reports on the level of anatomical phenomenon. The focus of the research was directed towards CHD owing to another disease, rheumatic fever (RF), which was raging around the world. In 1835, Bouillard noticed the relationship between the RF

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and pathological findings of the heart. Someone finally recognized that the inflammation of pericardium and heart valves was usually of rheumatic origin. Devastating impact of RF on the heart and its structures has made the studies of these acquired heart defects focused on the problems related to CHD. Helen Taussig, often called the mother of Pediatric Cardiology, began her career on one of the specialized clinics for RF. The greatest achievement of her engagement in the field of CHD was reflected in publishing the classification of CHD in 1932. CHD has become basic object of interest of pediatric cardiology after its triumph over RF.

Medical officials gave full financial support for the largest enthusiasts in the field of pediatrics and cardiac surgery to achieve their professional ideas. The first step was to integrate all previous knowledge. The following steps were very courageous: Forssman checked catheter function on his own heart in Boston in August 1938. Not much later, Gross [9] performed the first arterial duct ligation in 1939. In that way, CHD was transformed from an interesting entity to disease that can be treated. However, the initial enthusiasm was wearing thin at times because of high postoperative mortality. At the very beginnings of pediatric cardiac surgery, mortality was as high as 50% [10]. Postoperative mortality rate has dropped significantly due to a comprehensive knowledge of pulmonary vascular bed, and technological advance: the availability of NO_2 in the control of pulmonary hypertension and extracorporeal membrane oxygenation to support temporarily disabled myocardium.

From the very beginning, technological innovation was at the service of progress of pediatric cardiology. Introduction of echocardiography in the standard diagnostic procedure is one of the most revolutionary moments in its development because it provided a pictorial recording of the heart action, and understanding of the phenomenon of flow and hemodynamic consequences that are carried by some entities [11].

INCIDENCE OF CONGENITAL HEART DISEASE AS A SUBJECT OF STUDYING

The incidence of any disease is the essential information for the long-term planning of health care. In addition to the need for definition and classification of the anatomical heart defects, determination of the CHD frequency is fully recognized as actual. Frequency of CHD has been and will be the subject of research in countries in which the development of the health system is based on epidemiological indicators. Recognizing the evolutivity of idea of CHD from the anatomical curiosities to the formation of database on the number of children with CHD, we are able to follow the growth of awareness on the study of the incidence of CHD. The results of studies published in literature and our own research should be applied for reviewing the rational use of regional human and infrastructural resources.

Important studies of congenital heart disease incidence

Exploring the extent of problem in its all dimensions (medical, social, ethical, material) was a primary motivation for the implementation of the first large-scale population studies on this subject. These studies gave the answer to the question about the level of incidence of CHD. They also helped in streamlining the criteria and protocols for monitoring and treatment of certain defects as well as planning of staff dealing with CHD children.

A true pioneer in determining the CHD frequency was Swede Calgren [12], who calculated that the incidence of CHD was 4/1,000 live births through the period 1941–1950. First true population studies were conducted in the late 1950s by Sheila Mitchell et al. [13], who found that the incidence of CHD was 7.67/1,000 live births. Dickinson et al. [14] continued in determination of CHD frequency in Liverpool from 1960 to 1969. They came up with a number of 3.76/1,000 live births. Conducting tests in Denmark, Laursen [15] found out the highest incidence of CHD at that time, obtaining the prevalence of CHD of 4.3/1,000 live births. It should be noted that the clinical diagnosis alone was involved in all these studies in 30% to 48%.

Nadas et al. [16] used study of CHD incidence for further investigation of the natural evolution of some CHD. Keith et al. [17] made the first registry of heart diseases in Toronto and were among the first who identified a high mortality rate among children born with CHD. Although, from the present point of view and the amount of accumulated knowledge, all these conclusions are a bit naive, but being placed in a cognitive framework, they caused immeasurable interest and triggered an avalanche of new research in all aspects of CHD.

One of the most important and more comprehensive study ever conducted by the New England Regional Infant Cardiac Program (NERICP), was led by Fyler et al. [18]. The results of this study have not lost their significance over time. On the contrary, they have continued to serve as the basis of application of statistical methods for predicting the incidence of CHD in the USA. This long-term (from 1969 to 1977) and comprehensive study (1,528,686 children) with an incidence of 2.03/1,000 live births, has served as a basis for making an important conclusion: "... that the incidence of CHD is remarkably constant throughout the world for a number of years. There is no reason to expect any change soon. Not only is the overall CHD incidence relatively predictable, but the relative frequency of different CHD varies slightly (except subpulmonary VSD in Asia). If, says the author, there are reports of variations in incidence, they are usually attributed to weakness in the classification and inadequate data collection of the number of patients with CHD".

Very well defined study, conducted in Metropolitan Atlanta, collected data from 1968 to 1997 to determine the incidence of CHD and thereby pointed that the occurrence of major cardiac defects was almost tripled in this period. During this period, there were 5813 cases of CHD on a

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sample of 937,195 children with an overall incidence of 6.2/1000 live births [19].

Worth mentioning is the Baltimore-Washington Infant Study [20], a CHD regional epidemiological study conducted in the period from 1981 to 1989, with the values of incidence of 3.7/1000 live births.

Hoffman and Rudolph [21] conducted their first research with the aim to determine the frequency of CHD from 1959 to 1966, when he came to a figure of 4.11/1,000 live births. For them, determination of the CHD prevalence remained constant object of interest. Hoffman and Christiansen [22], in 1978, came to the prevalence of 8.56/1,000 live births. In 2002, Hoffman [23] started the trial called "The incidence of congenital heart disease", which was direct evidence that determination of the incidence of CHD was and has remained a challenge. This time, he clearly defined the objectives: to identify the reasons for diversity of published incidence of CHD and more precise determination of the incidence of CHD in the selected sample, as well as determination of the incidence of specific, significant CHD. Defined goals were the product of a major research experience in this field and understandable curiosity of that remarkable pediatric cardiologist. He reviewed 62 studies published from 1955 to 2002 to get insight into all previous studies on this important issue. Incidence disparity in different studies varied from 4/1000 to 50/1000 live births. Taking into account all the variations, Hoffman came to the conclusion: "...that there is no significant difference in incidence in different countries at different moments of time". He added that the lower incidence was attributable to the period when the pediatric cardiologists were less trained and when cardiosurgery was not developed enough. He suggested, like many, that the progress of echocardiography and color flow techniques enabled diagnosis of lesions that were clinically asymptomatic and harmless, even in the absence of murmur.

Its contribution to the study of CHD incidence was reflected in a study conducted in Montenegro, covering the period from 1995 to 2003 and 88,098 live births. During that period, there were 1,004 children with CHD, and the incidence was 8.8/1,000 births [24, 25, 26].

Factors influencing the incidence of congenital heart disease

Great hopes were placed in fetal echocardiography as one of the possible ways to reduce the birth of children with CHD. Various studies differently evaluated the influence of fetal echocardiography on incidence of CHD. Special study of CHD incidence in Atlanta that was carried out from 1990 to 1994 showed rapid increase in prenatal diagnosis of CHD of which even 12% were made *in utero*. However, not encouraging is the fact that only 6% of cases of transposition of great arteries (TGA) are diagnosed prenatally and 8% of women terminate the pregnancy after being informed on the presence of CHD [27]. The Boston study based on hospital data revealed different experience: 22% of pregnancies during which CHD was diagnosed *in utero* were terminated. The influence of selective abortion on the incidence of CHD varies from country to country, especially for certain defects. For example, the termination of pregnancy in case of TGA and hypoplastic left heart varies in different parts of France from 21% to 53%. Finally, we cannot predict the extent of decline of incidence due to therapeutic abortion because of the complex cardiac lesions which make a very low percentage of CHD. Some studies reported that their impact was negligible [28], while other studies suggested that therapeutic abortions substantially reduced the incidence of CHD [29].

Wren and Richards decided to use mathematical methods in order to approach this problem. They concluded that the detection of CHD using the 4-chamber section during routine obstetric examination and screening of pregnant women's serum for Down's syndrome indicated low effect on reduction of workload in the pediatric cardiology and cardiac surgery [30]. They substantiated it by the following calculation: assuming detectability of CHD in 20% and abortions in 67% of cases, the CHD incidence per 1000 live births would be decreased by 2%, the mortality rate of children with CHD would fall by 5% and the activities of pediatric cardiac surgery by 3%. Screening of pregnant women for Down's syndrome with 75% of comprehensiveness, 60% of detectability and termination of all pregnancies with this defect would reduce birth of children with Down's syndrome by 45%, the incidence of live births with CHD by 3.5% and cardiosurgery scope of work by 2.6%.

In our region, the University Children's Hospital and Clinic of Gynecology and Obstetrics in Belgrade have the greatest experiences in prenatal diagnosis of CHD. During 25 years of follow-up (from 1991 to 2013), 515 fetuses with CHD were detected in the prenatal period with sensitivity of 95.9% and specificity of 99.8%. In the early years of follow-up, the percentage of terminations of pregnancies for CHD was up to 60%, while in the past few years, that percentage dropped to 8% owing to advances in cardiac surgery [31].

In addition, prolonged life of patients with CHD and a chance to have offsprings may have significant effect on the incidence of CHD. Because of the parents (especially mothers) with CHD, there is also higher incidence of children with CHD [32, 33, 34]. Taking into account this fact, we may reasonably believe that the incidence of CHD will gradually grow in future. Carter predicted that the incidence would be doubled after seven generations [35]. We are aware of the fact that the advances have been achieved in other areas of medicine as well, so the women having diabetes mellitus can nowadays give birth to children with much less risk, which further increases the possibility of rise of CHD incidence. Today, many women opt for motherhood much later, there are more and more children born from *in vitro* fertilization, and the environmental factors have become more aggressive. All the above listed reasons may be potential causes of higher incidence of CHD.

Current overview of pediatric cardiology, cardiac surgery and congenital heart disease incidence studies

Advances in pediatric cardiology have created armies of millions of adults who had had CHD and had been operated in childhood. Now, for the first time, the number of adults with CHD is equal to the number of children with this problem. There are great variations in the complexity of CHD and for almost all of them there are well-established methods of correction or palliation, which have led to improved survival of patients with CHD. An example that illustrates best previous statement was a study conducted in Quebec, Canada. Comparing postoperative mortality of children with a significant CHD during the period 1987-1988 and 2004-2005 revealed that the number of deaths decreased by 67% [36]. At the University Hospital of Oslo, early mortality within 30 days after surgery was reduced from 10% in 1991 to below 1% in 2010 [37]. Thanks to the progress that has been made, there has been a growing number of patients with CHD, creating an entirely new and ever-increasing population of patients with "adult" CHD (ACHD). The prevalence of ACHD was estimated as 4/1000 after the age of 18 years in the study conducted in Quebec [36].

Unfortunately, early success of cardiac surgery in severe CHD did not guarantee good results in the long run. As operated patient grows, so he/she is faced with late complications. The survivors suffer from morbidity resulting from their circulatory abnormalities as well as from medical and surgical therapies they have undergone. Residues and sequelae of surgical procedures on the heart have become an important issue that affects their physical and mental development, psychosocial status and overall functioning. For example, some CHD are associated with the chronic heart failure, which is associated with depression, and which further interferes with the quality of life. The awareness of possible frequent hospitalizations, arrhythmias, and potential fatal outcome is always a fact that reduces the quality of life [38]. The struggle to maintain a certain level of quality of life is a new challenge for the health system operation. It is time to redefine traditional boundaries in providing health care.

Unfortunately, this is not the end of problems of adults with CHD. A moment of transition from pediatric to cardiologist for adults is considered a critical milestone in the implementation of health monitoring. Knowledge that there is no organized system of monitoring patients with ACHD is terrifying. All the aforementioned suggest that the level of health services provided to adults with acquired heart defects and children with CHD is much better than for the patients with ACHD. Cardiologists for adult patients have little experience in dealing with patients with complex CHD and do not understand the potential complications that can arise. The frequency of presentation of these patients to tertiary level institutions, with complications that could have been avoided if they had been monitored adequately, confirms the above statement.

The efforts of regional cardiac centers, the University Children's Hospital and the Institute for Mother and Child Healthcare of Serbia "Dr. Vukan Cupic" in Belgrade, to provide cardiosurgical support and monitoring of the clinical condition for adult patients with CHD, are encouraging.

PERSPECTIVES

Insufficiently precise and reliable record-keeping, which leads to underestimated number of patients with CHD, are the weaknesses observed during surveys of the CHD incidence. Therefore, multicenter clinical registries and administrative databases are just step away from networking in a single system. On the basis of the uniform criteria, this system will consolidate data of all patients with CHD in a particular geographical region and around the world. This is the only reliable way to overcome historical barriers in implementation of pediatric studies of CHD incidence and incidence of all other diseases. Relative scarcity and large heterogeneity of CHD, limited research infrastructure, costs, and ethical issues that arise in large trials were the limiting factors for achieving higher reliability of data on CHD incidence. Naturally, the data obtained in this way will enable the implementation of already defined regulatory requirements, which will specify the number of certified centers for dealing with pediatric cardiac surgery, and the minimum number of qualified surgeons in each center who will be engaged only in pediatric cardiac surgery. Nordic countries have adopted a standard that one pediatric cardiosurgery center is sufficient for the population of 5 million people, and the team has to consist of four cardiac surgeons practicing only pediatric heart surgery [39].

Solving problem of CHD in future lies in the implementation of knowledge that will be acquired within the human genome project. Great hope is to be taken in discoveries that will create the conditions for the realization of every doctor's dream: to stop the development of disease by preventive actions. Since it is still a far away future, the attention of medical professionals engaged in pediatric cardiac care will be focused on the outcome of treatment that guarantees a high quality of life. Quality of life is defined as an ability of the sick person to function in situational context, and get a personal satisfaction from the actual events [40]. Therefore, it is necessary to improve the health care system by making it available, in human and financial terms, to adults with CHD in order to maintain their functional status. Their functional status has been achieved with a lot of professional effort and huge financial resources. Improvement of treatment of the chronic heart failure, higher safety of cardio-pulmonary bypass application, better treatment of arrhythmias, better control of the pulmonary vascular resistance are the tasks requiring continuous action.

Special attention will be paid to further reduction of postoperative mortality of patients with CHD because there are still institutional differences in its rate. First of all, the attention will be paid to events that can be prevented and controlled during treatment and post-operative care. These are the events of technical and non-technical nature, which lead to failures resulting in patient death. The risks of technical origin will be reduced to negligible. Human factors will certainly become the focus of further develop-

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ment, i.e., decision-making that have crucial impact on the outcome of treatment. Specifically, 10 pediatric cardiologists have made 1000 decisions within 7 days, out of which only 3% were based on the results of large studies, and all others on the experience and results of small studies [41].

CONCLUSION

Every state has an obligation to its own population to monitor the incidence of major diseases that significantly affect the individuals, the family and wider community. The analysis of the CHD incidence is very important for each country. If there were no respective registry of CHD patients, the health system would be deprived of important information related to their population. Certainly, the data obtained from different countries, given their considerable uniformity, are good enough to serve the purpose of planning the quality of healthcare services. Well connected healthcare system with a clear assignment of responsibilities of medical professionals in providing health services for children with CHD, makes the health system safer, more accessible and of higher quality.

There is a possibility of improving the existing system to provide cardiac services to children with CHD at the regional level. The patterns that have been applied by Nordic countries and led to decrease of postoperative mortality rates, can serve as a model of regional institutional connection of medical institutions of neighboring countries that do not have children's cardiovascular surgery. Regionalization and rationalization of human and infrastructure resources is a model that may lead to an improved knowledge of CHD incidence, better quality and systematized treatment of CHD, and probably lead to information about possible causes of CHD.

The goal of all activities in pediatric cardiology and cardiac surgery is a decrease of the incidence of CHD and improved quality of life for children with CHD. Far behind are the times when the initiatives in this direction rested on one man. Now, there is quite complex composition of people involved in these activities, and the outcome now depends on collaboration and team work in this complex group of medical professionals. Geneticists and molecular biologists as representatives of the basic sciences that have the task to eliminate persistent frustration of pediatric cardiologists regarding the origin of CHD have joined the standard team. They are all convinced that near future will bring a lot of excitement in pediatric cardiology for patients they are dealing with now and for patients who are not born yet.

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Инциденција урођених срчаних мана: досадашња сазнања и перспективе

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КРАТАК САДРЖАЈ

Урођене срчане мане (УСМ) су најчешће од свих урођених аномалија и значајан су здравствени проблем у читавом свету. Ангажман медицинских професионалаца различитих профила довео је до драстичне промене у преживљавању и квалитету живота деце са УСМ. Мотив за извођење првих великих популационих студија на ову тему био је добијање одговора на питање о висини инциденције УСМ, али и усаглашавање критеријума и протокола за клиничко праћење и лечење болесника са УСМ, као и планирање кадра који се бави лечењем ове деце. Подаци говоре да је инциденција УСМ у распону 4–10 оболелих на 1.000 живорођене деце. На смањење инциденције УСМ могући утицај може имати фетална ехокардиографија. До повећања инциденције може доћи због могућности да деца са УСМ одрасту и сама стварају потомство, као и због других напредака у медицини који су омогућили рађање детета и поред постојања значајних здравствених проблема родитеља. Захваљујући прогресу који је остварен, све већи број болесника доживи одрасло доба, стварајући потпуно нову и стално растућу популацију са тзв. одраслом УСМ. Преживели пате од морбидитета који је резултат поремећаја њихове циркулације, као и од последица конзервативног и хируршког лечења којем су подвргавани. Примена достигнућа из Пројекта хуманог генома довешће до драстичних промена у приступу болеснику са УСМ. До тренутка када то буде могуће радиће се на даљем усавршавању постојећег система пружања услуга: умрежавању у јединствен систем мултицентричних клиничких регистара болесника са УСМ и унапређењу техничких и нетехничких услова за лечење ових болесника. Јасно је да се налазимо у времену промена без преседана, али смо заправо на крају почетка изградње институције педијатријске кардиолошке неге високе поузданости.

Кључне речи: урођене срчане мане; инциденција; ехокардиографија

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