Віддалені результати стану здоров'я дітей, що народились з вродженими вадами серця у львівській обласній клінічній лікарні, Львів, Україна

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Вступ. Вроджені аномалії є найпоширенішими медико-соціальними проблемами та основною причиною втрати плода та інвалідності дітей у світі. В Україні щороку народжується близько 5 тисяч дітей з вродженими вадами серця (ВВС). На сьогоднішній день інформація про майбутнє пацієнтів, які отримали медичну допомогу з ВВС, не систематизована і часто залишається недоступною.

Мета. Провести опитування жінок, які народили дітей з ВВС, та проаналізувати зібрану інформацію про віддалені результати здоров'я цих пацієнтів.

Методи дослідження. Були відібрані історії хвороби 170 дітей, які народилися з ВВС у Львівській обласній клінічній лікарні, Львів, Україна, у 2011-2015 роках. Критерієм включення до групи був клінічний діагноз «Вроджені вади розвитку системи кровообігу» на основі Міжнародної класифікації хвороб-10 (Q20 - Q28).

Результати. Згідно з даними анамнезу, що проаналізовані у нашому дослідженні, серед 170 дітей, народжених з ВВС, у 67 пацієнтів (39,4%) вади серця були виявлені на пренатальному етапі. Невідкладне оперативне втручання виконано 32 (18,8%) дітям. Приблизно 1/3 (27,6% – 47 випадків) жінок, які народили дітей з ВВС, повідомили в анкетах, що їхні діти в цілому здорові.

Висновки. Отримані дані про відділені результати ВВС у новонароджених Львівської області в Україні дозволять створити базу даних для майбутніх досліджень. Ці результати можуть допомогти розширити визначення груп ризику пренатальної патології та покращити сучасні підходи до медико-генетичного консультування та спеціалізованої допомоги вагітним жінкам із ризиком розвитку вроджених вад серця у плода.

Ключові слова: вроджені вади серця, новонароджені діти, фолієва кислота.

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Outcomes of congenital heart disease in newborns at Lviv Regional Clinical Hospital, Lviv, Ukraine

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Introduction. Congenital abnormalities are the most common medical and social problems and the primary cause of fetal loss and children's disability in the world. In Ukraine, about 5 thousand children with congenital heart disease (CHD) are born every year. Up to date, information on the future of patients who received medical care for CHD was not systematized and often remains unavailable.

The **aim** of the study: To conduct a survey of women who gave birth to children with CHD and collect information about distant health outcomes.

Methods. Medical history files of 170 children born with CHD in Lviv Regional Clinical Hospital, Lviv, Ukraine, between 2011-2015 were selected. The criterion for inclusion in the group was a clinical diagnosis of ''Congenital malformations of the circulatory system'' based on the International Classification of Diseases-10 (Q20 - Q28).

Results. According to the medical history files analysed in our study, among 170 children born with CHD, 67 patients (39.4%) had heart defects detected at the prenatal stage. Urgent surgical intervention was performed in 32 (18.8%) children. Approximately 1/3 (27.6% – 47 cases) of women who gave birth to children with CHD informed in the questionnaires that their children were generally healthy.

Conclusions. Collected data about the outcomes of CHD in newborns of Lviv region in Ukraine will allow creating a database for future research. These results could help expand the definition of prenatal pathology risk groups and improve current approaches in genetic counselling and specialized care for pregnant women at risk of fetal congenital heart disease.

Keywords: congenital heart disease, newborns, folic acid.

Introduction

Congenital abnormalities are the most common medical and social problems and a primary cause of fetal loss and children's disability in the world, and particularly in Ukraine [5, 10]. Recently, the prevalence of congenital heart defects (CHD) has dramatically increased among the known congenital abnormalities in general. Thus, the incidence of CHD in the world is 9 per 1000 neonates, and 8 per 1000 neonates in Europe, which is 4 times higher than the number of neural tube defects and 6 times higher than the frequency of chromosomal anomalies [9, 11]. In Ukraine, about 5 thousand children with CHD are born every year [2, 4, 5]. More than 90 types of CHD have been described either alone or in combination with other congenital abnormalities [16, 17].

Early detection, topical diagnostics, and evaluation of the hemodynamic disturbance allow choosing an adequate treatment method and the best time for the surgery [8]. The current progress in the field of cardiovascular surgery contributes to better CHD treatment outcomes in children, particularly neonates. About 75% of patients born with potentially "fatal" heart disease could be saved now with the methods of cardiac surgery developed in the last decade. Novel approaches of radical correction and the timelines of their application greatly improve the results of surgical interventions, reduce childhood and overall mortality, as well as the level of disability [7].

Situations that arise in the diagnostics of congenital disorders are usually so complex that they do not have an unambiguous solution. The general and economic state of society, the social security of its members, as well as the state of health, reproductive age, cultural and religious views of parents, and many other aspects define approaches to abortion or treatment of this pathology [14, 15, 18].

An important mission of medical genetic counselling is the optimization of diagnostics, prognosis and timely treatment of congenital cardiovascular malformations. Taking into consideration the increasing anthropogenic impact on the environment, it is critically important to evaluate each pregnancy for potential risk of birth defects and hereditary pathology [1, 3, 12]. Although CHD could be well diagnosed using non-invasive methods of prenatal diagnostics, late manifestations of this pathology often occur. Ultrasound screening of pregnant women helps to evaluate the intrauterine foetus at the time when most congenital defects can be visualized and identified as isolated or multiple. It is possible to detect the early manifestation of defects and make a decision about pregnancy management [6, 8, 19].

Although the survival of CHD patients in Ukraine has improved significantly due to advancements in cardiac surgery [7], the information regarding the future of patients who received medical care for CHD is still not systematized and often remains unavailable. Correspondence surveys, including in the form of questionnaires for CHD patients and their families, appear to be a very effective way to obtain missing information about medical outcomes for these patients [6, 21]. Taking into consideration that most congenital abnormalities, including CHD, can be prevented by the preconception use of folic acid (FA) [13, 18, 20], we included a question about diet supplementation with FA in our survey for women who gave birth to children with CHD. Our methodology of collecting medical information from families with children suffering from CHD could be successfully applied by medical geneticists, paediatric cardiologists, paediatricians, and cardiovascular scientists. Moreover, our newly developed survey methodology could be further developed to study the inborn abnormalities of other organs and organ systems. Thus, the aim of study was to perform a survey of women who gave birth to children with CHD and collect information about distant health outcomes.

Methods

Medical history files of 170 children born with CHD in Lviv Regional Clinical Hospital, Lviv, Ukraine between 2011-2015 were selected. The criterion for inclusion in the group was a clinical diagnosis of ''Congenital malformations of the circulatory system'' based on the International Classification of Diseases-10 (Q20 - Q28).

We developed a survey in the form of a questionary that followed up the development and health information of children born with CHD. The first part of the questionary related to pregnancy, such as pregnancy complications, use of medications, and delivery. We also introduced questions related to the consumption of

FA before and during pregnancy, including its dose and supplementation regimen. To confirm the prophylactic effect of FA on congenital fetal heart malformations, we collected and analysed data regarding diet supplementation with FA in doses of 1, 2, 4 or 5 mg daily during 2 to 3 months before conception and the first three months of pregnancy. The second part of the survey included questions about the child, such as the presence of a congenital abnormality, health condition at birth, and the subsequent disease progression. The questionnaire was mailed to families, the answers were recorded, and the data were subjected to statistical analysis using Statistica and Excel software.

Results

Out of 170 questionnaires sent to mothers of children with CHD, 140 (82.4%) were returned and 30 (17.6%) addressees failed to respond.

According to the medical history files analysed in our study, among 170 children born with CHD, 67 patients (39.4%) had heart defects detected at the prenatal stage. According to the generally accepted classification of CHD based on changes in hemodynamics [14], the following anatomical anomalies were diagnosed prenatally: valve or great vessel obstruction defects – 5 (2.9%), arteriovenous blood loss (left-right shunt, "pale") – 20 (11.8%), cyanotic defects with reduced or normal blood flow – 32 (18.8%), cyanotic defects with increased blood flow in a pulmonary circulation – 10 (5.9%).

Overall, out of 170 cases, heart defects with arteriovenous blood loss (left-right shunt, "pale") were predominant and found in 102 (60%) patients. Heart disease with reduced or normal blood flow was diagnosed in 49 cases (28.8%), including heart defects with reduced or normal blood flow in a pulmonary circuit of blood circulation accompanied by cyanosis in 44 cases (25.9%), and heart defects with cyanosis and increased blood flow in the pulmonary circuit of blood circulation (5 cases or 2.9%). Heart defects with complications related to the valve or large vessel obstruction development were less common (14 cases or 8.2%). Exceptionally rare heart defects were diagnosed only in 5 studied cases (2.9%).

Urgent surgical intervention was performed in 32 (18.8%) children, including 6 (3.5%) patients with aortic coarctation, 8 (4.7%) with pulmonary artery stenosis, and 18 (10.6%) with hypoplasia of left heart chambers. Surgical correction during the first month of life was necessary for 11 (6.5%) children born with a tetralogy of Fallot, and in 1 case (0.6%) with abnormal drainage of pulmonary veins.

In response to questions concerning the course of pregnancy, 22 (12.9%) women answered that they had had no complications during pregnancy. Early gestosis was noted in 12 (7.1%) women, with preeclampsia diagnosed in 5 cases (2.9%). Potential miscarriage was noticed in 14 cases (8.2%), the risk of preterm labour was detected in 17 women (10.0%) and 30 (17.6%) women failed to respond.

Importantly, approximately 1/4 of women who gave birth to children with CHD (43 cases – 25.3%) suffered from viral respiratory infection during pregnancy. One woman (0.6%) was infected with measles, and one with scarlet fever in the first trimester of pregnancy.

As for the medications taken during pregnancy, 31 (18.2%) respondents used antispasmodics, 14 (8.2%) progesterone, and 7 (4.1%) women received drugs for the treatment of diabetes mellitus.

Information about health outcomes of children born with CHD collected based on the results of the survey of their mothers is shown in Table 1.

Among 32 children who needed urgent surgical intervention, surgical correction of CHD was performed in 12 (7.1%) patients. The mortality analysis revealed that hospital mortality was observed in 22 (12.9%) cases. Particularly, 3 (1.8%) children died on the first day in the delivery unit, 8 (4.7%) during the first week after birth, 11 (6.5%) during their first month of life, including 2 patients (1.2%) who did not survive after surgical correction. Due to a critical health status that preceded lethality, 10 (5.9%) children were not operated on. Disability care is currently received by 51 (30.0%) children. Approximately 1/3 (27.6% – 47 cases) of women who gave birth to children with CHD informed in the questionnaires that their children were generally healthy.

Table 1

Follow up evaluation of the health status of children born with congenital heart disease according to the questionnaire

Answers	Number	
	Cases	%
Operated once	12	7.1
Operated twice	3	1.8
Not operated	10	5.9
Hospital mortality up to 7 days	11	6.5
Hospital mortality up to 1 month	11	6.5
Disability	51	30.0
Dispensary monitoring	71	41.8
Generally healthy	47	27.6

Table 2

Preventive consumption of folic acid (FA) by women who gave birth to children with CHD

Diet supplementation with FA	Number of cases	
	Cases	%
Supplementation		
Did not consume	49	28.8
Preconceptionally	18	10.6
Consumed before and during pregnancy	39	22.9
During pregnancy overall	34	20.0
Unknown	30	17.6
Dose		
A daily dose of 0.1 mg	24	14.1
A daily dose of 0.2 mg	43	25.3
A daily dose of 0.4 mg	15	8.8
A daily dose of 0.5 mg	9	5.3
Unknown	30	17.6
Duration		
1 month	21	12.4
2 months	43	25.3
3 months	27	15.9
Unknown	30	17.6

Notably, 16 women (9.4%) denied the presence of any congenital or hereditary pathology in a child at birth, despite the fact that CHD was diagnosed.

To estimate the effect of FA on CHD, questions about the consumption of this supplement before and during the first trimester of pregnancy were included in the questionnaire. Analysis of these answers is shown in Table II.

Discussion

CHD remains one of the most common medical and social problems in the world. Its incidence is 9 per 1000 neonates without any tendency to decrease [9]. This pathology is often the cause of childhood mortality and disability. Although the survival of CHD patients in Ukraine has improved significantly due to advancements in cardiac surgery [2, 6], the information regarding the future of patients who received medical care for CHD is still not systematized and often remains unavailable. Correspondence surveys, including in the form of questionnaires for CHD patients and their families, appear to be a very effective way to obtain missing information about medical outcomes for these patients [5, 17]. In our study, out of 170 children born with CHD, 32 (18.8%) needed urgent surgical correction for severe CHD. It is known that mortality in case of such pathologies is extremely high – from 55% to 70% of children do not

survive more than a year without surgical correction [3]. Surgical correction for CHD was performed in 7.1% of patients. Hospital mortality was 13%, (1.2% – postoperative). Obtained data correspond to several reports that the survival of CHD patients in Ukraine has improved significantly due to advancements in cardiac surgery [6, 14]. Another interesting fact is that about 1/4 of women who gave birth to children with CHD suffered from viral infection during pregnancy.

The analysis of results indicated the lack of knowledge about the preventive effect of FA supplementation on normal organ system development among women who gave birth to children with CHD. The majority of respondents were not aware that the recommended dose of FA is 0.4 mg daily for at least 2 months before pregnancy and the first 3 months during the first trimester [11, 13, 16, 17]. The results of our follow up study revealed that out of 170 studied cases of CHD, approximately 30% of children were generally healthy. However, 32 patients (18.8%) had severe CHD that required urgent surgical intervention, 12 children (7.1%) were operated on, and 2 (1.2%) were recovering after surgery at the time of the survey. Hospital mortality was registered in 22 cases (12.9%). Approximately 30% of children born with CHD were disabled. The analysis of answers provided by mothers of children with CHD has demonstrated the insufficient level of awareness about the preventive effect of FA on the development of congenital malformations.

Collected data about the outcomes of CHD in newborns of Lviv region in Ukraine will allow creating a database for future research. These results could help expand the definition of prenatal pathology risk groups and help improve current approaches in genetic counselling and specialized care for pregnant women at risk of fetal congenital heart disease.

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