



The commencement of congenital heart diseases registry in Isfahan, Iran: Methodology and design

Bahar Dehghan⁽¹⁾ , **Mohammad Reza Sabri**⁽²⁾ , Mohsen Hosseinzadeh⁽³⁾, Alireza Ahmadi⁽⁴⁾, Mehdi Ghaderian⁽⁴⁾, Nizal Sarrafzadegan⁽⁵⁾, Hamidreza Roohafza⁽⁶⁾

Original Article

Abstract

BACKGROUND: Reported prevalence of congenital heart diseases (CHDs) varies widely among studies worldwide. The incidence of CHD, total number of pediatric and adult grown-up congenital heart disease (GUCH), is not determined in Iran. Therefore, we have designed a system to register the information of patients with CHD for the first time in our country.

METHODS: CHD registry is a database in which the patients' data are collected by five pediatric cardiologists from three referral hospitals affiliated to Isfahan University of Medical Sciences, Isfahan, Iran, and five outpatient clinics. We enrolled patients with CHD either as new cases who were referred for evaluation of potential CHD or those who were being followed within the outpatient clinics and entered their whole information in a website specifically designed for it. All the information was collected from checklist by those pediatric cardiologists

RESULTS: From April 2017 to April 2020, after developing the forms and website, the Quality Control Committee evaluated the first 558 files. 73 files (13%) needed major revisions. Among them, 34 (46%) files were omitted totally and the 39 remaining files were revised and completed. After that revision, we changed our checklist and gathered about 1600 patients accordingly.

CONCLUSION: Registry of CHDs not only improves epidemiologic studies but also assists researchers to understand how much a disease management is useful and how to raise the quality of cares and outcomes. Moreover, this provides a better insight for policymakers to understand the extent of health-related problems as well as the issues related to the prevention and management of CHDs all around the world.

Keywords: Registries; Congenital; Heart Diseases; Data Collection

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Introduction

Congenital heart diseases (CHDs) are one of the most common congenital anomalies seen in human. Reported incidence of CHD varies widely among studies worldwide, on average 8 cases per 1000 live births.¹ Nowadays, the number of children born with CHDs is increasing (from 0.4 to 9 cases per 1000 live births).^{2,3} Great evolution in cardiovascular diagnostics and surgery methods has

enabled the physicians to identify CHDs that are more complex even during the intrauterine periods,

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1- Assistant Professor, Pediatric Cardiovascular Research Center, Cardiovascular Research Institute, Isfahan University of Medical Sciences, Isfahan, Iran

2- Professor, Pediatric Cardiovascular Research Center, Cardiovascular Research Institute, Isfahan University of Medical Sciences, Isfahan, Iran

3- Assistant Professor, Department of Pediatric Cardiology, School of Medicine, Ahvaz Jundishapur University of Medical Sciences, Ahvaz, Iran

4- Associate Professor, Pediatric Cardiovascular Research Center, Cardiovascular Research Institute, Isfahan University of Medical Sciences, Isfahan, Iran

5- Professor, Isfahan Cardiovascular Research Center, Cardiovascular Research Institute, Isfahan University of Medical Sciences, Isfahan, Iran

6- Assistant Professor, Cardiac Rehabilitation Research Center, Cardiovascular Research Institute, Isfahan University of Medical Sciences, Isfahan, Iran

Address for correspondence: Mohammad Reza Sabri; Professor, Pediatric Cardiovascular Research Center, Cardiovascular Research Institute, Isfahan University of Medical Sciences, Isfahan, Iran; Email: sabri@med.mui.ac.ir

and to refer those who need emergency therapeutic interventions. In addition, improvements in cardiothoracic surgery have led to a rise in their life expectancy and quality. According to the Civil Status Registration Organization, more than 1500000 babies are born every year in Iran.⁴ However, the incidence of CHD is not determined and also the total number of pediatric and adult grown-up congenital heart disease (GUCH) is unclear in our country as well.

Consequently, in order to find the real number of patients with CHDs and also not miss them, we designed a system to register these patients and collect their information for the first time in Iran, according to our knowledge. This will definitely help improve medical knowledge and prevent and treat patients.

Materials and Methods

CHD registry is a database in which the patients' data from hospitals or outpatient clinics are collected. It is a part of the biggest registry program in Iran - Persian Registry of cardioVascular disease (PROVE) - established by Isfahan Cardiovascular Research Center, a World Health Organization (WHO) collaborating center in Isfahan, Iran.⁵ We launched the registry program based on the following steps:

Upgrading the checklist: The CHD registration program first started in 2015 mainly with limited data but became more advanced by 2017. In the beginning, Ethics Committee affiliated to Isfahan University of Medical Sciences approved our program. We used a population-based protocol to register CHD. The Quality Control Committee, containing epidemiologists, statisticians, specialized physicians, and information technology staff,⁵ validated our checklist. It was developed to collect all the information about patients with CHD and categorized the obtained data into six domains: maternal history and birth, medical history, current clinical status, paraclinical data, cardiac diagnosis, and plan and medications which consist of: demographic characteristics and medical history of patient and parents including their job, complete pregnancy history including maternal diseases, used medications, and even address of the location where mother has lived during pregnancy, positive family history of CHDs in parents, other siblings, or close relatives, clinical presentations and diagnostic findings in current visit, and medications and recommendations for management.

All five pediatric cardiologists who are faculty

members at Isfahan University of Medical Sciences collected these data from the checklists as principle investigators and importers at three university-affiliated referral hospitals and five outpatient clinics.

Gathering information from the patient: In the next stage, referrals were enrolled either as new cases who were referred for evaluation of potential CHD or those who were being followed within the outpatient clinics. All patients or their parents received written informed consent to fill the checklist. The CHDs diagnosis has been principally made by echocardiography although more information was achieved from other diagnostic tests such as angiocardiology and catheterism, computed tomography (CT) and CT angiogram, or cardiovascular magnetic resonance imaging (CMR) in some cases. Then information was classified according to the International Classification of Diseases, 10th Revision (ICD-10) coding system (Q20-26).

Running website: Concurrently, a website was designed by Isfahan University of Medical Sciences to connect these five investigators and the data they gathered from every patient. Each investigator enters the information collected from checklists in this site and uses the patient's national code as an identification code. Furthermore, each importer makes an electronic diagnostic file and uploads it on the site. Therefore, other investigators may have online access to both patient's full history and the latest information especially beneficial in emergencies.

This website includes several parts such as patient's condition gathered from checklist, physical exam and the diagnostic tests, demographic, maternal, and birth history, medical history, current clinical and paraclinical data, cardiac diagnosis, and plan and medications. We also provided a part to enter the new information of old patients who have registered in previous visits and already come to follow up. It also allows the investigators to search patients not only by their names or codes but also by entering their ages or congenital heart diagnosis. Furthermore, it can sort the information based on the patient's arrival date and physician or city where the patient was registered.

In order to assimilate our records, we provided a specific dictionary and user manual, consisting of any specific information investigators need to know to complete the files.

The checklist, dictionary, and user manual are available on our website; however, for security reasons, the patient's data are not accessible except for those five principle investigators.

In addition to internal quality control (IQC) of

PROVE, this project was controlled by the team's supervisor evaluated by the committee which consisted of experienced and trained members who were not one of the PROVE executive members and were unaware of it. They performed an external and continuous control over the entire registry components from the beginning to the end.

Results

About 1600 patients with CHDs were recruited from April 2017 to April 2020, after developing the forms and website. The Quality Control Committee evaluated the first 558 files. 73 files (13%) needed major revisions. Among them, 34 (46%) files were omitted totally and the 39 remaining files were revised and completed. The information which our staff recorded from our primary checklist was too brief and was not compatible with our system. After the Quality Control Committee's assessment, we changed some parts of our checklist and designed it according to the experts' plan and scientific needs. We categorized the obtained data into six domains: maternal history and birth, medical history, current clinical status, paraclinical data, cardiac diagnosis, and plan and medications. Then all the information was recorded on the system.

After that revision, we changed our questionnaire and gathered about 1600 patients accordingly.

Discussion

To the best of our knowledge, it is the first registry database established for CHDs in Iran.

In this regard, there is now a lot of research that shows the prevalence of CHD in several parts of the world, extracted from the registry dataset. In one of the largest studies, van der Linde et al. demonstrated that in the course of time, the CHDs prevalence has increased considerably from 0.6 per 1000 live births in 1930 to 9.1 per 1000 live births after 1995. They also found remarkable geographical variances among their patients. The highest prevalence was in Asia and the lowest in Africa.² Dolk et al.⁶ reported the prevalence of CHDs which were diagnosed from prenatal to infantile periods, and also fetal and perinatal mortality due to CHDs in Europe with the usage of data from European Surveillance of Congenital Anomalies central database.

In China, Qu et al. established a hospital-based CHD registry in Guandong, China, from 2004, to study the epidemiology of CHD including stillbirths and live births and compare the incidence of CHDs in that region to the literatures in 2012. They could

find the prevalence of different CHDs' subtypes too.⁷ Tankeu et al. have conducted a study to find out the prevalence of CHDs and their different patterns inside African countries but their results have not yet been published.⁸

There are several studies reporting the prevalence of congenital anomalies all around the world through registry programs⁹ using a manual which concentrates on population-based and hospital-based surveillance programs¹⁰ and some countries have extracted specific data about congenital heart defects from these available resources. For example, Benavides-Lara et al. could describe the incidence of CHD in Costa Rica and assess their country's registry method.¹¹ In a large study, Rosa et al. reviewed the association between congenital cardiac and non-cardiac malformations based on congenital anomalies registries.¹²

Registry of diseases is useful for other purposes rather than epidemiologic studies. Nowadays, researchers can achieve more information about the efficacy of treatment by following up patients who registered before. American College of Cardiology (ACC) has published a literature reviewing the safety and efficacy of some specific congenital heart procedures by the use of ACC's Improving Pediatric and Adult Congenital Treatments (IMPACT) registry.¹³ In Sweden, a national registry of CHD has been established since 1990, but Bodell et al. recently evaluated the validity of data used in registry to employ them for medical and research purposes.¹⁴ As well, Pradat conducted a study to determine the association of specific CHDs with extracardiac malformations with the information from the registry.¹⁵ These registry programs are still running in different parts of the world; for example, in the United States of America (USA), Mahle has conducted a cohort as "Congenital Heart Disease Research Registry" in Emory University, Atlanta, since 2014. He not only used the participant's demographic and phenotypic data as well as medical records and assessment data, but also collected their blood samples and preserved them for potential future genetic research.¹⁶

Conclusion

Registry of CHDs not only improves epidemiologic studies but also assists researchers to understand how much a disease management is useful and how to raise the quality of cares and outcomes. Moreover, this provides a better insight for policymakers to understand the extent of health-related problems as well as the issues related to the

prevention and managements of CHDs all around the world.

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The study was reviewed by the Ethical Committee of Isfahan University of Medical Sciences (IR.MUI.MED.REC.1398.086) (Project registration code: 298005).

Conflict of Interests

Authors have no conflict of interests.

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