Incidence of Congenital Heart Disease in Jordanian Children Born at Jordan University Hospital; a Seven-Year Retrospective Study

Iyad AL-Ammouri [1], Fares Ayoub [2], Laila Tutunji [1]

Abstract

Objective: The purpose of this study is to report the incidence of congenital heart disease diagnosed in children born at Jordan University Hospital over seven-year period.

Methods: All echocardiographic studies performed for babies between August 2007 and November 2014 were reviewed. In addition, children who were diagnosed with congenital heart disease in outpatient department, and were born at Jordan University Hospital during the same period were included in the analysis. Number of newborns was determined from hospital data base. We report incidence, diagnoses, and age at diagnosis.

Results: Of 31,078 live births, we found 383 (12.3/1000 live birth) patients with congenital heart disease, 52% were males. In 268 patients (70%), the diagnosis was made in the neonatal period, including 16 patients with prenatal diagnosis that was confirmed after birth. In the remaining 115 patients (30%), the diagnosis was made in the outpatient department at a mean age of 7±11 months. The most common diagnosis was ventricular septal defect (43%), followed by atrial septal defect (20%). Cyanotic heart disease accounted for 11% of all congenital heart disease.

Conclusion: Congenital heart disease is present in 12.3/1000 Jordanian live births. It is likely that this incidence, although slightly higher than reported incidence worldwide, is an underestimate of the true incidence in Jordan due to under-diagnosis of asymptomatic lesions, presentation of patients to other institutions, or premature death without diagnosis. A national registry for congenital heart disease is needed to provide more accurate incidence, and help plan national health care policies.

Keywords: Congenital heart malformations, neonatal diagnosis, septal defects.

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Introduction

Congenital heart disease is the most common congenital anomaly with a global incidence of 6-9 per 1000 live births[1-3]. The reported incidence of congenital heart disease from a few middle-eastern countries appears to be higher than that reported from western

1. The University of Jordan, School of Medicine, Department of Pediatrics, Section of Cardiology. Amman 11942, Jordan.
2. The University of Jordan, Medical Student, School of Medicine, Amman 11942, Jordan.

**** Correspondence should be addressed to:***
Iyad AL-Ammouri, MD, FAAP, FACC
Associate Professor, Department of Pediatrics, Section of Pediatric Cardiology, School of Medicine, The University of Jordan, Amman 11942, Jordan.
E-mail: Iyad72@hotmail.com

countries, ranging from 9-12 per 1000 live births[4-7]. This may be attributed to different genetic background, environmental factors, and higher prevalence of consanguinity.

The incidence of congenital heart disease in Jordan is unknown. The purpose of this study is to report the incidence of congenital heart disease for live, Jordanian newborns delivered at Jordan University Hospital (JUH) during a seven year period from 2007-2014.

Methods

Electronic records were reviewed for all infants born at JUH, a large teaching hospital in the capital city of Jordan, between August 1, 2007 and November 30, 2014. In order to encompass all children with congenital heart disease, all echocardiographic studies performed for newborns at either the neonatal intensive care unit, or normal neonatal unit were reviewed. In addition, medical records of children who were diagnosed with congenital heart disease in the outpatient department were reviewed for the place of birth, and were included if they were born at JUH during the study period.

Definitive diagnosis of congenital heart disease was based on echocardiographic imaging. All echocardiographic studies were performed by a pediatric cardiologist at JUH and included standard two-dimensional imaging, and color, pulse and continuous wave doppler techniques. Two-dimensional echocardiographic loops were recorded in standard parasternal, apical, subcostal and suprasternal views.

Congenital heart disease was defined as an abnormality in cardio-circulatory structure that is present at birth, even if it is discovered much later. A wide range of abnormalities are included in this definition. Neonatal patent ductus arteriosus was not included in the final analysis of congenital heart disease unless it persisted beyond the seventh day of life. Patent foramen ovale was considered normal at any age. Other cardiac findings such as the presence of pulmonary hypertension, the presence of hypertrophic or dilated cardiomyopathy, or the presence of intracardiac masses without structural heart disease were not included in the analysis as congenital heart disease.

Congenital heart disease was classified into three categories based on severity of the lesions and the need for expert care. Classification used in this report is similar to that used in other reports.[2] Categories are summarized in Table 1. Statistical analysis was done using GraphPad statistical software (La Jolla, California, USA)

This study was approved by the institutional review committee at JUH.

Results

During the study period between August 2007 and November 2014, there were 31,078 live births at Jordan university hospital. Males were 16,140 (51.9%). Echocardiographic evaluation during the neonatal period was done for 1478 neonates for several indications. Of these, 807 were done for babies in the neonatal intensive care unit, and 671 in the normal neonatal ward. There were 321 abnormal studies; of which 268 were designated as congenital heart disease, including 16 patients who were diagnosed during fetal life and confirmed after birth. The other 53 abnormal studies included persistent pulmonary hypertension of the newborn (25 patients), left ventricular hypertrophy in infants born to diabetic mothers (23 patients), congenital dilated cardiomyopathy (3 patients) and intracardiac rhabdomyomas in two patients with tuberous sclerosis.

Table 1. Classification of congenital heart disease based on severity

Severe CHD
All cyanotic heart disease
Acyanotic CHD
AVC
Large VSD
Large PDA
Severe AS
Severe PS
Critical COA
Moderate CHD
Mild or moderate AS or AI
Moderate PS
Noncritical COA
Large ASD
Complex forms of VSD
Mild CHD
Small VSD
Small PDA
Small ASD
Mild PS
BAV with no stenosis or incompetence

AI: aortic insufficiency, AS: aortic stenosis, AVC: atrioventricular canal, BAV: bicuspid aortic valve, CHD: congenital heart disease, COA: coarctation of aorta, PDA: patent ductus arteriosus, PS: pulmonary stenosis, VSD: ventricular septal defect

Revision of medical records on children who were diagnosed with congenital heart disease in the outpatient department revealed another 115 children who were born at JUH during the study period. Diagnosis in these patients was made at a mean age of 7 ±11months.

The total number of patients diagnosed with congenital heart disease was 383 which makes an incidence of 12.3/1000 live births. The most common diagnosis was ventricular septal defect accounting for 43% of all congenital heart disease. Of the 165 patients with ventricular septal defect, 105 were muscular, 58 were perimembranous, and 2 were subarterial doubly committed (outlet septum) defects. The second most common diagnosis was atrial septal defect accounting for 20% of all congenital heart disease. Of the 78 patients with atrial septal defect, majority was secundum defects (75

patients), two patient had primum defects, and one patient had sinus venosus defect. Patent ductus arteriosus accounted for 9% of all congenital heart disease with 34 patients diagnosed during the study period; 17 patients were premature babies, where the diagnosis was made in the neonatal intensive care unit, and the ductus arteriosus persisted beyond the first week of life. The other 17 patients with patent ductus arteriosus were diagnosed in the outpatient setting. Since ductus arteriosus is a normal fetal structure, and spontaneous closure usually occurs in the first few days after birth, patent ductus arteriosus was considered normal if detected within the first week of life. Revision of echocardiographic studies done during the first week of life showed high prevalence of patent ductus arteriosus (28%). As expected, the prevalence of patent ductus arteriosus was more common when echocardiographic evaluation was done in the first day of life (174 of 493 studies, 35%) compared to the prevalence when evaluation was done beyond the first day of life (87 of 423 studies, 20%) (p= 0.0001)

Cyanotic heart disease collectively accounted for 11% of all congenital heart disease (N=42); the most common of which were tetralogy of Fallot, double outlet right ventricle, and transposition of great arteries with almost equal percentage of 2% each. Types and frequency of congenital heart disease are outlined in Table 2.

Table 2. Distribution of congenital heart disease among 383 patients diagnosed in children born at Jordan University Hospital between 2007 and 2014

Diagnosis	Number	Percentage
Ventricular septal defect	165	43.1
Atrial septal defect	78	20.4
Patent Ductus arteriosus	34	8.8
Pulmonary valve stenois	17	4.4
Bicuspid aortic valve	13	3.4
Coarctation of Aorta	10	2.6
Atrioventricular canal defect	8	2.1
Double outlet right ventricle	8	2.1
Tetrology of Fallot	8	2.1
D-Transposition of great arteries	7	1.8
Hypoplastic left heart syndrome	6	1.6
Aortic stenosis	5	1.3
Pulmonary atresia with Intact ventricular septum	5	1.3
Truncus arteriosus	3	0.8
Double inlet left ventricle/ Single ventricle	3	0.8
L-Transposition of great arteries	2	0.5
Congenital mitral regurgitation	2	0.5
Pulmonary atresia with ventricular septal defect	2	0.5
Subaortic membrane	2	0.5
Congenital coronary anomaly	2	0.5
Interrupted aortic arch	1	0.3
Ebstein anomaly	1	0.3
Congenital aorto-pulmonary fistula	1	0.3
Total	383	100

Patients with congenital heart disease were classified according to severity and the need for expert care into three categories (Table 1). Of the 383 patients, severe disease was present in 88 (23%), moderate disease in 113 (30%), and mild disease in 182 (48%). The incidence of congenital heart disease, which will likely need expert care and/or some form of intervention or surgical procedure during childhood (201 patient with severe and moderate disease combined), was 6.5 per one thousand live births. Mild diseases, not likely to require special care apart from follow up, were made up mainly of small ventricular septal defects (120 patients) and small atrial septal defect (40 patients). Others, included bicuspid aortic valve with no stenosis or regurgitation (11), small patent ductus arteriosus (6), and mild pulmonary valve stenosis (5).

Mean age of diagnosis of patients with severe disease (18 ±84 days) was significantly less than patients with mild or moderate disease (84 ±241 days) (p=0.013). Age at diagnosis of patients with cyanotic heart disease (4.8 ±16 days) was significantly less than patients with non-cyanotic heart disease (77 ±229 days) (p=0.043).

Discussion

The incidence of congenital heart disease in Jordan has not been reported previously. Like in other developing countries, challenges in determining incidence of congenital disease include, but not limited to, lack of registries for common diseases, under diagnosis of uncommon diseases, and unequal distribution of proper medical care on national level favoring urban areas and large cities. This may explain the lack of data in the literature from many developing and low income countries.[1,8,9] In this report we sought to estimate the incidence of congenital heart disease in Jordan by reviewing all patients diagnosed with congenital heart disease in a single large institution over a seven year period. This, although not inclusive of all nationwide live births, may provide an approximate estimation of the incidence in Jordan, which in turn helps guide future public health policy.

Global incidence of congenital heart disease has been rising for the last several decades.[1,2] The rise was mainly attributed to the method of diagnosis resulting in significantly more inclusion of small defects such as small atrial septal defects, ventricular septal defects and patent ductus arteriosus. Incidence has plateaued over the last two decade with a range of 6-9 per 1000 live births.[1-3] There is also variability among continents being highest in Asia and lowest in Africa.[1]

In our study, the incidence of congenital heart disease was 12.3/1000 live births. This is significantly higher than the worldwide incidence, but close to the reported incidence from some surrounding developing countries.[4-7] Incidence from Middle Eastern and Arab countries is summarized in Table 3. Of note, a few reports have lower incidence, which is probably due to different methodology.[10-13]

Despite the relatively high incidence of congenital heart disease in our cohort, it is noted that almost half of these patients have mild or trivial disease. In addition, most of the diagnoses were made during the neonatal period, reflecting that in large and tertiary care hospitals in urban areas, where services are readily available, it would be more likely to detect simple and asymptomatic lesions when compared to underserved areas. This also explains the relatively young age at diagnosis in our whole cohort. When considering patients who have moderate or severe disease, where an intervention is likely to be needed during childhood, our incidence was 6.5/1000, which is close to reports from other countries.[10]

Table 3. Reported incidence of congenital heart disease among Several Middle Eastern and Arab countries

Autor	Area	Period	Incidence (per 1000 live births)
Bitar FF, et al	Lebanon	1980-1995	11.5
Zaqout M, et al	Gaza	2010	10
Subramanyan R, et al	Oman	1994-1996	7.1
Andrej R, et al	Qatar	1984-1994	12.2
Rahim F, et al	Iran	1998-2007	12.3
Dorra A, et al	Tunisia	2010-2011	6.8
Al-Mesned A, et al	Saudi Arabia, Alqassim	2008-2010	5.4*
Baspinar O, et al	Turkey	1995-2002	7.8
Al-Ammouri I, et al	Jordan	2007-2014	12.3

* Incidence of severe congenital heart disease

Multiple factors may explain the higher incidence of congenital heart disease in Jordan compared to worldwide incidence: There is high rate of consanguinity in Jordan which increases the risk of genetic disorders.[14,15] It was shown in multiple studies that consanguinity increases the risk of congenital heart disease.[16,17] In addition, the prevalence of obesity and diabetes mellitus is relatively high in Jordanian women which may be an additional risk for development of congenital heart disease.[18, 19]

Despite the higher incidence of congenital heart disease reported in our study, it is likely an underestimate of the true incidence in Jordan due to multiple factors including under-diagnosis of asymptomatic lesions, presentation of patients to other institutions, or premature death without post-mortem diagnosis.

Finally, patients with both uncorrected and corrected congenital heart disease surviving into adulthood often require specialized care. A continued high incidence of congenital heart disease in Jordan, with continued high fertility, birth rate and population growth of 27 per thousand capita [20] will equate to more patients requiring specialized care in the future, particularly with the improvement of medical care and decreased mortality of these patients. The possible financial and resource burden on Jordan's health system must be considered in planning for future public health policies. An increased public awareness of the risks of consanguineous marriages and measures directed towards better family planning are essential to address this issue.

Despite being a hospital based study, this paper is the first one to report the incidence of congenital heart disease in Jordanian children. Another report from Jordan discussed the patterns of CHD in Northern Jordan.[21] The types and percentages of different common CHD were similar to what we found in our report, where VSD is the most common reported anomaly.

This study has a number of limitations; in addition to being a retrospective review, it involves a single institution in an attempt to provide a national incidence, which is clearly a major limitation. There is also a high likelihood of missing patients who were not detected in the neonatal period and were diagnosed with congenital heart disease at other institutions after discharge, and never referred back to our

institution. In addition, some patients may have asympomatic disease and are still undiagnosed. Despite these limitations, we believe that our study provides an estimate of the incidence of congenital heart disease in Jordan, which is important in providing a guide for planning health care policies, and stimulating development of national registries for congenital heart disease.

Conclusion:

The incidence of congenital heart disease in Jordan seems to be higher than the global incidence, but comparable to few other neighboring and Arab countries. There needs to be a national registry for congenital heart disease to better determine the incidence and to help plan health care policies.

Ethical standards

The authors assert that all procedures contributing to this work comply with the ethical standards of national guidelines and have been approved by the institutional Ethical Committee at JUH.

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نسبة حدوث أمراض القلب الخلقية للأطفال المولودين في مستشفى الجامعة الأردنية خلال فترة سبع سنوات

إياد العموري[1]، فارس أيوب[2]، ليلى توتونجي[1]

1- كلية الطب، الجامعة الأردنية.

الملخص

الهدف: تهدف هذه الدراسة لمعرفة نسبة حدوث أمراض القلب الخلقية للمواليد في مستشفى الجامعة الأردنية خلال فترة سبع سنوات.

الطريقة: تمت دراسة جميع صور الأمواج الصوتية التي أجريت لحديثي الولادة في مستشفى الجامعة الأردنية ما بين شهر آب 2007، وتشرين الثاني 2014 لمعرفة المرضى المصابين بأمراض القلب الخلقية. بالإضافة إلى ذلك، تم مراجعة بيانات المرضى الذين تم تشخيصهم بأمراض القلب الخلقية في عيادة قلب الأطفال حيث تم إضافتهم لعينة الدراسة في حال كانت ولادتهم في مستشفى الجامعة الأردنية خلال فترة الدراسة. نعرض في هذه الدراسة نسبة حدوث أمراض القلب الخلقية عند الأطفال وطبيعتها، ومتوسط العمر عند التشخيص.

النتائج: من بين 31078 مولودا في مستشفى الجامعة الأردنية خلال فترة الدراسة، تم تشخيص أمراض القلب الخلقية لدى 383 حالة (نسبة الحدوث 12.3 لكل ألف مولود حي). كانت نسبة الذكور 52%. وكان أكثر الأمراض شيوعا الثقب البطيني وشكل ما نسبته 43% من مجمل الحالات، يتبعه الثقب الأذيني بما نسبته 20%. أما بالنسبة لأمراض الزرقة القلبية فشكلت ما نسبته 11% من مجمل الحالات. تم التشخيص خلال فترة ما بعد الولادة مباشرة في 268 حالة (70%)، بينما تم تشخيص باقي الحالات وعددها 115 حالة في عيادة قلب الأطفال في متوسط عمر 7 أشهر (±11).

الاستنتاج: أمراض القلب الخلقية تحدث في 12.3 لكل ألف ولادة حية في الأردن. وهذه النسبة تعد أعلى قليلاً من حالات الإصابة المبلغ عنها في بقية أنحاء العالم. يوصي الباحثون بضرورة إنشاء سجل وطني لأمراض القلب الخلقية لما لذلك من أهمية في رسم سياسات الرعاية الصحية الوطنية للأطفال المصابين.

الكلمات الدالة: عيوب القلب الخلقية، نسبة أمراض المواليد، الثقوب القلبية.