Epidemiology of Congenital Heart Disease in the Kingdom of Bahrain

Abhinav Agarwal, MBBS MD, FPC (RGUHS)* Suad R. Al Amer, MD, DCH, SSC-P, SF-Ped (Card)**
Neale Nicola Kalis, MBChB, M Meds (Paeds), FCP (Paeds) SA***

Background: Congenital heart diseases (CHDs) are the most common congenital anomaly in the newborn population with significant childhood morbidity and mortality. There is no published data for the patterns of CHD available in Bahrain.

Objective: To evaluate the incidence and trends of CHD and its subtypes over 17 years.

Design: A Retrospective Observational Study.

Setting: Pediatric Cardiology Department, Mohammad Bin Khalifa Bin Salman Al Khalifa Cardiac Center (MKCC), Bahrain.

Method: The study was performed from January 2000 to December 2016. Two thousand one hundred eighty-nine cases of CHDs were reviewed. All cases of CHD were confirmed by echocardiography and/or cardiac catheterization.

Result: Two thousand one hundred eighty-nine cases of CHD were diagnosed over 17 years with a cumulative incidence of 7.54/1000 live births; 1774 (81%) were acyanotic and 415 (18.9%) were cyanotic heart diseases. There was an increase in the incidence of CHD during the study period. Five hundred forty-five (24.9%) patients were ventricular septal defect (VSD), and 171 (7.8%) were Tetralogy of Fallot (TOF). The incidence rates of VSD, transposition of the great arteries, pulmonary atresia, Ebstein anomaly, and truncus arteriosus were lower than previously published studies. TOF and aortic stenosis were higher than in previous studies.

Conclusion: There is an increase in the incidence of CHDs. As the incidence and patterns differ from other studies, follow-up studies of this population in the future are indicated. There is a need to evaluate the population group and develop health policies.

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Congenital heart disease (CHD) is defined as a structural abnormality of the heart or intra-thoracic great vessels that is potentially of functional significance[1]. CHD is the most common congenital anomaly affecting newborns with an incidence of 4-5/1000 live births[2,3]. There is variation in the incidence and prevalence; recent studies reported it to be as high as 5/1000 live births[4]. Reasons for this increase could be due to the rising awareness of CHD, its treatment, widespread availability of echocardiography, and inclusion of milder lesions that were excluded before the availability of echocardiography[5].

Many CHD requires advanced medical support. Modern treatment strategies have resulted in rising adults with CHD compared to children. Adults with CHD constitute 60% of the total CHD population in the United States of America[6]. They have higher morbidity and mortality than the general population[7]. It is necessary to estimate the true incidence and prevalence of this group to evaluate their needs. The development of health policies is dependent on epidemiologic indicators, and as such, research is necessary to aid in the rational use of local and regional human and infrastructural resources.

The total number of infants born in the Kingdom of Bahrain is about 20,000 annually[8]. Accordingly, the previous estimates of CHD incidence was 5/1000 live births[3]; children born annually with CHD in Bahrain should only be 100. Clinicians, however, are encountering numbers larger number than that.

The aim of this study was to evaluate the incidence of various types of CHD and the trends over the study period.

METHOD

The study was performed from the year 2000 to 2016. The MKCC is the only tertiary care referral cardiac center catering

* Senior Resident
Pediatric Cardiology
Mohammed bin Khalifa bin Salman Al-Khalifa Cardiac Center
Bahrain Defence Force Hospital
** Consultant Pediatric Cardiologist and Senior Lecturer
*** Consultant Pediatric Cardiologist and Associate Professor
Mohammed bin Khalifa bin Salman Al-Khalifa Cardiac Center
Bahrain Defense Force Hospital and Royal College of Surgeons of Ireland Medical University of Bahrain (RCSI-MUB)
P.O. Box 28743 Kingdom of Bahrain
E-mail: nnkalis@batelco.com.bh

to the population and responsible for all invasive treatment of cardiac illnesses. The study includes all the cases that were referred for diagnosis or further management from peripheral centers. Neonates that were delivered in the center diagnosed with CHD pre or postnatally were included in the study.

Suspected cases of CHD were all confirmed with echocardiography and rarely additional cardiac catheterization is needed. In patients with multiple lesions, a diagnosis was labelled with only the primary dominant lesion. Data were directly recorded into a Microsoft Excel spreadsheet (Microsoft Inc., Redmond, Washington, USA) for analysis. Statistical analysis was performed using IBM SPSS 25.0 software and descriptive analyses were performed.

Exclusion criteria were functionally insignificant abnormalities (left superior vena cava, bicuspid aortic valve without stenosis, mitral valve prolapse without mitral regurgitation); arrhythmias without structural heart diseases; premature infants (<36 weeks' gestation) with patency of the arterial duct; small defects (patent foramen ovale and patency of the arterial duct) with spontaneous closure before 3 months of life and non-pathological mild regurgitant lesions.

RESULT

Two thousand one hundred eighty-nine cases of CHD were diagnosed over 17 years with a cumulative incidence of 7.54/1000 live births; 1,774 (81%) were acyanotic heart diseases and 415 (18.9%) were cyanotic heart diseases. Distribution and trend over the years is shown in figure 1.

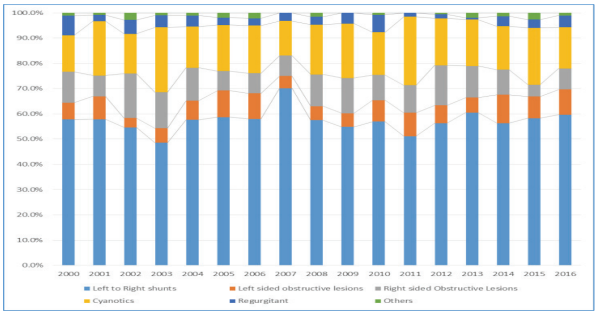

Figure 1: Distribution of CHD

CHD was divided into categories: one is left to right shunt lesions (L to R), such as ASD, ventricular septal defects (VSD), patent ductus arteriosus (PDA), atrioventricular septal defects (AVSD), aortopulmonary window (APW) and partial anomalous pulmonary venous drainage (PAPVD); two is left sided obstructive lesions (LOL), such as mitral stenosis (MS), aortic stenosis (AS), interrupted aortic arch (IAA) and coarctation of the aorta (CoA); three is right sided obstructive lesions (ROL), such as double chambered right ventricle (DCRV), pulmonary stenosis (PS) and peripheral pulmonary stenosis (PPS); four is cyanotic lesions, such as Tetralogy of Fallot (TOF), double outlet right ventricle (DORV), univentricular hearts, hypoplastic left and right hearts, transposition of great arteries (TGA), total anomalous pulmonary venous drainage (TAPVD), pulmonary atresia (PA), Ebstein anomaly and truncus arteriosus); and regurgitant lesions isolated pathological mitral regurgitation (MR), isolated tricuspid regurgitation (TR) and isolated aortic regurgitation (AR).

Cases which could not be classified according to the above were grouped as others, such as aortic arch anomalies other than coarctation of the aorta, anomalous left coronary artery from the pulmonary artery (ALCAPA), aortic root dilatation, coronary artery fistulae, idiopathic pulmonary artery dilatation and cor triatrium.

Figure 2 illustrates the trends of incidence of CHDs. It reveals an increase in the total incidence of CHDs (6.45/1000 live births in 2000 to 9.46/1000 live births in 2016). This increase corresponds to the increment in the incidence of left to right shunt lesions (3.73/1000 live births in 2000 to 5.64/1000 live births in 2016). There were no prominent changes seen in incidence of cyanotic, obstructive or regurgitant lesions.

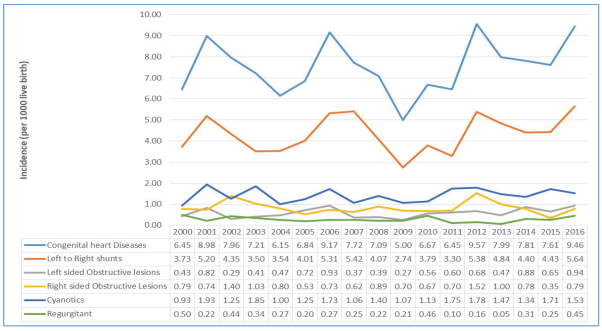

Figure 2: Trends in the Incidence of CHD Over 17 Years

Table 1: Total Number, Percentages and Incidences of Anatomical Subtypes of Congenital Heart Disease

Anatomical Subtypes	Number	Percentage	Incidence (per 1000 live births)
Ventricular Septal Defects	545	24.9%	1.88
Atrial Septal Defect	347	15.9%	1.19
Patent Ductus Arteriosus	261	11.9%	0.90
Pulmonary Stenosis	182	8.3%	0.63
Tetralogy of Fallot	171	7.8%	0.59
Atrioventricular Septal Defects	99	4.5%	0.34
Univentricular Heart/ Hypoplastic Left Heart Syndrome/ Tricuspid Atresia	97	4.4%	0.33
Coarctation of Aorta / Interrupted Aortic Arch	87	4.0%	0.30
Aortic Stenosis	84	3.8%	0.29
Mitral Regurgitation	62	2.8%	0.21
Peripheral Pulmonary Stenosis	53	2.4%	0.18
Double Outlet Right Ventricle	59	2.7%	0.20
D- Transposition of Great Arteries	27	1.2%	0.09
L- Transposition of Great Arteries	16	0.7%	0.06
Total Anomalous Pulmonary Venous Drainage	16	0.7%	0.06
Pulmonary Atresia	13	0.6%	0.04
Ebstein Anomaly	9	0.4%	0.03
Truncus Arteriosus	7	0.3%	0.02
Miscellaneous	54	2.5%	0.19
Total	2189	100%	7.54
Cardiomyopathies	51		0.18
Isolated Dextrocardia without any cardiac anomaly	14		0.05

The total number, percentage, and incidence of specific anatomical lesions are shown in table 1. The most common six lesions are VSD, ASD, PDA, PS, TOF, and AVSD, which make 73.3% of total CHD cases.

Cases that had very low incidence were grouped under miscellaneous category, such as isolated pathological TR 11 (0.5%), aortic arch anomalies 9 (0.4%), ALCAPA 7 (0.3%), pathological AR 6 (0.3%), PAPVD 4 (0.2%), aortopulmonary window 3 (0.1%), DCRV 3 (0.1%), aortic root dilatation 3 (0.1%), coronary artery fistulae 3 (0.1%), idiopathic pulmonary artery dilatation 3 (0.1%) and cor triatrium 2 (0.09%).

Fifty-one (2.3%) cases of cardiomyopathies (incidence=0.18/1000 live births) and 14 (0.6%) cases of isolated dextrocardia without any cardiac anomaly (incidence=0.05/1000 live births) were also diagnosed in that time but not included in the calculations.

The six most common subtypes of CHD revealed a gradual decline in the incidence of VSD whereas increment was noticed for ASD and PDA, see figure 3. There was no change in incidence for PS, TOF and AVSD. The incidence of cyanotic lesions showed no significant change over the study time, see figure 4.

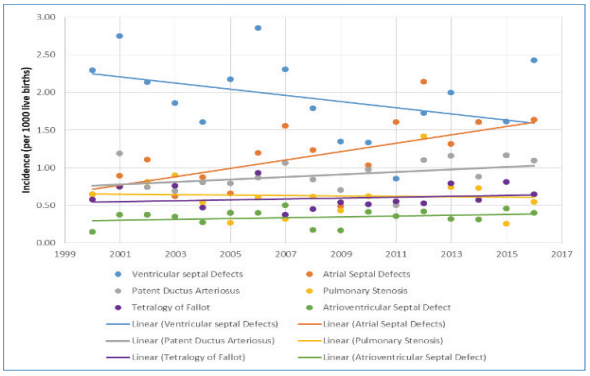

Figure 3: Incidence of Most Common Anatomical Subtypes of CHD

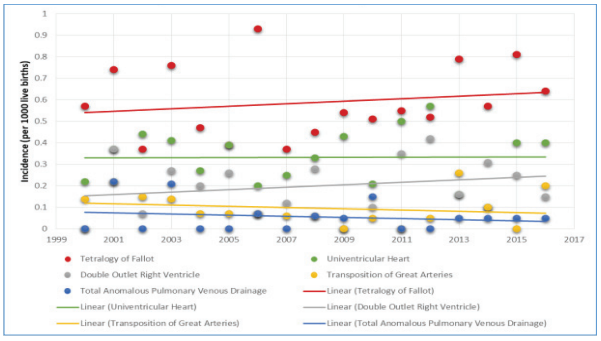

Figure 4: Timeline of Incidence of Cyanotic Lesions

DISCUSSION

All births in Bahrain are recorded and followed up, and all suspected cases of CHD are screened, identified and managed. It is an ideal condition for an epidemiological study.

The survival and life expectancy of children born with CHD have improved significantly; therefore, adults with CHD are more numerous than children with CHD[6]. The prolonged lifespan of patients with CHD and a chance of having offspring may have a significant effect on the incidence of CHD. It has been predicted to double over seven generations[10].

Other reasons that may contribute to the increase in the incidence of CHD are an increase in in-vitro fertilization and late pregnancies, safe delivery of complicated pregnancies, such as gestational diabetes (GDM), and environmental pollution[5]. Fetal diagnosis in the estimation of incidence also plays a role[11].

The incidence of a disease is essential information in the long term planning of healthcare and it varies depending on ethnicity and geography.

We compared the incidence of each subtype in our study with three different studies: Qu et al from Guangdong Registry of Congenital Heart Disease (GRCHD), the European Registry of Congenital Anomalies (EUROCAT), and a review by Hoffman et al[5,12,13].

As the above three studies incorporated data from different ethnic populations, there are some variations in incidence data. Hence, the incidence from our study was considered higher only when it was greater than all three studies and lower when it was lower than all three studies[5,12,13].

In our study, the incidence of VSD was considerably lower than the other studies[5,12,13]. The reason for this could be that the babies with small VSDs that did not require surgery or go for spontaneous closures were likely managed at peripheral hospitals and not referred and hence not included in our data. Hoffman et al found that isolated small VSDs are the most common form of CHD with incidence varying from 2% to 5% and inclusion of these cases can easily alter the incidence data[5]. If they were included, it would have resulted in a higher incidence of isolated VSDs and total CHD incidence in Bahrain.

In our study, the incidence of D-TGA, pulmonary atresia, Ebstein anomaly, and truncus arteriosus were lower than the international data[5,12,13]. However, the incidence of Tetralogy of Fallot and Aortic Stenosis was found to be higher than other studies[5,12,13]. The ethnic and genetic differences from the populations might be the cause of this variation. For the remaining diseases, the incidence was comparable to international data[5,12,13].

Trend analysis found a gradual decline in the incidence of VSDs reported at our center. ASD showed a gradual increase after 2010 because we started managing adults with CHDs in 2010 and that led to many adult patients with ASD being included in our study that was missed during childhood. There is no significant change in the incidence of TOF, AVSD, PS and other cyanotic heart lesions. Similar trends were found in other studies[12,14].

The study is limited by being passive and only suspected cases of CHD were referred to us. In addition, our data is from a tertiary care hospital and does not represent the community population, thus, there might be some selection bias. We might have missed some minor heart ailments that were not referred to us and were managed at other centers. In addition, the various difficulties in detection CHD and its incidence will

always remain and be responsible for the variation of incidence rates in different studies[5].

CONCLUSION

CHD is the most common congenital anomaly affecting the newborn population, with high morbidity and mortality. This study showed that there is a gradual increase in the incidence of CHDs. It is essential to assess the needs of the population and develop health policies. We recommend a countrywide population-based study to evaluate the incidence of CHDs in Bahrain.

Author Contribution: All authors share equal effort contribution towards (1) substantial contributions to conception and design, acquisition, analysis and interpretation of data; (2) drafting the article and revising it critically for important intellectual content; and (3) final approval of the manuscript version to be published. Yes.

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Competing Interest: None.

Sponsorship: None.

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