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Research Article

Congenital Heart Diseases in Saudi Arabia: Epidemiologic Overview during the Period of 1988 - 2008

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Abstract

Background: Congenital heart disease is the most common congenital disorder in newborns. The prevalence of congenital heart disease is unclear, with underestimates of patients who actually have the diseases. In this study, we present an overview of the frequency, prevalence, geographical distribution, extra cardiac disorders and some factors associated with congenital heart diseases in children in Saudi Arabia.

Aim: To describe frequency, distribution and prevalence of congenital heart diseases and associated extra cardiac diseases in children during the period of 1988 - 2008 in Saudi Arabia.

Methods: Descriptive retrospective study of congenital heart diseases for databases collected from all paediatric and maternal hospitals in Saudi Arabia. A total number of 4465 patients were reported from all regions in Saudi Arabia. All patients with congenital heart defects had their diagnoses confirmed by paediatric cardiologist with use of at least echocardiography.

The frequency and prevalence of the anomalies were classified according to ICD-10 classification of congenital heart diseases. Types of congenital heart diseases were compared between geographical areas.

Findings: A total number of 4465 patients were diagnosed with congenital heart diseases, with prevalence of 2.01 per 1000 in children age 0- 4 years of age. The majority of them of them 3436 (77%) were reported from central region. The stratification of the diseases was as fellow: 1078 (24.1%) VSD, 786 (17.6%) PS, 493 (11%) ASD, 473 (10.6%) PDA, 229 (5.1%).

TOF and 202 (4.5%) HLHS. There was an overall statistically significant difference in reported CHD diseases (51% vs. 49%; 0.05) for male to female respectively. Several extra cardiac anomalies were observed associated with CHD, the most frequent were, chromosomal anomaly which was mainly Down syndrome 32.3%, genito-urinary abnormality 14%, Hematological disorders 9.4%, metabolic and endocrine disorders 9.2% and central nervous system defect 8.5%. Ventricular septal defect was the most frequent cyanotic anomaly, and tetralogy of Fallot was the most frequent cyanotic anomaly.

Conclusion: Children with congenital heart defects were mainly reported from central region, ventricular septal defect was the most frequent heart defect, Down syndrome predominant extra cardiac disease.

Keywords: *Congenital Heart Disease; Epidemiology; Extra-Cardiac Anomalies*

Background

Congenital heart diseases and great vessels anomalies are the most frequently diagnosed congenital malformations among neonate. It is usually defined as clinically significant structural heart disease present at birth [1,2] and causes common morbidity and high mortality in the first year of life [3]. The incidence of congenital heart disease at birth depends on how a population is studied [3,4]. Prior to emerge

of echocardiography, estimated incidence of congenital heart diseases ranged from 5 to 8 per 1 000 live births but better diagnosis has detected many more with milder forms, so that current estimates range from eight to 12 per 1 000 live births [5]. Numerous studies conducted with specific populations have had an incidence of between 2 and 10 per 1,000 live births [6].

Aim of the Study

The aim of this study was to describe frequency, distribution and prevalence of congenital heart diseases and associated extra cardiac diseases in children during the period of 1988- 2008 in Saudi Arabia.

Methods

During the period of 1988 to 2008, data of babies born in maternal and pediatric hospitals in all regions of Saudi Arabia was collected. Detailed history was obtained from the parents. All patients underwent clinical assessment, and electrocardiographic examinations, with color Doppler and spectral analysis at our service under supervision of expert pediatric cardiologists.

The diagnosis of cardiac and great vessel anomalies was based on the echocardiographic study. All patients had their diagnoses confirmed by senior pediatrics cardiologist. Cardiac diseases were classified according to ICD-10-CM classification of congenital heart diseases. Extra-cardiac diseases were assessed by clinicians from relevant specialty and classified according based on ICD-10. Data manage med and analyzed were performed by used of SPSS IBM™ Version 22. The frequency and prevalence, types of congenital heart diseases were compared between geographical areas and extra-cardiac diseases were described. Data were presented in tables and graphs in form of number and percentages. Prevalence of the congenital heart diseases was calculated for children 0 - 4 year of age based on demographic indicators according to General Authority of Statistics, Kingdom of Saudi Arabia [7].

Results

A total number of 4465 patients were diagnosed with congenital heart diseases, of them (2189, 51% *versus* 2276, 49%; *P* = 0.0.5) were males and female respectively. The majority 77% of patients were reported from central region, 13% from eastern region, 6.9% from western region, 2.6% from southern region and few cases 0.5% from northern region (Figure 1).

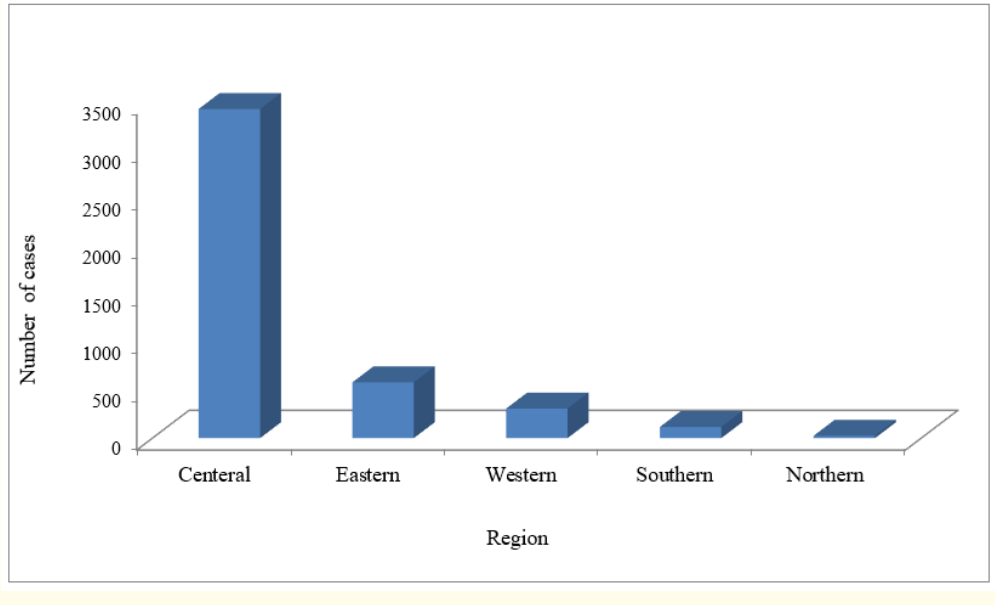

Figure 1: *Distribution of Congenital Heart Diseases by Region in Saudi Arabia*

Overall prevalence of congenital heart diseases was 2 per 1000 in children age 0 - 4 years of age in the country and there was variation in prevalence rate between the regions (Table 1). The highest prevalence was observed in eastern region while the lowest was observed in northern region.

Region	Number of reported Cases	Percentage	Estimate number of children 0 - 4 year	Prevalence per 1000
Central	3436	77.0	671412	5.1
Eastern	582	13.0	796696	0.7
Northern	24	0.5	114045	0.2
Southern	116	2.6	352468	0.3
Western	308	6.9	344886	0.9
Saudi Arabia	4465	100.0	2279506	2.0

Table 1: *Distribution of prevalence of congenital heart diseases in the 5 regions of Saudi Arabia (n = 4465).*

The most prevailing diseases were ventricular septal defect 24.1%, pulmonary stenosis 17.6%, atrial septal defect 11% and patent ductus arteriosus 10.6% (Table 2).

Anomalies	Number of cases	%
Ventricular septal defect (VSD)	1078	24.1
Pulmonary Stenosis (PS)	786	17.6
Atrial Septal Defect (ASD)	493	11.0
Patent Ductus Arteriosus (PDA)	473	10.6
Tetralogy of Fallot (TOF)	229	5.1
Unclassified	205	4.6
Hypoplastic Left Heart Syndrome (HLHS)	202	4.5
Atrioventricular Septal Defect (AVSD)	191	4.3
Dextro transposition of great artery (DTGA)	178	4.0
Aortic Stenosis (AS)	126	2.8
Coarctation of Aorta (COA)	116	2.6
Hypertrophic cardiomyopathy (HCM)	115	2.6
Double Outlet Right Ventricle (DORV)	77	1.7
Truncus Arteriosus (TA)	46	1.0
Pulmonary Atresia (PA)	37	0.8
Dextrocardia (DEX)	29	0.7
Tricuspid Stenosis (TS)	29	0.7
Single Ventricle (SV)	14	0.3
Mitral Valve Prolapse (MVP)	12	0.3
Ebstein Anomaly	8	0.2
Total Anomalous Pulmonary Venous Drainage (TAPVD)	7	0.2
Miscellaneous	14	0.3

Table 2: *Frequency of congenital heart diseases anomalies (n = 4465).*

There was no predominance of gender in the diseases occurrence, 49% were males and females respectively. Proportion of patients with congenital heart defects was highly significant in single pregnancy (96%) compared to 4% in twin, 0.2% in triplet pregnancies and 0.3% for other.

Table 2 shows frequency of congenital heart disease is described in whole country, the most frequent congenital heart defects were ventricular septal defect 24.1%, pulmonary stenosis 17.6%, atrial septal defect 11.0%, persistent ductus arteriosus 10.6%, tetralogy of Fallot (5.1%) hypoplastic left Heart syndrome 4.6%) and atrioventricular septal defect 4.3%.

Several different extra- cardiac anomalies existed within congenital heart diseases in this study. These anomalies were associated with about 22% of diagnosed congenital cardiac diseases. The most common diseases related disorders were Down syndrome consisted 32% of total disorders and associated with 7.1% of all diagnosed CHD, genito-urinary anomalies, hematological disorders, metabolic disorders, central nervous system disorders and respiratory disorders (Table 3).

Anomalies	No	% from total CHD
EENT Disorders	9	0.2
Skin Disorders	13	0.3
Miscellaneous	25	0.6
Gastro-Intestinal Disorders	31	0.7
Growth and Development Disorders	53	1.2
Musculo-Skeletal Disorders	59	1.3
Respiratory Disorders	69	1.5
Central Nervous System Disorder	83	1.9
Metabolic and Endocrine Disorders	90	2.0
Hematological Disorders	92	2.1
Genito-Urinary Disorders	136	3.0
Down syndrome	315	7.1
Total	975	21.8

Table 3: *Extra-cardiac congenital anomalies associated with congenital heart diseases.*

Discussion

This is the one of largest sample size nationwide multi-centers analysis to investigate congenital heart diseases, and the associated extra-cardiac disorders. This study of 4465 of age 0 - 4 year, the majority of the patients were reported central region, and this normally existed due to concentration of population in the capital as well as due to availability of major hospitals. In previous study by W Greer., *et al.* they reported that 26% of the registered cases were from central region [8]. In our study, the overall prevalence of CHD was 2% which was similar to had been reported by in previous study [9]. Several studies reported big discrepancy in incidence of CHD range from 4 to 50 per 10^3 live births [10-14].

Among children with congenital heart disease, the most prevalent of the diagnosis of congenital heart disease in infants coincided with the clinical manifestation of heart defects such as ventricular septal defect, pulmonary stenosis and persistent, atrial septal defect and patent ductus arteriosus, which were prevalent in our study.

Epidemiological studies have shown varied frequency and prevalence of congenital heart diseases. The technological advance and routine use of echocardiography have contributed to improvement in the establishment of the diagnosis, and, therefore, to increase the prevalence of some heart defects [15]. Ventricular septal defect, frequency of 24%, was the first most common defect in our study, similarly to the results of other studies with accidence rates of 22.5% to 43% in the country of this study [16-23].

Pulmonary stenosis anomaly was 17.6% placed in the order of diagnosed diseases in this study, this finding was higher compare to previous studies in which PS, Alabdullager, in a previous study PS was reported 8.9%, while Al-Mesned A., *et al.* described that 7.6% of cases were PS, Alnajjar., *et al.* presented a proportion of 7.9% and Jaiyesimi., *et al.* reported 9% [16,19,21,24]. The third common defect was atrial septal defect 11.04%, this result was comparable to what had been reported by numerous studies which reported broad array of proportions ranged from 9.3% to 26% [16-23].

Complicated heart defects which manifesting in the first days of life, such as hypoplastic left heart syndrome, Tetralogy of Fallot and dextro transposition of great artery were 5.3%,4.5 and 4% respectively. They were less commonly prevailed in our study compared to previous studies [15-23]. Since patients with such diseases die early, and therefore we presumed that a lot of them wouldn't receive optimum treatment and numerous pass away without a diagnosis.

Several extra cardiac anomalies were associated with CHD, the most frequent were, chromosomal anomaly which was mainly Down syndrome was 32.3% of total cases. Recent studies indicate that approximately 5% of all congenital heart defects are associated with some form of chromosomal abnormality, the majority of which are Down syndrome. Reports of the incidence of congenital heart disease in patients who have Down syndrome have varied, but it is commonly accepted to be 50% and 40-50% of babies diagnosed with trisomy 21 have heart defects [25]. In this study genito-urinary tract anomalies were 14% of total diagnosed cases, they were less than found by Kenna., *et al.* in which they reported 20% of infants had major defects notably, genito-urinary and nervous systems associated with CHD [26].

We found hematological disorders of 9.4% while Zabala LM., *et al.* reported that 9.2% of patients had metabolic and endocrine disorders congenital heart disease have complex alterations in their whole blood composition and coagulation profile due to long-standing hypoxemia [27]. In this study we found 2% of cases had metabolic and endocrine disorders mainly thyroid disorders, this finding is in line with the findings of Meissner T., *et al.* in Germany, where they found that patients with congenital heart defects had persistent hyperinsulinism and hypothyroidism [28]. Central nervous system disorders were associated with 1.9% of diagnosed cases in this study without determined pattern neurologic diseases. However, de Los Reyes., *et al.* stated that various neurologic complications occurred in association with congenital heart disease, including cognitive impairment and ischemic stroke. They concluded that likelihood of stroke is greatest in individuals with severe structural cardiac defects such as tetralogy of Fallot, transposition of the great arteries, or hypoplastic left heart syndrome while a persistent foramen ovale adds little or no additional stroke risk unless it is associated with an atrial septal aneurysm or other anomaly [29].

Gastrointestinal disorders were less frequently prevailed in this study only 0.7% of diagnosed patients and consisted about 3.7% of total associated disorders with CHD.

Congenital anomalies of the gastrointestinal tract/abdominal wall were associated with congenital heart disease in 38% followed by anal atresia 30.5% [30]. Duess JW., *et al.* reported that Hirschsprung's disease and associated congenital heart disease ranged from 20 to 80% with an overall prevalence 51% [31]. Respiratory and growth disorders consisted 7.1% and 5.4% of total detected disorders and they associated with 1.5% and 1.2% of all diagnosed CHD in this study respectively. It is evident that children with congenital heart disease with great left-to-right shunt, heart failure, and pulmonary hypertension usually have growth delay. The degree of hypoxia, however, does not show a linear correlation with the degree of impairment [32].

Conclusion

Children with congenital heart defects were mainly reported from central region, ventricular septal defect were being the most frequent anomaly, followed by pulmonary stenosis, atrial septal defect and patent ductus arteriosus. Chromosomal anomaly mainly Down syndrome, genitourinary disorders, haematological disorders and metabolic disorders are the commonest extra –cardiac disorders associated with congenital heart diseases.

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Volume 7 Issue 2 February 2020