

SPECTRUM OF CONGENITAL HEART DISEASES IN BASRA: AN ECHOCARDIOGRPHY STUDY

Jawad Khadim¹, Sawsan Issa²

ABSTRACT

This is a retrospective study that was set out to describe the spectrum of congenital heart disease using echocardiography in two main centers in Basra over a period of 24 months from June (2006-2008). Children with the diagnosis of congenital heart disease were selected; information obtained from their records included age, gender, clinical diagnosis and echocardiography finding. Five hundred seventy patients had congenital heart disease making 40% of 1414 examined children. There were 309(54%) males and 261(46%) females (ratio 1.2:1); their ages ranged from 3 days to 19 years. Two hundred forty four (42.8%) children referred for echocardiography before the age of one year and two hundred one (35.2%) were 1-4 years of age. Ventricular septal defect was the most common congenital heart disease present in 247 patients (43.3%), of these (81.8%) were membranous in type, 41(16.6%) were muscular, and 4(1.6%) were supracristal type. The second commonest congenital heart disease diagnosed by echocardiography was tetralogy of Fallot in 72(12.6%) of examined children. Sixty eight (11.9%) had atrial septal defects, out of which 55(80.9%) were of secundum type, 7(10.3%) were primum, 5(7.4%) had sinus venosus defect and only one case with coronary sinus defect. Distribution of specific lesions and sex distribution were similar to findings from other parts of the world; However, the overall detection rate at 1 year of age was lower, with increasing availability of echocardiography facilities more cases of congenital heart defects are likely to be identified early.

INTRODUCTION

Congenital heart defects (CHD) represent some of the more prevalent malformations among live births and remained the leading cause of death from congenital malformations.^[1,2] CHD defined as an abnormality in cardio circulatory structure or function that is present at birth, even if it is discovered much later.^[3] Congenital heart disease occurs in 0.8-1% of live births. The incidence is higher in still births (3-4%), spontaneous abortion (10-25%) and premature infants (2%) excluding patent ductus arteriosus. It has a wide spectrum of severity in infants; about 2-3/1000 newborn infants will be symptomatic with heart disease in the first year of life.^[2] CHD causes the deaths of thousands of children in developing countries. According to data from WHO; in north America, heart defects account for more than one third of infant deaths due to congenital anomalies and for approximately one tenth of all infant deaths, these estimate are similar for other areas of the world.^[4] Over the past 20-40 years, major advances have been made in the diagnosis and treatment of CHD. Echocardiography is a major mode of cardiovascular imaging with versatile applications, it is a cheap and non invasive technique for the investigation of cardiac diseases with advantage of reproducibility of results, instantaneous images and reliable levels of accuracy.^[5] There are limited data on

echocardiography findings in children with heart disease in Basra, so this study was set out to describe the spectrum of CHD seen in two echocardiography centers in Basrah city.

PATIENTS AND METHODS

This is a retrospective study; transthoracic echocardiography (TTE) data collected over 24 months (June 2006-June 2008) were reviewed. Patients with ECHO diagnosis of CHD were selected. The study was carried out at Echocardiography Clinic in two main hospitals (Basra maternity and children hospital and Al port hospital). Information obtained from patients records included age, gender, clinical diagnosis and echocardiography findings. The presence and severity of any cardiac malformation was analyzed according to recommendations of the American Society of Echocardiography.^[6] Presence of atrioventricular septal defect (AVSD) was determined as either complete if a single common atrioventricular valve was present or partial if both atrioventricular valves (mitral and tricuspid) were seen with primum atrial septal defect, inlet ventricular septal defect or cleft anterior leaflet of mitral valve the patient was placed under the dominant defect. All cyanotic CHD except tetralogy of Fallot (TOF) and transposition of great arteries (TGA) were categorized as complex CHD, which included

common arterial trunk, double outlet ventricles, double inlet ventricles, hypoplastic right ventricle, hypoplastic left ventricle, atrio-ventricular valve atresia and anomalous venous connections. The miscellaneous group included all acyanotic CHD that could not be categorized into one of the other groups. Persistent foramen ovale in the neonate and patent ductus arteriosus (PDA) in premature babies were not included in the analysis, however these were considered if they persisted beyond the first four weeks of life. Echocardiographic modalities applied included M mode, two dimensional and Doppler echocardiography were done with 3 and 7 MHz sector transducer. The data were analyzed using SPSS version 10 soft ware.

RESULTS

A total of 1414 echocardiography examinations were done over 24 months, five hundred seventy (40%) patients had CHD. There were 309 males and 261 females (ratio 1.2:1), their ages ranged from 3 days to 19 years. The mean age was (2.68±3.37) years.

Table 1, Illustrates the age and sex distribution of the patients. Two hundred forty four (42.8 %) children were less than one year old and 445(78%) under 5 years.

Table 1. Age and sex distribution amongst patients with congenital heart disease

Age (years)	Males	Females	Total (%)
<1	135	109	244(42.8)
1-4	110	91	201(35.2)
5-9	47	42	89(15.6)
10-14	13	14	27(4.7)
15-19	4	5	9(1.5)
Total	309	261	570(100)

Table 2, shows the distribution of various congenital heart disease according to the gender. VSD was the most common CHD diagnosed by echocardiography and accounts for (43.3%) of the patients, followed by TOF 72(12.6%), ASD 68(11.9%), PDA 54(9.4%), AVSD 37(6.4%), PS 22(3.8%), complex heart defects (2.6%) and miscellaneous group (3.5%) The distribution of CHD in general was almost predominance for male with VSD, TOF, ASD,

D-TGA while females with PDA were predominant.

Table 2. The distribution of the various congenital heart disease by gender among all patients

Type of CHD	Males	Females	Total (%)
VSD	137	110	246(43.3)
TOF	46	26	72(12.6)
ASD	38	30	68(11.9)
PDA	20	34	54(9.4)
*AVSD	20	17	37(6.4)
D-TGA	14	10	24(4.2)
PS	11	11	22(3.8)
AS	6	5	11(1.9)
Complex	10	5	15(2.6)
Miscellaneous	10	1	20(3.5)
Total	309	261	570(100)

VSD= ventricular septal defect TOF= tetralogy of Fallot ASD= atrial septal defect PDA=patent ductus arteriosus *AVSD=Complete atrioventricular septal defect D- TGA=dextro- transposition of great arteries PS= pulmonary stenosis- AS= ps.aortic stenosis

Out of two hundred forty seven cases of VSD; 202(81.8%) were membranous, 41(16.6%) were muscular in type and 4(1.6%) were of supracristal type. There were 68 patients with ASD, out of which 55(80.9%) were of secundum type, 7(10.3%) were primum type, rare types sinus venosus and coronary sinus were (7.4%) and (1.5%) respectively. Fourteen patients had pulmonary hypertension, eight of them had VSD, four had AVSD and two had ASD. A six-years-old female had Eissenmenger complex in AVSD.

DISCUSSION

This study highlights the common variants of CHD seen in Basrah. The presence and severity of any cardiac malformation was analyzed with similar approach had been followed in American Society of Echocardiography^[6] and other studies.^[7,8] Two thirds of children presented for echocardiography examination before the age of five years and by 1 year of age in about 1/3 of patients; which reflect late presentation and diagnosis of patients with

CHD. Similar results had been conducted by Mahoud et al in Nigeria^[9] but in contrast to other researchers in Saudi Arabia; their results from three centers show early detection of CHD before one year age in (60%) of patients.^[10-12] In other reports the diagnosis of CHD is established by one week of age in 40-60% of patients and by one month of age in 50-60%.^[2,13] Unfortunately; this is the scenario that operates in most developing countries because of the lack of skilled personal, equipment and facilities for diagnosis. Patients presented for echocardiography for the first time at late age in about (22%) in this study, probably due to difficulties in accessing care; neglect of the family to search early medical help. There was a slight male predominance; this is consistent with reports from Mahoud et al,^[9] Al-abdulghader and Reinhold.^[14,15] The distribution of males with CHD suggest an important causative link that is not well understood. When all isolated lesions are classified by embryonic timing of disturbed organogenesis, male had to predominate in those that are developed later in gestation.^[16] Acyanotic CHD was more common than cyanotic CHD, the relative frequencies of individual CHD are consistent with reports from Nigeria,^[17,18] Saudi Arabia^[14] and other parts of the worlds^[2] With VSD being the commonest acyanotic lesion and TOF was the second most common cyanotic lesion in this series, this is consistent with other reports.^[2,9] Concerning cyanotic CHD; TOF constitutes 12.6% of all CHD in Basrah, other study conducted in Nigeria reported a higher frequency 26.2%.^[9] Echocardiography definition of TOF is still lacking as some authors still consider double outlet right ventricle with pulmonary stenosis as TOF, even if most of aortic emanates from right ventricle, this will exaggerate the frequency of TOF. On the other hand still lower frequency of TOF recorded in Saudi Arabia 3.5%^[14] and in Japan 5.8%^[19] Atrial septal defect was the third most frequent lesion 11.9% compared to 5.3% in California^[20] and 12.3% in Nigeria.^[9] Ostium secundum defect being most common type of ASD worldwide.^[2] The frequency of PDA was 6.8% overall, excluding PDA of premature neonate.^[2] In this series; it was 9.4%, changing frequency in different reports may be attributed to differences in definition of CHD, study methodology and

diagnostic accuracy with emphasis on certain exclusion has significant impact on accurate calculation of the real frequency of PDA. Atrioventricular septal defect, a characteristic lesion of Down syndrome, accounted for 6.4% of all CHD, fluctuations of its frequency in different studies is well known, 3.5% in Saudi Arabia^[14] and 8.2% in Nigeria.^[9] Transposition of great arteries was found with a higher frequency in this study compared to other studies done by Alabdalgadar et al (2.1%) in Saudi Arabia^[14] and Hoffman (2.6%) in USA.^[20] The presence of environmental causative factors might have contributed to high frequency of TGA in Basrah, it had been suggested that the interaction between genetic and environmental factors plays a major role in its etiology.^[21] Also it shows a male predominance; which was first noted by MacMahon et al. in 1953^[22] and has been supported by number of subsequent studies.^[23] Regarding obstructive lesion of pulmonary artery and aorta it had been recorded with lower frequency; in contrast to other studies in Japan,^[19] USA^[20] and Saudi Arabia.^[14] With advances in both palliative and corrective surgery in the past 20 years in developed countries, the number of children with CHD surviving to adulthood has increased dramatically, however; despite these advances, CHD remain the leading cause of death in children with congenital malformations.^[2] Only 40% of referred children with suspected heart disease had a congenital heart disease, this burden of unnecessary referrals could be minimized by improving clinical skills for recognizing these conditions. Pediatric echocardiography as diagnostic tool should be made more widely available especially in tertiary institutions to enable early diagnosis and, screening for possible cardiac defects using echocardiography during pregnancy, this can improve the outcome of some fetuses with severe cardiac malformations. parents who have a child with CHD require genetic counseling regarding the probability of cardiac malformation occurring in subsequent children. There is an urgent need for the government to establish pediatric cardiac surgical centers with specialized medical cardiology, intensive care, imaging and interventions.

REFERENCES

1. Jenkins KJ, Correa A, Feinstein JA. Non-inherited Risk Factors and Congenital Cardiovascular Defects: Current Knowledge: A Scientific Statement From the American Heart Council on Cardiovascular Disease in the Young. *Circulation*. 2007; 115(23): 2995-3014.
2. Bernstein D. Congenital heart disease in: Behrman RE, Kliegman RM, Jenson HB (eds). *Nelson's Textbook of Pediatrics*. 17ed. Philadelphia. WB Saunders CO 2004:1499-1554.
3. Brickner ME, Hillid, Lange RA. Congenital heart disease in adults. *N England J Med*. 2000; 342: 256-263.
4. Botto LD, Correa A. Decreasing the burden of congenital anomalies: an epidemiologic evaluation of risk factors and survival. *Progress in Pediatric Cardiology*. 2003; 18(2): 111-121.
5. Agomuoh DI, Akpa MR, Alasia DD. Echocardiography in the University of Port Harcourt Teaching Hospital: April 2000 to March 2003. *Niger J Med*. 2006; 15:132-136.
6. American Society of Echocardiography. Recommendations for continuous quality improvement in echocardiography. *J Am Soc Echocardiogr* 1995; 8: S1-28.
7. Grabitz RG, Joffres MR, Collins-Nakai RL. Congenital heart disease: incidence in the first year of life. *The Alberta Heritage Pediatric Cardiology Program*. *Am J Epidemiol* 1988; 128: 381-388.
8. Bosi G, Scorrano M, Tosato G, Forini E, Chakrokh R. The Italian Multicentric Study on Epidemiology of congenital heart disease: first step of the analysis. *Cardiol Young* 1999; 9: 291-299.
9. Mahmoud U, Mariya M. Spectrum of CHD in tropical environment: an echocardiography study. *J National Med. Association*. 2007; 99:665-669.
10. Alabdulgader AAA. Congenital heart disease in 740 subjects: epidemiological aspects. *Annals of tropical pediatrics*, 2001; 21(2):111.
11. Abbag F. Pattern of congenital heart disease in the south-western region of Saudi Arabia. *Annals of Saudi medicine*. 1998; 18:393-395.
12. Bhat BA. Pattern of congenital heart disease among children in Madinah Munawara. *Journal of the Saudi Heart Association*. 1997; 9:9- 16.
13. Robida A, Folger GM, Hajar HA. Incidence of congenital heart disease in Qatari children. *International J of Cardiology*. 1997; 60(1):19-22.
14. Alabdulgader A.A.A. Congenital heart disease in Saudi Arabia: current epidemiology and future projections. *East Med Health J*. 2006; 12 (Supp2)157-167.
15. Reinhold RL, Fischer A, Schneider-OJ. Congenital heart defects. Frequency at autopsy. *Teratology*. 1987; 35:305-307.
16. Gensburg LJ, Marshall EG, Druschel CM. Examining potential demographic risks factors for congenital cardiovascular malformations on a time development model. *Pediatric and peri-natal epidemiology*, 1993; 7(4):434-449.
17. Jaiyesinmi F, Antia AU. Congenital heart disease in Nigeria: a 10- year experience at UCH Ibadan. *Ann Trop Paediatr*. 1991;1:77-85.
18. Bode Thomas F, Okolo SN, Ekedigwe JE, et al. Paediatric echocardiography in Jos University Teaching hospital: Problems, prospects and preliminary audit. *Nig J Pediatrics*. 2003;30:143-149.
19. Nakazawa M, Seguchi M, Takao A. Prevalence of congenital heart disease in Japan. In: Clark EB, Takao A, eds. *Developmental cardiology: morphogenesis and function*. Mount Kisco, New York, Futura Publishing Co., 1990; 541-548.
20. Hoffman JIE. Congenital heart disease: incidence and inheritance. *Pediatric clinics of North America*. 1990, 37(1):25-43.
21. Nora JJ, Nora AH. The evolution of specific genetic and environmental counseling in congenital heart disease. *Circulation*. 1985; 57(2): 205-213.
22. MacMahon B, McKeown T, Record RG. The incidence and life expectation of children with congenital heart disease. *British Heart Journal*. 1953; 15(2):121-129.
23. Perry LW. Infants with congenital heart disease: the cases. In: Ferencz C et al., eds. *Perspectives in pediatric cardiology. Epidemiology of congenital heart disease: the Baltimore-Washington infant heart study, 1981-1989*. Mount Kisco, New York, Futura Publishing Co. 1993; 33-62.