

ORIGINAL ARTICLE
SPECTRUM OF ADULT CONGENITAL HEART DISEASE AND THE INDICATIONS FOR ECHOCARDIOGRAPHY REFERRAL IN LAGOS, NIGERIA: A SINGLE TERTIARY INSTITUTION'S EXPERIENCE

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ABSTRACT

BACKGROUND: *Echocardiography is an essential diagnostic tool for congenital heart diseases (CHD). Though present at birth, CHD are sometimes diagnosed in adult life and the diagnosis may be incidental, during evaluation of other cardiac or non-cardiac conditions. There is therefore a need to evaluate the spectrum of CHD and the reasons for referral for echocardiography.*

OBJECTIVES: *To determine the spectrum of adult congenital heart disease and the indications for referral for echocardiography.*

METHODS: *A retrospective study of all patients who had transthoracic echocardiography done at the echocardiography laboratory of the Cardiology Unit of the Lagos State University Teaching Hospital (LASUTH) over a five -year period (January 2015 to December 2019). Echocardiogram records were retrieved and analyzed using SPSS version 20.0 software.*

RESULTS: *Four thousand, eight hundred and forty (4,840) transthoracic echocardiographs were done over the 5 years of study, 129 (2.67%) had diagnosis of congenital heart disease. The mean age of the study population was 38.10 ± 17.67 years (range 12-78 years). Fifty nine (45.7%) were males while 70(54.3%) were females. The commonest CHD were ventricular septal defect (39.5%), atrial septal defect (14.7%), tetralogy of Fallot (12.4%), patent ductus arteriosus (10.9%) and bicuspid aortic valve (7.8%). About 13% of cases were diagnosed incidentally while only 21.7% of patients with CHD were referred with a clinical suspicion of CHD.*

CONCLUSION: *This study showed that ventricular septal defect is the most frequently encountered adult CHD in our center. Many patients with echocardiographic diagnosis of CHD were not referred for echocardiography with a prior clinical suspicion of CHD. We advocate more clinical surveillance for adult CHD.*

KEYWORDS: echocardiography, adult congenital heart disease, ventricular septal defect, atrial septal defect, tetralogy of Fallot, patent ductus arteriosus

INTRODUCTION

Congenital heart diseases (CHD), though present at birth, are sometimes diagnosed in adult life.¹⁻³ CHD is the most common congenital birth defect and it is estimated to affect about 1-2% of all live births.⁴ A high index of suspicion is important for early diagnosis and echocardiography is an essential diagnostic tool.⁴ Although there are regional differences in the prevalence and incidence of CHD, the global estimated incidence is about 8-10/1000 live births which represents almost about 1.35million newborns yearly.⁵ A recent study has estimated the prevalence of CHD in children to be about 6.6 per 1000 Nigerian children.⁵

The prevalence of adult congenital heart disease is on the rise probably because of the widespread use of echocardiography and also because of the recent advances and breakthroughs in the medical and surgical care of neonates born with CHD.^{2,3,6} This is promoting a growing population of adult survivors making the prevalence of adult congenital heart disease rise at a higher rate thereby outpacing the relatively dwindling prevalence of pediatric congenital heart disease.² It is estimated that adult cases now surpass children's CHD by a ratio of 2:1 in Canada.²

Clinical presentations in adult life are diverse

and complex, making suspicion for diagnosis sometimes difficult.³ CHD in children may be asymptomatic and patients may have a near-normal childhood.^{5,7} Some may present with atypical signs while some present with frank symptoms of cyanotic CHD.^{5,7} In the absence of prompt diagnosis and interventions, many patients present with severe complications like Eisenmenger's complex, heart failure, pulmonary hypertension or sudden cardiac death with diagnosis made at autopsy.^{1,8,9}

There are growing areas of interest in the management of adult CHD targeted at retardation of disease progression, management of pregnancy, preparation for non-cardiac surgery, managing arrhythmias and endocarditis prophylaxis.³ Hence, there is a need for prompt diagnosis of CHD in order to prevent negative outcomes.^{1,4} Despite these growing areas of interest, many cases of CHD are missed in early life and some cases are diagnosed incidentally or on routine echocardiography in adult life.⁶ Late presentation and delayed diagnosis are known causes of the morbidities and mortalities associated with these conditions.^{3,6}

¹⁰ Also, patients with congenital heart disease diagnosed in childhood and given life-saving surgeries, now live into adulthood.²

Though prevalence and pattern of CHD had previously been reported, there is however no study that has reported the spectrum as well as the indications and reasons for referral of patients diagnosed with adult CHD in an echocardiography laboratory in this environment.⁹⁻¹⁴ This is important in order to determine the percentage of referral that were based on prior suspicion of CHD. This study therefore set out to determine the spectrum of CHD and the reasons for referral for echocardiography in an adult echocardiography laboratory of a tertiary hospital in Lagos, Southwest Nigeria. This result of this study will add to the available

knowledge will go a long way in increasing advocacy for more surveillance of CHD among adults.

METHODS

Study design and study population

This is a retrospective descriptive study of all patients who had transthoracic echocardiogram done at the adult echocardiography laboratory of the cardiology unit of Lagos State University Teaching Hospital (LASUTH), Ikeja, over a period of five years (January 2015 to December 2019). LASUTH is the only Lagos State Government-owned tertiary health institution, attending to the over 25 million inhabitants of the metropolitan city.

Sample size determination and data collection

The minimum sample size of 100 for this study was calculated using the formula for finite population size.¹⁵ A standard normal deviation of 1.96 which corresponds to 95% confidence level and a precision of 5% were used. We also used a prevalence of 6.6% obtained from a previous study on the prevalence of congenital heart disease.⁷ A consecutive retrieval of all echocardiography records over a 5-year period met this sample size.

Echocardiography was done by all the unit Cardiologists, over the period of study, with a Sonoscape S40 machine equipped with 3.5MHz transducer with capacities to perform M-mode, two-dimensional (2D) and Doppler examinations including tissue Doppler. All measurements were taken from a standard view according to the recommendation of American Society of Echocardiography.¹⁶ Contrast echocardiography, using agitated saline, was used for some difficult-to-visualize shunts.¹⁶

Patients' demographic data, indications for referral and echocardiographic records were retrieved from their individual records, inputted and stored in a personal computer and analyzed and those with incomplete data were excluded.

Ethical consideration

Ethical approval was sought and obtained from the Lagos State University Teaching Hospital Ethics and Research Committee. Patients' identifiers were removed

Data analysis

Statistical analysis was done using the Statistical Package for Social Sciences (SPSS) version 20.0 (Chicago IL. USA). Quantitative data (age distribution) was summarized as mean \pm standard deviation, while qualitative data (diagnosis with their respective gender distributions as well as indications for referral) were summarized in frequencies and percentages. All results were presented in tables.

RESULTS

Four thousand, eight hundred and forty (4,840) transthoracic echocardiograms were done over the 5 years of the study of which 129 (2.67%) had a diagnosis of congenital heart disease, making up the study population. The mean age of the study population was 38.10 ± 17.67 years (range 12-78 years). Fifty nine subjects (45.7%) were males while 70 (54.3%) were females.

Table 1: Age and gender distribution of subjects

Age(years)	Male(n)	Female(n)	Total n (%)
11-20	14	17	31 (24.03)
21-30	18	21	39 (30.23)
31-40	10	11	21 (16.28)
41-50	9	11	20 (15.50)
51-60	4	4	8 (6.20)
61-70	3	5	8 (6.20)
71-80	1	1	2 (1.55)
Total	59 (45.7%)	70 (54.3%)	129 (100)

Table 2 showed that the commonest CHD were ventricular septal defects (VSD) in 39.5%, atrial septal defect (ASD) in 14.7%, tetralogy of Fallot (TOF) in 12.4%, patent ductus arteriosus (PDA) in 10.9%, isolated pulmonary stenosis (IPS) 7.8% and bicuspid aortic valve (BAV) 7.8%. Patent foramen ovale (PFO) 3.9%, dextrocardia and cor triatriatum were rare (1.55% each).

Table 2: Diagnosis and gender distribution of subjects

Diagnosis	Male (n)	Female (n)	Total n (%)	p-value
Ventricular septal defect	20	31	51 (39.54%)	0.67
Atrial septal defect	8	11	19 (14.73%)	0.61
Tetralogy of Fallot	6	10	16 (12.40%)	0.60
Patent Ductus Arteriosus	5	9	14 (10.85%)	0.60
Bicuspid Aortic valve	8	2	10 (7.75%)	0.48
Isolated Pulmonary Stenosis	7	3	10 (7.75%)	0.59
Patent foramen Ovale	2	3	5 (3.88%)	0.87
Dextrocardia	1	1	2 (1.55%)	0.89
Cor Triatriatum Sinistrum	2	0	2(1.55%)	0.49
Total	59	70	129 (100%)	0.48

Table 3 showed the age and gender distributions for the spectrum of all the congenital heart disease.

Table 3: Age and Gender distributions for each congenital heart disease.

	Age Distribution (years)														Total
	11-20		21-30		31-40		41-50		51-60		61-70		71-80		
	M	F	M	F	M	F	M	F	M	F	M	F	M	F	
VSD	5	5	8	14	6	8	1	4	0	0	0	0	0	0	51
ASD	0	0	0	1	4	0	3	6	1	2	0	2	0	0	19
TOF	3	5	3	4	0	1	0	0	0	0	0	0	0	0	16
PDA	2	7	3	1	0	1	0	0	0	0	0	0	0	0	14
BAV	0	0	0	0	0	0	3	0	3	0	1	1	1	1	10
IPS	3	0	2	1	0	0	0	1	0	0	2	1	0	0	10
PFO	0	0	0	0	0	0	2	0	0	2	0	1	0	0	5
Dext	0	0	1	0	0	1	0	0	0	0	0	0	0	0	2
CTS	1	0	1	0	0	0	0	0	0	0	0	0	0	0	2
Total	14	17	18	21	10	11	9	11	4	4	3	5	1	1	129

VSD=ventricular septal defect, ASD=atrial septal defect, TOF=tetralogy of Fallot, PDA=patent ductus arteriosus, BAV=bicuspid aortic valve, IPS=isolated pulmonary stenosis, PFO=patent foramen ovale, Dext= dextrocardia, CTS=cor tritratum sinistrum, M=male, F=female

Of note is that only 21.7% of these patients were referred with clinical suspicion of CHD. However, the majority (65.1%) of cases were referred on suspicion of cardiac diseases (Table 4). Other reasons for referral were non-cardiac complaints (10.08%) and routine tests (2.33%) where the diagnoses of congenital heart disease were made incidentally.

Table 4: Indications for referral for Echocardiography

Indications for Echocardiography	Male N	Female N	Total N (%)
Evaluated for Suspected Congenital Heart Disease	16	12	28 (21.70)
Evaluated for other Cardiac Disease	36	48	84 (65.12)
Evaluated for Non - cardiac Disease	6	7	13 (10.08)
Routine	0	3	3 (2.33)
Not stated	1	0	1 (0.78)
Total	59	70	129 (100)

DISCUSSION

The objective of this study was to determine the spectrum of adult CHD and the reasons for referral for echocardiography in the adult echocardiography laboratory of Lagos State University Teaching Hospital.

CHD occurred in 2.6% of all echocardiography done which was similar to previous works of Adebayo et al and Ogah et al in Southwest-Nigeria but less than that of Mahmud Sani et al (9.3%) in Northern Nigeria^{9,11,13}. This disparity may have been as a result of the age difference of the population, as many of their subjects were less than 35 years of age⁹. Studies in the pediatric population have reported different prevalence rates of CHD depending on the population studied.^{5, 7, 17, 18} Also, the high prevalence of congenital heart disease in

Northern Nigeria may be attributed to younger maternal age, malnutrition and low vaccination rates among these young mothers.⁴ This study showed that more females were affected with congenital heart disease which is similar to previous studies on congenital heart diseases.^{5,9,14,17,18} This may however be adduced to the fact that there were more females in the overall population of the echocardiograms done in the study period.

Previous studies by Kolo et al and Buba et al reported a high prevalence of VSD and TOF compared to other CHDs.^{12,19} Our work also showed that VSD was the commonest adult congenital heart diseases reported followed by atrial septal defects and tetralogy of Fallot. Studies in the pediatric population by Ekure et al, Otaigbe et al and Ibadin et al reported a similarly high prevalence of VSD.^{5,14,18} Bicuspid

aortic valve had previously been referred to as the commonest congenital heart disease in the Caucasian however this was not so in our study.²⁰

²¹ Bicuspid aortic valve was not very common in this study. A low prevalence of bicuspid aortic valve had also been reported in other studies done in this environment and may therefore be due to epidemiological differences in disease conditions.^{9,12-14}

The main indications for echocardiography in the pediatric population as reported by Bode-Thomas et al were rheumatic valvular heart diseases, CHD and for evaluation of cardiac murmurs.¹⁷ However, our study showed that the main indication for echocardiography was for other cardiac reasons and not primarily for suspicion of CHD. Less than a quarter of our study population was referred for echocardiography on account of a suspected CHD while more than 10% were referred on account of non-cardiac related issues. About 2.33% of our patients had the diagnosis of CHD made on routine screening. These routine screening included individuals who had pre-employment investigations and those who wanted echocardiogram done prior to rigorous sporting activities. Incidental diagnosis of adult CHD had also been reported in about 1.05% of Koreans on routine screening of 27,897 patients.²²

The guidelines recommend that echocardiography is mandatory for clinical diagnosis of CHD.^{10, 16} Also, echocardiography remains crucial to the overall surveillance of patients with adult CHD because it provides a comprehensive assessment of the morphology, physiology, pathophysiology and function of the heart.^{10, 16} In spite of the foregoing, the clinical suspicion necessitating referral for echocardiography in patients with CHD seems to be low in this study. The appropriate use of echocardiography has been shown to contribute immensely to the overall clinical outcome of patients with CHD.^{2,3,6}

The pediatric population seems to receive more clinical suspicion and a higher rate of diagnosis of CHD probably because of the congenital defect screening at birth and because patients present with more classical clinical features than adults.^{5, 7, 17} This may imply that there is more awareness of CHD in pediatric practice than in adult medicine even though the population of adult patients with CHD may overwhelm the existing health structure in the near future.¹⁻³

Complex live-saving cardiac surgeries have made many otherwise fatal congenital heart diseases diagnosed early in life survive into adulthood, however, this study did not identify any subject with prior surgical intervention. This may be due to the fact that this is a retrospective study and some data may have been incomplete or lost and thus excluded or that many of these complex childhood surgeries are yet to be routinely done in this environment, and so it is possible that many of these patients die in childhood.

There is thus a need to increase awareness of CHD among physicians so as to have a high index of suspicion for the diagnosis of CHD. This will make early intervention possible in order to reduce adverse outcomes of CHD. Also, a multicenter prospective study will be needed to further corroborate the findings.

CONCLUSION

This study showed that ventricular septal defects, atrial septal defects and tetralogy of Fallot are the most frequently encountered adult CHD in our center.

The majority of adult patients with CHD were not referred for echocardiography based on a prior clinical suspicion of CHD; rather many CHD diagnoses were made incidentally.

We advocate more clinical surveillance of CHD in adult patients.

CONFLICT OF INTEREST

There are no conflicts of interest

FINANCIAL DISCLOSURE

The authors have none to declare

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