Spectrum of Congenital Heart Disease in a Tropical Environment: An Echocardiography Study

Mahmoud U. Sani, MBBS, FWACP; Mariya Mukhtar-Yola, MBBS, FMCpaed; and Kamilu M. Karaye, MBBS, MSc, FWACP

Echocardiography is a major mode of cardiovascular imaging with versatile applications. Modern two-dimensional echocardiographic techniques provide a comprehensive means for evaluating virtually all forms of congenital heart disease (CHD) found in both adults and children. CHD is an abnormality in cardiocirculatory structure or function that is present at birth, even if it is discovered much later. We set out to describe the spectrum of CHD using echocardiography in two centers in Kano, northern Nigeria.

In this retrospective study, transthoracic echocardiography (TTE) data collected from two echocardiography laboratories in Kano over a period of 48 months (June 2002 to May 2006) were reviewed. Patients with diagnosis of congenital heart disease were selected. Information obtained from the records included the age, gender, clinical diagnosis and echocardiographic findings.

One-hundred-twenty-two patients had CHD, making 9.3% of the 1,312 patients with abnormal echocardiograms. There were 73 males and 49 females (ratio 1.5:1); and their ages ranged from nine days to 35 years. Forty-one (33.6%) children presented for echocardiography before the age of one year, and 69% presented before the age of five years. Thirteen (10.6%) were ≥18 year. Ventricular septal defect (VSD) was the most common echocardiographic diagnosis present in 56 patients (45.9%). Thirty-two (26.2%) had tetralogy of Fallot, and 15 (12.3%) had atrial septal defect (ASD). Ten (8.2%) had endocardial cushion defect, and nine (7.4%) had other congenital heart abnormalities. Coarctation of the aorta and aortic stenosis were rare.

CHD is a common cardiovascular problem in our setting, and a number of patients were diagnosed in adulthood. With increasing availability of echocardiographic facilities, more cases of CHD are likely to be identified early.

Key words: congenital heart disease ■ echocardiography ■ ventricular septal defect

 From the Cardiology Unit, Department of Medicine (Sani, senior lecturer/consultant physician and cardiologist; Karaye, lecturer I/consultant physician and cardiologist) and Neonatology Unit, Department of Pediatrics (Mukhtar-Yola, lecturer I/consultant pediatrician and neonatologist), Aminu Kano Teaching Hospital, Kano, Nigeria. Send correspondence and reprint requests for *J Natl Med Assoc.* 2007;99:665–669 to: Dr. Mahmoud U. Sani, Cardiology Unit, Department of Medicine, Aminu Kano Teaching Hospital, PMB 3452, Kano, Nigeria; phone: 234 803 3479179, 234 64 971434; fax: 234 64 663354; e-mail: sanimahmoud@yahoo.com

INTRODUCTION

Congenital heart disease (CHD) is defined as an abnormality in cardiocirculatory structure or function that is present at birth, even if it is discovered much later.[1] These malformations remain one of the most frequent birth defects, with a live born prevalence of about 5–8 per 1,000 (0.5–0.8%) live born infants in western countries, where such prevalence data are tracked.[2-5] Since the introduction of fetal echocardiography almost 20 years ago, fetal surveillance from numerous centers indicates that among some forms of congenitally malformed hearts, there is increased fetal death.[6] Thus, for certain congenital heart malformations, the live born prevalence is lower than the fetal prevalence. In the United States, the incidence of CHD in stillborns is 3–4%, in abortuses 10–25% and in premature infants 2% [excluding patent ductus arteriosus (PDA)]. About 2–3 in 1,000 newborns will be symptomatic with heart disease in the first year of life. The diagnosis is established by one week of age in 40–50% of patients with CHD and by month of age in 50–60% of patients.[7]

CHD causes the deaths of thousands of children in developing countries. Gupta and Antia estimated the incidence of congenital heart defects in Nigeria to be 3.5 per thousand.[8] This value is high considering the rate of deliveries in the country. Due to inadequate diagnostic facilities, only a minority of cases are detected, and often at late stages of the diseases.[9]

Over the past 20–30 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 noninvasive technique for the investigation of cardiac diseases with the advantages of reproducibility of results, instantaneous images and reliable levels of accuracy.[10] Modern two-dimentional echocardiographic techniques provide a comprehensive means for evaluating virtually all

forms of CHD found in both adults and children.[11] Additionally, it can be used to evaluate repaired and palliated CHD. There are limited data on the echocardiographic findings in children with heart disease in Nigeria, especially from the northern part. This is attributable to the fact that pediatric echocardiography has not been widely available in this country. Even when available, the optimal use of this diagnostic tool is often hampered by factors such as cost, scarcity of skilled personnel and the absence of appropriate probes. We set out to describe the spectrum of CHD seen in two echocardiographic centers in Kano, northern Nigeria.

PATIENTS & METHODS

In this retrospective study, transthoracic echocardiography (TTE) data collected over 48 months (June 2002 to May 2006) were reviewed. Patients with echo diagnosis of CHD were selected. The study was carried out at the echocardiography laboratory of Aminu Kano Teaching Hospital and a private echocardiography laboratory, both located in Kano. During the study period, the two echocardiography laboratories served all the echo requests in the state in addition to receiving referrals from three neighboring states. Kano, the study site, is one of the most populous states in Nigeria, with a population of about 10 million people.

Information obtained from the records included age, gender, names of referring hospital/physician, clinical diagnosis and echocardiographic findings. Data was analyzed using SPSS® version 10.0 software.

Echocardiographic modalities applied included M-Mode, two-dimensional and Doppler studies. Two-dimensional and Doppler echocardiography were done with 5-MHz sector transducer. Complete two-dimensional echocardiography examination was performed according to the recommendations of the American Society of Echocardiography (ASE).[12] Doppler indices of LV diastolic filling were obtained. Complete Doppler study was done according to the recommendations of the ASE.[13]

RESULTS

A total of 1,499 echocardiographic examinations were done in the two centers over the four-year study period. Two-hundred-ninety-one patients were ≤18 years. Of the total number that had echocardiograms, 1,312 (87.5%) had abnormal findings, of which 122 (9.3%) had CHD. There were 73 males and 49 females (ratio 1.5:1); and their ages ranged from nine days to 35 years. Table 1 shows the age and sex distribution of the patients. Forty-one (33.6%) children presented for echocardiography before the age of one year, and 69% presented before the age of five years.

The main clinical indications for the echocardiography included CHD (unspecified) 44 (36.1%), ventricular septal defect (VSD), 37 (30.3%), congestive cardiac failure 12 (9.7%), tetralogy of Fallot (TOF) eight (6.6%), PDA five (4.1%), failure to thrive four (3.3%), Down's syndrome six (5.0%), congenital rubella syndrome four (3.3%) and dextrocardia two (1.6%). The test was requested by a wide range of medical practitioners, including general practitioners, Physicians (noncardiologists) and cardiologists.

VSD was the most common echocardiographic diagnosis, which occurred in 56 (45.9%) of the patients, followed by TOF 32 (26.2%), atrial septal defect (ASD) 15 (12.3%) and then endocardial cushion defect 10 (8.2%). The distribution of the various CHDs according to sex is shown in Table 2. Of the 56 cases of VSD, 49 (87.5%) were membranous, two (3.6%) were in the muscular interventricular septum and five (8.9%) were of the maladie de Rogers type. There were 15 patients with ASD, out of which 12 (80%) were of the secundum type, two (13.3%) were primum type, and one (6.67%) was a sinus venosus defect.

Eleven patients had Eisenmenger complex, all of whom were older children and young adults. Three of them had VSD, three had ASD, and four had endocardial cushion defect.

Table 3 shows CHD among patients ≥18 years. Six of the 13 (46.2%) had TOF; one had TOF and secundum ASD (Fallots pentology); and another had TOF, ASD and cor triatriatum sinister.

Four patients (three VSD, one TOF) came back for a postoperative echocardiogram after surgical intervention abroad.

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

Age (Years)	Males	Females	Total (%)
<1	26	15	41 (33.6)
1–4	28	15	43 (35.2)
5–9	7	6	13 (10.6)
10–14	4	7	11 (9.0)
15–19	4	2	6 (4.9)
20–24	1	2	3 (2.4)
>25	3	2	5 (4.1)
Total	73	49	122 (100)

DISCUSSION

This study highlights the common variants of CHDs seen in Kano. Acyanotic CHD was more common than cyanotic heart disease. The relative frequencies of individual CHD are consistent with reports from other parts of Nigeria[14-16] and the world,[17-21] with VSD being the most common acyanotic lesion and TOF the most common cyanotic lesion. VSD defect accounted for 56 (45.9%) of all the cases, making it the most common CHD in this series, with membranous VSD accounting for 87.5% of all the cases. Small VSDs are usually benign and frequently decrease in size, and 30–50% undergo spontaneous closure during the first two years of life. Small muscular defects are, however, more likely to close (80%) than membranous VSDs (35%).[7] Larger defects may lead to congestive heart failure and infants succumb in the early months of life either to cardiac failure or to superimposed respiratory infection.[22] Those who survive infancy but have marked pulmonary hypertension will develop increasing pulmonary vascular resistance. This leads ultimately to shunt reversal together with the late onset of cyanosis, usually after the age of 5 years and sometimes not until early adult life.

About 2–3 in 1,000 newborns will be symptomatic with heart disease in the first year of life. The diagnosis is established by one week of age in 40–50% of patients with CHD and by one month of age in 50–60% of patients in the western world, where diagnostic facilities are readily available.[7] In this study, a total of 41 (33.6%) children presented for echocardiography before the age of one year, and 69% presented before the age of five years. This late presentation often worsens the prognosis for survival. Unfortunately, this is the scenario that operates in most developing countries because of delayed diagnosis, and lack of skilled personnel, equipment and facilities for surgery. Eleven (9%) patients had Eisenmenger complex secondary to VSD, ASD and endocardial cushion defect. These were older children and young adults with shunt reversal. These patients would have benefited from early surgery to prevent this complication. With advances in both palliative and corrective surgery in the last 20 years in developed countries, the number of children with CHD surviving to adulthood has increased dramatically. However, despite these advances, CHD remains the leading cause of death in children with congenital malformations.[7]

There was a general male preponderance of 1.5:1, which is consistent with reports from Reinhold et al.[23] and Akang et al.[24] Coarctations of the aorta, and aortic and pulmonary stenosis were uncommon findings in this study. Other reports from Africa had similar findings.[14,17]

Though the incidence of CHD in the tropics may not be different from that of the rest of the world, the relative frequencies of the individual defects may vary in different races. Congenital aortic stenosis and coarctation of the aorta are rarer in the black race than in the Caucasian race.[25]

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

Type of CHD	Males	Females	Total (%)
VSD	31	25	56 (45.9)
TOF	21	11	32 (26.2)
ASD	8	7	15 (12.3)
Endocardial cushion defect	7	3	10 (8.2)
PDA	0	3	3 (2.5)
Congenitally corrected TGV	1	0	1 (0.8)
Bicuspid aortic valve	1	0	1 (0.8)
Others	3	1	4 (3.3)
Total	73	49	122 (100)

CHD; congenital heart disease; VSD; ventricular septal defect; TOF; tetralogy of Fallot; ASD; atrial septal defect; PDA; patent ductus arteriosus; TGV; transposition of great vessels

Table 3. Distribution of congenital heart disease by gender among patients >18 years

Type of CHD	Males	Females	Total
TOF	4	2	6
PDA	0	2	2
VSD	1	1	2
Congenitally corrected TGV	1	0	1
TOF & ASD	1	0	1
TOF, ASD & cortriatriatum	1	0	1
Total	8	5	130

CHD; congenital heart disease; TOF; tetralogy of Fallot; PDA; patent ductus arteriosus; VSD; ventricular septal defect; TGV; transposition of great vessels; ASD; atrial septal defect

The etiology of malformations of the heart and great vessels is not known, although certain relationships have been established in some defects. Maternal rubella in the first trimester of pregnancy results in a high incidence of cardiac defects such as PDA, or pulmonary stenosis in the fetus. Certain chromosomal defects such as trisomy 21, 18 are associated with specific cardiac defects. In the majority of cases, however, the exact cause is unknown and believed to be multifactorial: drugs, genetics, parental age and the environment. This study did not set out to establish the etiological factors in patients with CHD; however, the clinical indication for echocardiography in this study was Downs syndrome in six (5%) patients (four cases of VSD, two cases of endocardial cushion defect) and congenital rubella syndrome in four (3.3%) patients (one case of PDA, three cases of VSD).

CHD was diagnosed in 13 (10.7%) patients who were aged >18 years. These patients presented for echocardiography for the first time at this late age, probably due to difficulties in accessing care and the availability and cost implications of investigations.

To our knowledge, only four patients had access to surgery, and this was done abroad because of nonavailability of such centers in the country. These surgeries were paid for partly by the parents assisted by some nongovernmental associations (NGOs) such as Save a Child's Heart and the Kanu Heart Foundation. Although these NGOs assist in transporting few patients abroad for surgery, the demand for such treatment is more than they can cope with. Therefore, our healthcare planners must be convinced of the need not only to control communicable diseases but also to allocate resources for the diagnosis and treatment of noncommunicable diseases such as CHD that have for too long been neglected in our healthcare delivery system. Centers of excellence for cardiac surgery should be instituted to enable early diagnosis and treatment.

Parents who have a child with CHD require genetic counseling regarding the probability of a cardiac malformation occurring in subsequent children. In the United States, where such data is available, the incidence of CHD in the normal population is 0.8%, and this incidence increases to 2–6% for a second pregnancy after the birth of a child with a CHD or if a parent is affected.[7] When two first-degree relatives have CHD, the risk for a subsequent child may reach 20–30%.[7] With the establishment of more tertiary healthcare institutions in Nigeria, there will be an increasing number of pediatric cardiologists and echocardiographic facilities in the near future; therefore, more cases of CHD are likely to be identified earlier. Pediatric echocardiography as a diagnostic tool should be made more widely available, especially in tertiary institutions to enable early diagnosis. Centers lacking purpose-built echocardiography equipment could in the short term improvise by using the general-purpose ultrasound machines that are currently available in many centers. Although they are usually fitted with the larger abdominal probes that may not be ideal for the pediatric chest and may not be equipped to provide detailed hemodynamic information, such machines are still useful in the confirmation of simple abnormalities such as large septal defects and effusions.

There is an urgent need for the government to establish a well-equipped cardiothoracic surgical center to cater for these patients either free of charge or at highly subsidized rates.

REFERENCES

1. Brickner ME, Hillis LD, Lange RA. Congenital Heart Disease in adults: I. *N Engl J Med.* 2000;342:256-263.

2. Ferencz C, Rubin JD, Loffredo CA, et al. Epidemiology of congenital heart disease: the Baltimore-Washington Infant Study 1981–1989. In: Anderson RH, ed. Perspectives in Pediatric Cardiology. Mount Kisco, NY: Futura Publishing; 1993:353.

3. Carlgren LE. The incidence of congenital heart disease of children born in Gothenberg, 1941–1950. *Br Heart J.* 1959;21:40–50.

4. Fyler DC, Buckley LP, Hellenbrand WE, et al. Report of the New England regional infant cardiac program. *Pediatrics.* 1980;65(suppl):375–461.

5. 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.

6. Hoffman JIE. Incidence of congenital heart disease: II. Prenatal Incidence. *Pediatr Cardiol.* 1995;16:155–165.

7. Daniel Bernstein. Congenital heart disease. In: Nelson Textbook of Paediatrics. Behrman RE, Kliegman RM, Jenson HB, eds. 17th ed; 2004:1501-1509.

8. Gupta B, Antia AU. The incidence of congenital heart disease in Nigerian children. *Bt Heart J.* 1967;29:906-909.

9. Jaiyesimi F. Congenital cardiac malformations: a serious health problem in Nigeria? *Nig J Paediatr.* 1982;9:67–70.

10. 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.

11. Hope UC, Dederichs B, Deutsch HJ, et al Congenital heart disease in adults and adolescents: comparative value of transthoracic and transesophageal echocardiography and MR imaging. *Radiology.* 1999;199:669-677.

12. Henry WL, DeMaria A, Gramiak R, et al. Report of the American Society of Echocardiography (ASE) Committee on Nomenclature and Standards in Two Dimensional Echocardiography. *Circulation.* 1980;62:212-217.

13. Quinones MA, Otto CM, Stoddard M, et al. Recommendations for Quantifications of Doppler Echocardiography. A report from the Doppler Quantification Task Force of the Nomenclature and Standards Committee of the American Society of Echocardiography. *J Am Soc Echocardiogr.* 2002;15:167-184.

14. Jaiyesinmi F, Antia AU. Congenital heart disease in Nigeria: a 10-year experience at UCH Ibadan. *Ann Trop Paediatr.* 1991;1:77-85.

15. Olowu AO. Clinical profile of congenital heart disease in Sagamu. *Nig J Paediatr.* 1995;22:26-29.

16. Bode Thomas F, Okolo SN, Ekedigwe JE, et al. Paediatric echocardiography in Jos University Teaching hospital: Problems, prospects and preliminary audit. *Nig J Paediatr.* 2003;30:143-149.

17. Alarbbulgader AA. Congenital heart disease in 740 subjects: epidemiological aspects. *Heart.* 2004; 90:1321-1325.

18. Miyague NI, Cardoso SM, Meyer F, et al. Epidemiological study of congenital heart defects in children and adolescents. Analysis of 4,538 cases. *Fam Pract.* 2000;17:394-4000

19. Jaiyesinmi F, Ruberu DK, Misra VK. Pattern of congenital heart disease in King fahad Teaching Hospital Buraidah. *Ann Saudi Med.* 1993;13:407-411.

20. Bannerman CH, Mahalu W. congenital heart disease in Zimbabwean children. *Ann Trop Paediatr.* 1998;18:5-12.

21. Hoffman JI, Christianson MA. Congenital heart disease in cohort of

19,502 births with long term follow up. *Am J Cardiol.* 1978;42:641-647.

22. Antia AU, Wilkinson JL, Jaiyesinmi F. The Cardiovascular System. In: Paediatrics in the tropics. Hendricksen RG, Barr GD, Matthews TS, eds. Blackwell Scientific Publications; 1991:240-242.

23. Reinhold RL, Fischer A, Schneider-OJ. Congenital heart defects. Frequency at autopsy. *Teratology.* 1987; 35:305-307.

24. Akang EE, Osinusi KO, Pindiga HU, et al. Congenital malformations: A review of 672 autopsies in Ibadan, Nigeria. *Zentralbl Allg Pathol.* 1987;133:253-261.

25. Okoroma EO. Congenital Heart Disease and Arrhythmias. In: Paediatrics and Child Health In A Tropical Region. Azuibuike JC, Nkangieneime KEO, eds. African Education Service; 1999:273-275. ■