INTRODUCTION

Acquired heart diseases (AHD) known to have their origins predominantly in childhood are still major public health problems in the developing world. The patterns of childhood AHD also vary in different parts of the world. Reports dating back three or four decades show that “idiopathic cardiomegaly,” endomyocardial fibrosis (EMF), rheumatic heart disease (RHD) and infective pericarditis were the most commonly encountered AHD among Nigerian children. More recent reports indicate that there may not only be geographical differences in patterns of disease within the country, but that some diseases, notably EMF, may be disappearing. The reports on childhood AHD from North-Central Nigeria have been preliminary. In this 10-year retrospective review of our echocardiography records, we compare the pattern of childhood AHD in our center in North-Central Nigeria to the historical and contemporary patterns reported from other parts of the country, and aim to highlight possible regional differences and changes in trend.

MATERIALS AND METHODS

We reviewed our pediatric echocardiography records over a 10-year period from December 1999, when the service became available at our center, the Jos University Teaching Hospital (JUTH), Jos, Plateau State, until November 2009. The pediatric cardiology unit of JUTH receives referrals from several states in North-Central Nigeria, parts of North-Western and North-Eastern Nigeria, and also from the Federal Capital Territory. All children referred to the unit for management or for echocardiography (ECHO)
undergo clinical, radiologic (chest radiograph), and electrocardiographic (ECG) evaluation before an ECHO is performed.

During the period under review, three different ECHO machines were in use in the hospital. From December 1999 to July 2000, a Hewlett-Packard (HP) Sonos 1250 was used. The second and third machines, a HP Sonos 1500 and a Logic Expert GE (General Electrics) ultrasound system, were in use from July 2000 to February 2006 and from July 2007 to November 2009 respectively (no machine was available for a 16-month period). All three machines had facilities for two-dimensional (2D), M-mode and color flow Doppler imaging. The latter two also had facilities for pulsed and continuous wave Doppler.

Depending on the age and size of the child and the quality of image produced, 3.5 or 5 MHz frequency transducers were used for the majority of children examined. Occasionally a 2.5 MHz frequency transducer had to be used for an older child. The number of pediatric ECHO procedures performed was limited by factors such as the availability of skilled personnel, cost of the procedure, and periods of equipment breakdown.9 Multiple users (the same machines were used by various other services and departments in the hospital) and unstable power supplies were other factors that not only limited the access of pediatric patients to echocardiography but also contributed to recurrent breakdown of equipment.9

Over the 10-year period, up to four different pediatricians performed ECHO examinations at different times (each after a period of training and supervision) and after the first few years, at least two of them were usually present at each ECHO session. Echocardiographic diagnoses were based on standard guidelines and on the diagnostic criteria for each acquired heart disease (AHD).13-17 Rheumatic heart disease (RHD) was defined by the presence of any definite evidence of valve regurgitation or stenosis seen in two planes on Doppler examination, and at least two morphologic abnormalities, such as restricted leaflet mobility, focal or generalized valvular thickening, and abnormal subvalvar thickening of the affected valve.14 In the presence of poor LV contractility (fractional shortening less than 28%), dilated cardiomyopathy (DCM), another frequently encountered lesion, was diagnosed if the left ventricular (LV) dimensions were above the upper limit of normal for age.16 While nondilated cardiomyopathy (NDCM) was diagnosed if the LV dimensions were within normal limits for age.17 Information obtained from our records included age at the time of ECHO, gender, clinical indication for the ECHO, and the echocardiography findings and diagnosis. This paper is a retrospective review based primarily on ECHO diagnosis. Thus children with normal ECHO were excluded, except there was irrefutable clinical evidence of an acquired heart disease such as infective endocarditis or Kawasaki disease.18,19

The data were analyzed using the statistical software Epi Info® version 3.5.1 of the Centres for Disease Control (CDC) Atlanta, Georgia, USA. Descriptive statistics including means, standard deviations, and frequencies were generated and stratified by age, gender, and heart lesions as appropriate. Means of continuous variables were compared using Student’s t-test while proportions were compared using the chi-square test. The level of statistical significance was set at P<0.05. Permission to publish the data was obtained from the hospital ethical committee.

RESULTS

A total of 580 children aged 2 weeks to 18 years had echocardiographic examinations performed during the period covered by this report, of which 167 (28.8%) had various forms of acquired heart disease (AHD) as their primary diagnoses, 390 (67.2%) had congenital heart disease (CHD) only, 7 (1.2%) children had both AHD and CHD, while 16 (2.8%) had normal ECHO findings. One of the latter, a female infant, had irrefutable clinical features of Kawasaki disease. The seven children with coexisting CHD and AHD were RHD with atrial septal defect (ASD) – two cases; dilated cardiomyopathy (DCM) with ASD – two cases; and three cases of infective endocarditis associated with patent ductus arteriosus, congenital mitral incompetence and Fallot tetralogy respectively.

Thus a total of 175 children were diagnosed with AHD during this period. The commonest indications for ECHO in these children are displayed in Table 1. They comprised 80 (45.7%) males and 95 (54.3%) females and were aged between 4 weeks and 18 years (mean 9.84±4.5 years). There was no significant age difference between males (9.6±4.5 years) and females (9.9±4.5 years) – P=0.31. The relative frequencies, sex distributions, and mean ages of

Table 1: Indications for echocardiography in 175 children with acquired heart disease

| Indication          | Heart lesion | Percentage |
|---------------------|--------------|------------|
| Suspected RHD       | 43           | 23.4       |
| CCF                 | 40           | 22.9       |
| Heart murmur        | 31           | 17.7       |
| Cardiomegaly        | 13           | 7.4        |
| Recurrent chest pain| 8            | 4.6        |
| Suspected Cor pulmonale | 8 | 4.6       |
| Pericardial effusion| 7            | 4.0        |
| Suspected EMF       | 3            | 1.7        |
| Palpitations        | 2            | 1.1        |
| Arrhythmia          | 2            | 1.1        |
| DCM                 | 2            | 1.1        |
| Myocarditis         | 2            | 1.1        |
| Kawasaki Disease    | 1            | 1.1        |
| Missing data        | 15           | 8.0        |
| **Total**           | **375**      | **100**    |

RHD - Rheumatic heart disease, CCF - Congestive heart failure, EMF - Endomyocardial fibrosis
children with different acquired heart lesions are displayed in Tables 2-4.

**Rheumatic heart disease**
Rheumatic heart disease (RHD) was the most common AHD encountered. It was diagnosed in 101 (57.7%) children, including the two with coexistent atrial septal defect (ASD). No RHD patient was less than 5 years old and they were significantly older than those with other acquired heart diseases (10.8±3.2 versus 7.7±5.4 years) – *P* < 0.001 [Tables 2 and 4]. The mitral valve (MV) was the most commonly affected valve – in 95% (96 cases) of the 101 RHD patients and in 96 (47.5%) of a total of 202 diseased valves. The most common isolated valve lesion was mitral incompetence (MI) which was present in 34 (32.3%) of the 101 patients with RHD while the least common was tricuspid stenosis (TS), seen in only one patient (0.95%). Furthermore, regurgitant lesions predominated while mixed valvular lesions (incompetence plus stenosis) were more frequent than isolated stenosis [Table 5]. No patient had isolated pulmonary valve pathology. All four valves were diseased in 16 (15.5%) of patients [Figure 1]. All the mitral and aortic valve lesions as well as the isolated tricuspid stenosis were associated with leaflet or cusp thickening or deformities, while majority (64.8%) of the tricuspid and pulmonary incompetences were not associated with these abnormalities, suggesting they were predominantly functional, or secondary to other valve abnormalities.

**Cardiomyopathies**
Cardiomyopathy was present in 37 (21.1%) subjects. Those with dilated cardiomyopathy (DCM) were slightly younger (mean age 6.8±4.9 years) compared with other acquired heart diseases and accounted for 33 (89.2%) of all subjects with cardiomyopathy [Tables 3 and 4]. Eight (21.6%) of the children with ECHO features of DCM (dilated and poorly contractile left ventricles) were thought to have possible etiologies were found. However, viral pericarditis was considered very likely in the absence of any other systemic disease manifestations.

**Other acquired heart diseases**
Other AHD diagnosed included cor pulmonale and pericardial disease in 16 (9.1%) and 15 (8.6%) respectively. Infective endocarditis was diagnosed in eight (4.4%) subjects; it was associated with rheumatic heart disease in four children, congenital heart disease in three and dilated cardiomyopathy in one child. Rarer forms of AHD encountered were aortic aneurysm and nondilated cardiomyopathy (two subjects each), and one each of endomyocardial fibrosis, arrhythmogenic right ventricular dysplasia and Kawasaki disease – Table 3.

Cor pulmonale was secondary to pulmonary tuberculosis in four children (two of whom also had human immunodeficiency virus (HIV) infection), sickle cell anemia in two and to poorly treated bacterial pneumonia and suspected broncho-pulmonary dysplasia (the latter in a former preterm) in one case each. The aetiology in the other eight children could not be ascertained.

The 15 patients with pericardial disease included two (13.3%) cases each of constrictive pericarditis (with pulmonary tuberculosis), myo-pericarditis (massive effusions in association with poorly contractile ventricles) and possible pyogenic pericardial effusion (ECHO evidence of organization of the effusion). In the remaining nine children, no direct pointers (clinical or ECHO) to the possible etiologies were found. However, viral pericarditis was considered very likely in the absence of any other systemic disease manifestations.

| Age group (years) | Heart lesion | Male N (%) | Female N (%) | Total N (%) | χ² | P value |
|------------------|--------------|------------|--------------|-------------|-----|---------|
| 0-4              | RHD          | 0 (0)      | 0 (0)        | 0 (0)       | —  | —       |
|                  | Other        | 19 (10.9)  | 33 (17.4)    | 52 (28.1)   | 4.24| 0.04    |
| 5-9              | RHD          | 11 (6.3)   | 22 (12.0)    | 33 (18.9)   | —  | —       |
|                  | Other        | 7 (4.0)    | 3 (1.7)      | 10 (5.4)    | 0.63| 0.87    |
| 10-14            | RHD          | 25 (14.6)  | 28 (16.0)    | 53 (29.3)   | —  | —       |
|                  | Other        | 9 (5.4)    | 10 (5.7)     | 19 (10.3)   | 0.65| 0.83    |
| 15-18            | RHD          | 5 (2.9)    | 9 (5.1)      | 14 (7.6)    | —  | —       |
|                  | Other        | 4 (2.3)    | 6 (3.4)      | 10 (5.3)    | 2.70| 0.10    |
| Total            | RHD          | 41 (24.0)  | 59 (34.5)    | 100 (58.3)  | —  | —       |
|                  | Other        | 39 (22.8)  | 32 (18.7)    | 71 (41.5)   | —  | —       |

RHD - Rheumatic heart disease, Other - Other acquired heart lesions, PE - Pericardial disease, DCM - Dilated cardiomyopathy; Four female subjects had no age records (one each of RHD, PE, DCM, endocarditis)
Four of the eight children with infective endocarditis had visible vegetations. These were on the anterior mitral leaflets of a 12-year-old girl with multivalvular RHD, a 9-year-old boy with NDCM and a 2-year-old girl with DCM; the fourth was in the main pulmonary artery of a boy with a patent ductus arteriosus. The other four children with no vegetations had underlying congenital (two boys) or rheumatic (two girls) heart disease but with compelling clinical features of infective endocarditis. All four eventually responded to prolonged high dose intravenous broad-spectrum antibiotic therapy, but only one of them had a positive blood culture (this was a 9-year-old boy with Fallot tetralogy whose blood culture grew Staphylococcus aureus).

### DISCUSSION

Although based only on patients that came to specialist medical attention and thus not representative of the community prevalence of the various acquired heart diseases (AHD) in the region, our data nevertheless provide insight into the types of AHD common among children in Jos, North Central Nigeria, and by extension the rest of the region. They also provide a basis for comparison with other populations.
similar studies; for example, we found relatively fewer children with AHD than with congenital heart disease (CHD) – in tandem with recent similar studies from other parts of the country and older studies from some developed countries. It however contrasts with community-based data from Egypt as reported by Refat et al., who screened 8,000 school children and found predominantly rheumatic heart disease (RHD). The difference highlights the afore-mentioned varying emphases of hospital-based studies such as ours (which largely reflect the healthcare seeking behavior of the population) compared with community-based ones that better reflect disease epidemiology. This conclusion may however not be as straightforward as might at first appear.

School-age children are recognized as having the highest risk and prevalence of RHD but only the symptomatic ones are likely to present in hospital. Thus milder cases may not be captured in a hospital-based study. Also, the natural history of CHD is such that its prevalence reduces with age since the most severe cases die early without intervention. Others develop dramatic symptoms at a young age, prompting their families to seek early medical attention. RHD on the other hand, may evolve rather insidiously, with many patients presenting to hospital only when the disease is relatively advanced or complications have set in. Finally, cases from a wide area (whether congenital or acquired) would tend to congregate, but in a random manner, in a referral center such as ours. All these underscore the fact that hospital and community-based studies both have their different uses, with the former providing useful information in terms of disease pattern while the latter may provide more reliable data for prevalence studies and estimation of disease burden.

Like Refat et al., we found more girls with AHD than boys, particularly rheumatic heart disease (RHD) in the age range 10–18 years. RHD has been reported by various other authors to be more severe and more likely to occur in females. In consonance with some other Nigerian authors, we found RHD to be the most frequent AHD in children in our center. However, a report from Lagos cited effusive pericarditis as the dominant AHD among children and RHD as the second most frequent. The trend in the present report is also similar to that from other parts of the developing world and is an indication of the persistence of the poor social conditions that favor transmission of the group A β haemolytic streptococcal pharyngitis that leads to rheumatic fever (RF) and RHD. By contrast, Kawasaki disease (KD) is now the most frequent AHD affecting children in developed countries such as the USA and Japan. Our clinical diagnosis of Kawasaki disease in only one child might be indicative either of a true rarity of the condition in our environment or of a very low index of suspicion among physicians and other health workers. Our patient presented to specialist attention at a very early stage of the disease and was treated vigorously with aspirin which resulted in rapid and complete resolution of all signs and symptoms. The clinical cardiovascular and echocardiography findings in this child were normal throughout the acute phase of the illness and for several months thereafter. Coronary aneurysms, the major ECHO feature of Kawasaki disease, are seen only in the later stages and are usually asymptomatic. Thus children diagnosed with Kawasaki disease must be consistently followed up for several months and screened thoroughly for coronary aneurysms because of the risk of sudden death from coronary thrombosis and myocardial infarction. Kawasaki disease however has its highest incidence in people of Asian ancestry.
majority of the cases are usually of viral aetiology.²¹ Eight of our patients with DCM had preceding symptoms of acute upper respiratory tract infection prior to the onset of congestive heart failure (CHF), suggesting the possibility of viral myocarditis.²¹ The significant family history in one of our patients and recent literature suggest that HIV infection is an increasingly significant etiologic factor that should be considered in African children with DCM.³⁰,³¹ DCM is also known to have familial or genetic forms, while recent reports suggest that selenium deficiency is another possible etiologic factor.³²,³³ With our limited diagnostic facilities we were unable to establish any of these diagnoses. However, micronutrient deficiencies are common among children in sub-Saharan Africa.³⁴ Public health measures aimed at promoting good nutrition in the community in general and children in particular will therefore be helpful in preventing nutritionally derived DCM. The present study also showed that DCM was more common in younger children, while RHD occurred more frequently in school-aged children and adolescents, a trend that is similar to reports from other parts of Nigeria and Africa.¹⁰,¹¹,¹² These however were the ages at the time of ECHO diagnosis and not at the onset of illness.

Cor pulmonale was present in 15 (8.7%) of our patients with AHD. Much fewer cases have been reported from other parts of Nigeria.⁸,¹¹ This calls for further investigation, as it is possible that with the high altitude in our locality, the effects of relative hypoxia such as vasoconstriction may lead to an increase in pressure in the pulmonary vascular bed.³⁵ The fact that we found no apparent cause in many of our subjects may support this theory. The few etiologic or associated factors found were poorly treated bacterial pneumonia, tuberculosis, and sickle cell anemia, all of which are recognized causes of cor pulmonale.⁶,³⁷ Human immunodeficiency virus (HIV) infection is increasingly recognized as an important cause of cor pulmonale in children, either in isolation or in combination with tuberculosis.³⁰ Two of our patients diagnosed with cor pulmonale secondary to pulmonary tuberculosis also had HIV infection. However, during the period under review, we did not routinely screen all our echocardiography or cardiac patients for HIV and so may have missed some other cases.

We saw relatively fewer children with pericardial disease (15 cases or 8.6% of all children with AHD) compared with Lagos where it was the most frequent AHD and Ibadan where it was the second most frequent.⁸,¹¹,¹³ On the other hand, Asani et al.¹⁰ in Kano reported no cases of pericardial disease at all. These apparent differences may be random or related to geographical variations within the country. Tuberculosis was a predisposing illness in two of our children with pericardial disease. Two of the other cases had associated fever and echocardiographic features of an organizing effusion which would suggest a purulent bacterial aetiology. Although there were no bacterial growths on culture, antibiotic treatment resulted in remarkable improvement, supporting a bacterial infective process and in keeping with the other Nigerian literature.⁸,¹¹ HIV infection, which we did not routinely screen for, is also an increasingly recognized cause of pericardial disease in African children.³⁰,³¹

We found only one child with EMF, as did Okoromah et al.,¹¹ while Asani et al.¹⁰ reported no such case. By contrast, Antia²⁵ had in 1972 reported 15 cases from Ibadan, while Jaiyesimi⁴⁰ in 1982 reported 52 cases from the same institution. The rarity of EMF in this and the other recent reports, may lend credence to the observations of other recent researchers that EMF may be a disappearing disease in Nigeria.³⁹ Reasons adduced for this apparent change include general improvements in nutrition and socioeconomic conditions. Deficiencies of the amino acids leucine, valine, and tryptophan had earlier been implicated in the aetiology of EMF in adults in East Africa.⁴⁰ Also, basic sanitation has improved, while better education including health education may have effected some behavioral changes that might have led to a reduction in the incidence of parasitic infections suggested in the past to be associated with EMF.⁴¹

The relative rarity of endocarditis in our study is not surprising as very few cases have been reported from other parts of the country.¹³,⁴²,⁴³ This may be the result of a low index of suspicion coupled with the fact that vegetations are usually seen on echocardiography only in advanced cases. The overall incidence in Nigeria and other developing countries is not known. As with our study the reports from Ibadan and Zaria documented endocarditis mainly in children with underlying heart disease.⁴¹,⁴²

The other AHD we encountered, namely aortic aneurysm, ARVD and NDCM are all said to be rare in children.²¹,⁴⁴ Apart from reports of left ventricular aneurysms from Ibadan,²¹,⁴⁵ reports from other parts of Nigeria have not included any of these heart lesions, a reflection perhaps of their rarity.¹⁰,¹¹

**CONCLUSION**

RHD is still the most common AHD among children in our center, particularly among older children, while DCM is most common among younger children. There is urgent need for an improvement in living conditions and also a well-planned primary prevention program aimed at prompt and appropriate treatment of streptococcal sore throat alongside secondary prevention of RHD.²⁵,⁴⁶ These will reduce the incidence of rheumatic fever and its cardiac sequel RHD, as well as arrest disease progression, thus reducing the number of children and young adults requiring expensive heart valve surgeries (which are virtually unavailable in the country) or dying prematurely from chronic heart failure.⁵,⁴⁷ There is also a need for more
vigorously implemented the National Tuberculosis Control Programme as this will help to reduce the incidence of its cardiovascular complications such as cor pulmonale and tuberculous pericarditis. Vigorous pursuit of the millennium development goals and a general improvement of child health in the developing world will also have an impact on the incidence of acquired heart diseases in children as the majority of these are of infectious aetiology. We also recommend multicenter collaborative studies and community surveys of heart disease prevalence with a view to ascertaining the true burden, spectrum and epidemiology of heart diseases in Nigerian children and to more accurately assess regional differences and changes in trend.

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All three authors have fulfilled the criteria for authorship, have read and approved the final manuscript and believe that all the data presented therein are true and represent their honest work.

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