ORIGINAL ARTICLE

CHILDHOOD DILATED CARDIOMYOPATHY IN JOS, NIGERIA

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SUMMARY

Objective: ***to study the pattern of childhood dilated cardiomyopathy in Jos University Teaching Hospital, Nigeria.***

Methodology: ***Paediatric echocardiography records (January 2000 to June 2002 – 2 and ½ years) were reviewed and cases of dilated cardiomyopathy (DCM) identified. Clinical, radiologic, electro- and echocardiographic data of identified cases were reviewed.***

Results: ***childhood DCM (6 cases) was the commonest acquired heart condition diagnosed in children under the age of 5 years. Four patients presented with recurrent episodes of congestive heart failure (CHF) while 2 presented during their first episode. DCM was initially considered in only 1 child. Another child suffered a cerebro-vascular accident (CVA) before the diagnosis was made.***

Conclusions: ***DCM should be considered early in young children with long-standing or recurrent CHF. A high index of suspicion, early diagnosis and appropriate management should reduce morbidity, prevent complications and prolong survival.***

INTRODUCTION

Childhood DCM is the main indication for paediatric heart transplantation in Europe and North America.[1] Its incidence there is said to vary from 2-8/100,000 while its prevalence is approximately 36/100,000.[2,3] The disease is characterized by dilatation of the cardiac chambers particularly the left sided ones. Indices of poor left ventricular contractility are usually demonstrable at echocardiography.[4] Seventy-five percent of cases of DCM in developed countries are diagnosed within the first 2 years of life.[1] The aetiology in a significant proportion of cases is undetermined. However, 40-45% of cases have been attributed to myocarditis (biopsy proven) and up to 30% to genetic factors[1-3]. The latter could be in the form of specific cardiomyopathies as seen in association with inborn errors of metabolism or neuromuscular diseases, or be directly attributable to autosomal (dominant or recessive), X-linked or mitochondrial modes of inheritance. Also, the HLA loci DR4 and DQB1 located on chromosome 6 are thought to be possible genetic markers for susceptibility to DCM.[2]

By contrast, while DCM is also a common cause of congestive heart failure (CHF) in adults in Nigeria,[5-7] reports of childhood DCM in this country have been rare. Bronchopneumonia is recognized as the foremost cause of CHF in Nigerian children,[8] while congenital heart disease (CHD) and rheumatic heart disease (RHD) are the leading intrinsic heart diseases that are usually responsible for childhood CHF. Therefore, the incidence, prevalence and aetiology of childhood DCM in our environment remain largely undefined.

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In 1969, Antia and his co-workers[9] at Ibadan, reported their observations on 13 children with 'idiopathic cardiomegaly'. The clinical features were largely consistent with what is today called DCM. The authors leaned in favour of an infectious aetiology because 12 of the children had had fever at the onset of their illnesses, while 3 had leukocytosis and 1, leucopenia. Infectious diseases remain highly prevalent in our environment and are therefore likely to have continued to play a major aetiologic role. The contribution of genetic disorders remains difficult to evaluate because of paucity of the appropriate diagnostic modalities. Antia[10] had in an earlier paper in 1968, highlighted the problems of the clinical diagnosis of the condition. There has been no significant improvement in the diagnosis of this condition in the country since then, since echocardiography which has become the standard diagnostic mode in other countries remains outside the reach of the majority of our population. This may account for the paucity of reports since the work of Antia et al, leading in turn to a low index of suspicion.

Since late 1999 it has become possible for more children to undergo paediatric echocardiographic examinations in our hospital. We then began to encounter cases of DCM, up to a total of 6, in 2 and ½ years. We here report their clinical, radiologic, electro- and echocardiographic findings, as well some of our observations concerning their outcome and follow-up.

MATERIALS AND METHOD

We reviewed our paediatric echocardiographic (ECHO) records spanning a period of 2-and ½ years (January 2000 to June 2002) and identified all patients with a diagnosis of dilated cardiomyopathy (DCM). The diagnostic criteria

were a left ventricular internal dimension in diastole [LVIDd] exceeding the 95th percentile for age, associated with poor LV contractility as evidenced by fractional shortening [FS] less than 25% and ejection fraction [EF] below 50%. The ECHO studies were performed using Hewlett Packard ultrasound imaging systems (model 77020A from January to July 2000 and model 77035A or *SONOS* 1500 from November 2000 to June 2002. Both were usually fitted with 3.5 MHz transducers when used on our children. Intracardiac dimensions were measured as recommended by Sahn *et al.*[11] Neither of these echo machines was, however, fitted with a printer. It was consequently not feasible to record any still images.

We retrieved the hospital records of the identified cases of DCM, for a detailed review of their clinical, radiologic, electro- and echocardiographic data. Information obtained with respect to each patient included age, gender, duration of illness before presentation, symptoms, previous treatment if any, findings on physical examination, diagnosis at presentation, treatment received, clinical course and the current status at follow-up. Others were the cardiac size (from the chest radiograph), electrocardiographic [ECG] abnormalities, and ECHO data which included the intracardiac dimensions plus any other abnormalities found on 2-D and Doppler echocardiography. All the results were tabulated and any visible trends noted. Where appropriate, the mean and/or median values of continuous variables or the frequencies of categorical variables were also determined and recorded.

RESULTS

A total of 93 paediatric echocardiograms (excluding repeats) were performed during the period under review. Six (6.5%) of these satisfied the stated criteria for the diagnosis of DCM. They accounted for 12.3% of the 49 children diagnosed with acquired heart diseases (AHD) during the period, and for 5 (45.5%) of the 11 cases of AHD who were aged less than 5 years. The other AHD diagnosed in children less than 5 years of age during this period were: cor pulmonale (3 cases), tuberculous pericarditis with effusion (1), septal hypertrophy – possible hypertrophic cardiomyopathy (1), and a severe mitral regurgitation whose cause was not immediately apparent (1).

Clinical characteristics: The 6 DCM patients ranged in age from 32 to 72 months (mean 43±15.3, median 36 months), and comprised 1 male and 5 females. Their median duration of illness before presentation was 1.5 months (range 2 weeks to 18 months, mean 5.5 ± 7.45 months). Some of their clinical data are summarized in Table I. Cough, dyspnoea, facial and leg swellings were present in all the 5 patients whose case notes were available for review, while 4 (80%) had a history of fever at the onset of the illness, and presented with anorexia, vomiting and abdominal pain. Other symptoms noted were: orthopnoea (3 patients), paroxysmal nocturnal dyspnoea, chest pain, excessive sweating and lethargy or reduced physical activity (2 patients each). The most frequently observed physical signs were: a bulging praecordium, displaced apex and hepatomegaly in all 5 patients, a third heart sound in 4 (80%) and a mitral incompetence murmur in 3 (60%). Praecordial hyperactivity was observed in 2 patients (40%) and distended neck veins and ascites in only one (20%) each.

Laboratory data: Review of the chest radiographs revealed that all 6 patients had globular cardiomegaly (figure 1). The ECG abnormalities found in the children together with some of their echocardiographic parameters at the time of diagnosis are shown in Table II. Three (50%) of the 6 children exhibited T-wave abnormalities in lead V_6. Figure 2 shows one such ECG tracing. All 6 children had dilated and poorly contractile left ventricles, and dilated left atria – in keeping with the diagnosis of DCM.

Follow-up: All our patients were initially stabilized on standard anti-CHF drugs. The parents of patient number 4 elected to continue follow-up at the referring mission hospital for convenience. Patients 1 and 3 defaulted early and we recently received a report that patient 3 had died at home. Patient 2 improved enough to be taken off medication and then defaulted. However, patient 5 despite excellent compliance continued to deteriorate, required increasing dosages of medication, and eventually died. This patient had had a younger sibling with history of a similar illness, who died at the age of 3 months. No genetic studies could be carried out. Currently, only patient 6, who had earlier suffered a CVA is still being followed up. She is stable on medication but relapses if for any reason 1 or 2 days of treatment are missed.

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TABLE I: Clinical data in 6 children with dilated cardiomyopathy

S/N	ID	Age (Months)	Gender	Illness duration	Aetiologic clue	Weight (kg)/ % EWA	Prevention diagnosis/treatment	Clinical diagnosis at presentation	Interval before echo	Treatment received in JUTH	Clinical Course	Present status
1.	MM	48	M	6 weeks	Negative	14 / 87.5	SCD / 2 blood transfusions	CCF / RHD	8 day	Diuretics, digoxin	Poor compliance; relapses.	Defaulted
2.	MA	36	F	2 weeks	Preceding viral URI	10 / 114.5	Bronchopneumonia	CCF / AGN	2 weeks	Diuretics, digoxin	Improved; medication discontinued after 1 year.	Defaulted
3.	CM	36	F	NA	NA	NA	NA	CCF / DCM	<1 week	Diuretics, digoxin, captopril	Early default	Died
4.	HAB	72	F	18 months	Negative	11 / 61.1	Recurrent CCF; 12 admissions, 2 blood transfusions, anti-tuberculous chemotherapy (6/12).	RHD / EMF	2 weeks	Diuretics, digoxin, captopril, ASA	Follow-up in peripheral hospital.	NA
5.	NP	32	F	7 months	3 month old sib died of similar illness	10.7 / 82.3	Recurrent bronchopneumonia, pulmonary tuberculosis.	CCF / VSD, BPn	9 days	Diuretics, digoxin, captopril, ASA	Relapses; increasing doses of medication.	Died
6.	PC	34	F	3 weeks	Negative	10.6 / 77.4	Recurrent bronchopneumonia	Recurrent BPn / CCF	3 months	Diuretics, captopril, ASA	CVA	Stable

Key: *CCF = congestive cardiac failure*
SCD = sickle cell disease
RHD = rheumatic heart disease
EMF = endomyocardial fibrosis
AGN = acute glomerulonephritis
SD = ventricular septal defect
DCM = dilated cardiomyopathy
BPn = bronchopneumonia
ASA = acetyl salicylic acid
CVA = cerebrovascular accident
JUTH = Jos University Teaching Hospital
URI = upper respiratory infection
NA = data not available
EWA = expected weight for age.

TABLE II: ECG abnormalities and some echocardiographic parameters in 6 children with dilated cardiomyopathy

S/No	ID	Age (Months)	Gender	ECG abnormalities	RVIDd (cm)	LVIDd (cm)	LVIDs (cm)	LVPWd (cm)	IVSd (cm)	LA (cm)	Ao (%)	FS (%)	EF (%)	Other ECHO findings
1.	MM	48	M	LAE	-	5.10	4.30	0.90	0.90	0.90	1.70	15.7	40.1	IAS bulging into RA
2.	MA	36	F	Inverted T in V6	1.1	4.58	3.73	0.52	0.58	2.65	1.37	18.6	46	-
3.	CM	36	F	NA	-	506	4.29	0.65	0.77	4.07	1.43	15.2	39.1	Small ASD II, paradoxical interventricular septal motion
4.	HAB	72	F	RAE, RVH, LVH	1.02	5.68	4.75	0.72	0.72	4.83	2.20	16.0	34.0	-
5.	NP	32	F	Niphasic T Vt, inverted T in III	-	6.51	5.76	0.75	-	3.73	1.83	12.6	26.7	-
6.	PC	34	F	Inverted T in V6	0.95	4.88	4.68	0.95	0.48	-	-	4.1	9.8	Apical LV Thrombus

Key:
RVIDd = right ventricular internal dimension in diastole
IVSd = interventricular septum dimension in diastole
LVIDd = left ventricular internal dimension in diastole
LVIDs = left ventricular internal dimension in systole
LVPWd = left ventricular posterior wall dimension in diastole
LA = left atrial dimension
Ao = aortic root dimension
NA = data not available
ASD II = ostium secundum atrial septal defect
IAS = interatrial septum
RA = right atrium
LV = left ventricle

DISCUSSION

Our finding of 6 cases of childhood DCM within a 2 and ½ year period suggests that this condition may not be as rare in Nigerian children (at least in our part of the country) as might have been presumed. The paucity of further reports since the total of 17 cases reported by Antia et al[9 12] may not be a reflection of the true situation, but of the lack of appropriate and specific diagnostic modalities - in this case, echocardiography. This may have led to a relatively low index of suspicion for childhood DCM: it is very likely that as a result, most of the cases of longstanding or recurrent heart failure in young children have been presumed to be secondary to acyanotic congenital heart disease.

This report has however revealed DCM as the commonest acquired heart condition among under-5 children in our centre, accounting for 45.5% of the cases of AHD diagnosed in this age group. Our oldest patient was aged 6 years at presentation. This is similar to the findings of Antia et al, whose patients ranged from 11 months to 6 years of age,[9] and in consonance with reports from other countries to the effect that childhood DCM is predominantly a disease of younger children.[1]

As expected, our patients generally presented with symptoms of CHF,[1-3, 9] that had largely been recurrent or longstanding, and had frequently been mis-diagnosed as acyanotic CHD with or without pneumonia. Although CHDs remain the commonest and most important causes of heart disease in young children, our findings show that DCM should be more readily considered as a differential diagnosis of recurrent and longstanding heart failure in this age group. This is even more important when the physical findings are not fully consistent with any of the common (acyanotic) CHD that manifest in early childhood. Such conditions as ventricular septal defects (VSD) or patent ductus arteriosus (PDA) for example, are each usually associated with their typical murmurs.

Three of our patients had murmurs suggestive of mitral incompetence (MI). This is a frequent observation in patients with DCM, and should be differentiated from organic MI, which in our environment, is usually secondary to RHD but could occasionally be attributable to a congenital cleft in the anterior mitral leaflet (AML), or to mitral valve prolapse. No organic mitral valve disease was found at ECHO in any of our DCM cases. Dilatation of the mitral valve ring secondary to severe LV dilatation is the usual explanation. The presence of a murmur of MI in young children with CHF could therefore be a possible pointer to DCM. Talierco and co-workers[4] have however observed that severe MI was present only in those of their patients who ultimately died.

The presence of globular cardiomegaly in all 6 of our patients is consistent with the findings of Antia et al[9], and in keeping with standard teaching.[2,3] The main differential diagnosis is that of a massive pericardial effusion (PE). This we were able to exclude relatively easily, by means of 2-dimensional echocardiography. We, however, accede to the strong scientific value of clinical ECHO photographs in delineating our current series had this been feasible. ECHOs can also usually be done using any simple ultrasound machine - even when the full ECHO study that includes Doppler and M-mode echocardiography cannot be done. Apart from excluding PE, simple ultrasound while not providing the quantitative data on which the definitive diagnosis of DCM is usually based, does provide very useful qualitative information – that of a dilated, poorly contractile heart. With the increasingly widespread availability and use of upper abdominal ultrasound, manpower for this simplified form of cardiac ultrasound is (with a little additional training) currently available in most hospitals where ultrasound can be done. The definitive diagnosis of DCM is however made by means of accurate measurements of the intracardiac dimensions during M-mode echocardiography – this is best carried out by a cardiologist or trained echocardiographer. Therefore suspected cases should as much as possible be referred for the appropriate definitive diagnosis.

The ECG findings in DCM are usually non-specific and include sinus tachycardia (which was present in all our patients), hypertrophy signs, arrhythmia, low QRS voltages and ST segment and T-wave abnormalities[2,3]. Fifty percent of our subjects had biphasic or inverted T-waves on their surface left chest leads. This is comparable with the observation of Antia et al[9], who reported this finding in 8 (61.5%) of their original 13 patients. We suggest that this finding, when combined with careful clinical observations, could be another useful clue that to help differentiate DCM from left-to-right shunt CHD (which are usually associated with tall T-waves), especially when cardiac ultrasound is not readily available.

The prognosis of childhood DCM is at best, guarded. Except in rare instances such as systemic carnitine deficiency, there is currently no available cure.[2] In advanced countries, the mean survival rates are approximately 70% at 1 year, 60% at 5 years 50% at 10-11 years after diagnosis.[2] One third of affected children are expected to die within

the first 2 months of diagnosis[2]. Another 1/3 are expected to remain the same while the remaining third improve or recover completely[2,3]. Improvement would usually be evident within the 1st 6 months, which was the case with our patient number 2. A repeat ECHO study to fully substantiate this was however not possible. History of a recent viral illness (less than 3 months prior to DCM symptoms) is a good prognostic factor[2,4] and was present in this patient. Familial DCM tends to carry a poor prognosis. Patient 5 seems to fit into this category, even though unconfirmed.

The mainstay of management is to control heart failure and any significant arrhythmias that may arise, and to reduce the risk of thrombo-embolic complications.[2] The high default rate in our series is in sharp contrast to the experience of Antia et al,[9] who were able to follow their patients for 2-7 years, and may be a reflection of the current socio-economic conditions in the country.

Finally, this report has shown DCM to be the commonest acquired heart disease (AHD) among children under the age of 5 years in our centre. Careful clinical, radiologic and ECG observations provide helpful clues in trying to clinically differentiate childhood DCM from the acyanotic CHD that may present in a similar manner and in the same age group. We advocate a higher index of suspicion so that all suspected cases could be subjected to simple cardiac ultrasound or referred for echocardiography to establish the diagnosis. This point is underscored by our patient number 6, who suffered a CVA as a result of delayed diagnosis. Once the diagnosis is confirmed, appropriate treatment to control heart failure and low dose aspirin to prevent thrombo-embolic complications should be instituted.[2]

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