Endomyocardial fibrosis in Sub Saharan Africa: The geographical origin, socioeconomic status, and dietary habits of cases reported in Yaounde, Cameroon

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ABSTRACT

Background : **Endomyocardial fibrosis (EMF) is a neglected heart condition of the inter-tropical regions. Numerous hypotheses suggest a relationship between its geographical distribution in the affected regions and other etio-pathogenic factors such as dietary habits, infectious causes, and geochemical causes. Knowledge of its epidemiology in Cameroon remains limited, which is why we decided to describe the profile of a paediatric series of EMF in Yaoundé.**

Patients and Methods : **A retrospective study was carried out on EMF in 54 patients diagnosed from 1 January 2006-31 December 2014 in a Paediatric Centre of Yaoundé. Diagnosis was mainly echocardiographic. We compiled data on the geographic origins of the patients, their dietary habits and the socioeconomic profile of their families.**

Results : **The patients' ages ranged from 2 to 17 years, most of whom (83.3%) were between 5 and 15 years. For geographical distribution, all came from three tropical forest zones where they have lived since their childhood. These were Center (32/54), South (12/54), and East (10/54). All families had a moderate income, consumed tubers at least twice a week especially cassava (43/54) and had low sources of proteins.**

Conclusion : **Apart from geographical similarities all patients of our series shared the same dietary habits. Our study was conducted in a hospital setting; therefore a screening of the disease in the whole national territory would enable a more reliable mapping.**

Keywords : Cameroon, dietary habits, endomyocardial fibrosis, geographical origin, socioeconomic status, Yaounde

INTRODUCTION

First described in the late 40s in Uganda, endomyocardial fibrosis (EMF) is a neglected disease and remains a mystery till date.[1,2]

It is a restrictive cardiomyopathy characterized by a fibrotic thickening of the endocardium and the myocardium. It affects the apices and the inflow tract of one or both ventricles.[2-4]

The etiology remains unknown, the pathophysiology not fully clear. The most widespread hypotheses in relation to the predisposing factors include nutritional deficiencies, frequent consumption of certain foods, especially cassava, parasitic infections, which lead to eosiniphilia as well as geochemical soil characteristics from which patients get their foods.[2,5]

Endomyocardial fibrosis is mainly found in regions of the tropical belt.[6] In Asia, it has mainly been described in China and India[7,8] In Latin America, most cases were mostly from Brazil[9] and Venezuela.[10] The disease is most common in Africa, south of the Sahara where it mostly affects children and young adults in the lower

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socioeconomic group.[2,6] Literature is full of series reports from Uganda and Mozambique.[11-13] Other countries are, however, mentioned (Cote d'Ivoire, Nigeria, Ghana).[2,9,14] In countries where the disease has been studied, the authors noticed a marked variation of prevalence from one geographic region to another.[2,7,15] Cameroon is not cited as one of the numerous African countries in which the disease exists due to lack of published data.[2,9] We collected data on EMF in a series of patients followed up in a popular pediatric hospital in Cameroon. The geographic origin of patients was determined to draw out the mapping of this disease in Cameroon. Information on the main foods normally consumed as well as their socioeconomic characteristics were gathered.

PATIENTS AND METHODS

Type of study

We conducted a retrospective study using echocardiography and hospitalization records, as well as files of patients with EMF seen between January 1, 2006 and December 31, 2014.

Site of the study

The study took place at the Mother and Child Centre of the Chantal Biya Foundation (MCC/CBF). The MCC/CBF is a Children's Hospital located in the heart of Yaoundé, the political capital of Cameroon, a city of nearly 2½ million inhabitants. It is a University Teaching Hospital with a 260-bed capacity, the busiest pediatric hospital structure in the country. About 30.000 children are consulted every year and about 9.000 are admitted. The center is highly publicized because of its affordability. It, therefore, receives children from all social strata coming from all parts of the country. In January 2006, a Pediatric Cardiology Unit was opened. The team was led by a trained pediatric cardiologist, assisted by a general practitioner and a pediatric resident. This unit, exclusively dedicated to children, is one of the two in the country, whose tasks include: Outpatient consultations, noninvasive cardiologic explorations and treatment of hospitalized patients.

Study population

In January 2006, a register for heart diseases was opened in the cardiology unit where all children with heart disease were routinely recorded. A special section in this register is reserved for EMF. Furthermore, an iconography is used in the archiving process, and recording of at least two telephone numbers of parents/relations of each patient is implemented. The patients are contacted when necessary, either to remind them of their appointments or to get additional information when needed.

A complete physical examination is performed, and pulse oximetry is carried out on all patients with heart disease at the outpatient consultations and in the wards. To patients in whom EMF is diagnosed during the course of an echocardiography, an appointment is given to come back and complete information necessary for the medical file. A food survey is done. For this, parents and/or children of more than 10 years of age are asked, to list the three main foods that make up their usual meals according to priority. They are also asked the different foods, how often they consume these foods weekly, and their weekly sources of proteins. We also gather information on their usual place of residence since birth, their ethnicity, and the occupation of the family head and sources of revenue for the family. A survey is done on their home environment, the equipment found there (to know to their standard of living), the usual mode of transport.

All data are recorded in medical versions in both soft and hard copies and archived.

To diagnose EMF we use a commercially available echocardiography equipment (ACCUSON Cypress Siemens) and a 3-7 Megahertz transducer in bi-dimensional, Doppler and M-modes. tests are performed according to the American Heart Association.[16] Echocardiography examination was performed in the subcostal view, parasternal long axis, short axis, apical four and five chambers, and suprasternal views. The diagnosis of EMF was posed in patients if we have a combination of a more or less extensive retraction of at least one ventricle, apical obliteration of at least a ventricle by a hyper echoic mass, regurgitation in the atrio-ventricular valve and a corresponding atrial enlargement.

Other tests performed include a plain chest X-ray, an electrocardiogram and a full blood Hyper-eosinophilia is inferred if eosinophil values are above 500/μl.

Ethical considerations

The ethical committee of the Faculty of Medicine and Biomedical Sciences of the University of Yaoundé approved this study.

Statistical analyses

Data were analyzed using Epi Info version 3.5.1 (CDC, Atlanta) software. Heart lesions and hyper eosinophilia were described according to the sociodemographic characteristics of the children and the main type staple in the families. Continuous variables were expressed as medians and intervals ranges while categorical variables were expressed as percentages. The level of significance was set at $P < 0.05$.

RESULTS

Between January 1, 2006 and December 31, 2014, 54 patients with the diagnosis of EMF were recruited

in our hospital in the echocardiography base. During this period, our site recorded 273.117 consultations of patients <16 years of age. Among them, 1.666 were diagnosed of heart disease. EMF, therefore, accounted for 0.019% of consultations and 3.241% of all heart diseases.

Residence

All the patients of EMF in our series lived permanently in only 3 of the 10 regions in Cameroon and were from the respective tribes found in those regions [Figure 1]. The regions included: Center region (32/54), South region (12/54), and East region (10/54). Forty-one (76%) patients lived in rural areas. This distribution of cases of EMF limited to 3 out of 10 regions of the country marked contrast to the geographical origin of patients with other heart diseases who came from all parts of the country [Figure 1 and 2]. Of the 1666 patients of heart disease in fact, 1612 had other heart diseases than EMF. Fifteen of them were from the neighboring countries of Cameroon. The national cases (1597) came from all parts of the country including those where no case of EMF had been reported. Although the central region remained clearly predominant (59.3%), all other regions were involved. Thus, noted in order of decreasing frequency the western regions (11.2%), coastal (9.9%), South (5.3%), the Far North (3%), North (2.9%), East (2.6%), Adamawa (2.4%), the Southwest (2%) and Northwest (1.4%).

Sex

Our study sample was made up of 27 girls and 27 boys, a sex ratio of 1:1.

Age at diagnosis

The youngest patient was 2-year-old and the oldest was 17-year-old at the time of diagnosis. The majority of patients were between 5 and 15 years (83.3%). The mean age was 10 (interquartile range of 7-13 years).

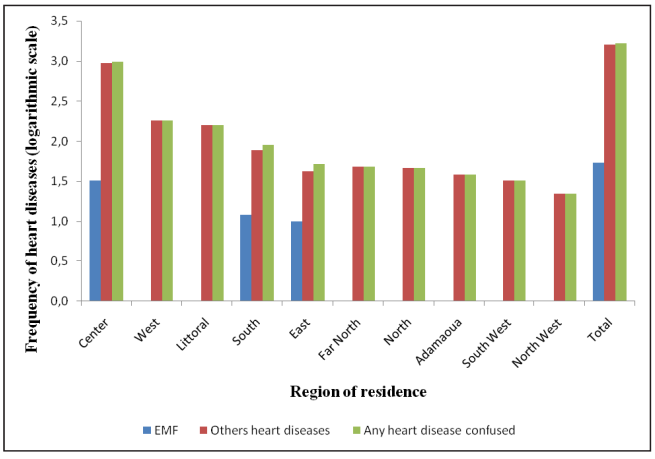

Figure 1: Distribution of the patients with endomyocadial fibrosis and others cardiac diseases with regard of the region of residence

Family origin of patients

Of the patients registered, The 54 came from 48 different families. Three belonged to the same family while eight were from four different families (two per family). Patients of the same family had always lived together in the same environment from birth.

Socioeconomic status of families

Taking into consideration those in charge of patients, we realized that the patients were all from economically deprived social strata. No family head owned a car. Only four families (8.3%) lived in a house with running water, four families (8.3%) had fridges and 32 families (66.6%) had no electricity in their homes.

Nine patients (16.6%) were orphans; 6 with no father and 3 with no mother. The head of the family was a subsistence farmer in 64.5% of cases. Other parents did small jobs like petty trading (10.4%), bricklaying (8.3%); night-watch (6.3%); low status civil servants on retirement (6.3%); household cleaners (4.2%).

Family meals

The main staple foods were limited to three tubers [Figure 3]: Cassava (*Manihot esculenta*); plantain (*Musa paradisiaca*); cocoyams (*Xanthosoma sagitifomium*). One of any of these tubers was consumed in a meal at least 6 times a week. The first staple food was cassava. It was the main menu for 40 patients where it was eaten at least 4 times/week. Second place was plantain, and the third place was cocoyams. The other foods (rice, sweet potatoes, yams, maize) were consumed occasionally. Tubers were eaten alone or with sauce. The main sauces were cassava leaves (64.8%), groundnut paste (22.2%), melon seeds (7.4%) or rarely other varieties of vegetables (5.6%). Only 10 patients (18.5%) could have at least one meal containing animal sources of proteins per week. Generally, sources of proteins were essentially made up of beans, occasionally dry and fresh fish, game.

Clinical features

Failure to thrive was a constant finding in our series. Other signs were: Hyperpigmentation of the lips in all the patients despite a normal oxygen saturation level; facial swelling; absence of lower limb oedema even with voluminous ascites [Figure 4]; signs of heart failure; arrhythmia. We noted a muffling of heart sounds in 24 patients. Heart murmurs were noted in 22 patients. These were most often tricuspid incompetence (45.5%) followed by mitral insufficiency (18.2%), pulmonary insufficiency (13.6%) and a combination of tricuspid and mitral incompetence (22.7%). The splitting of the second heart sound was noted in 16 children. Since 2006, 12 patients were lost to follow-up, 32 died and 10 are still being followed

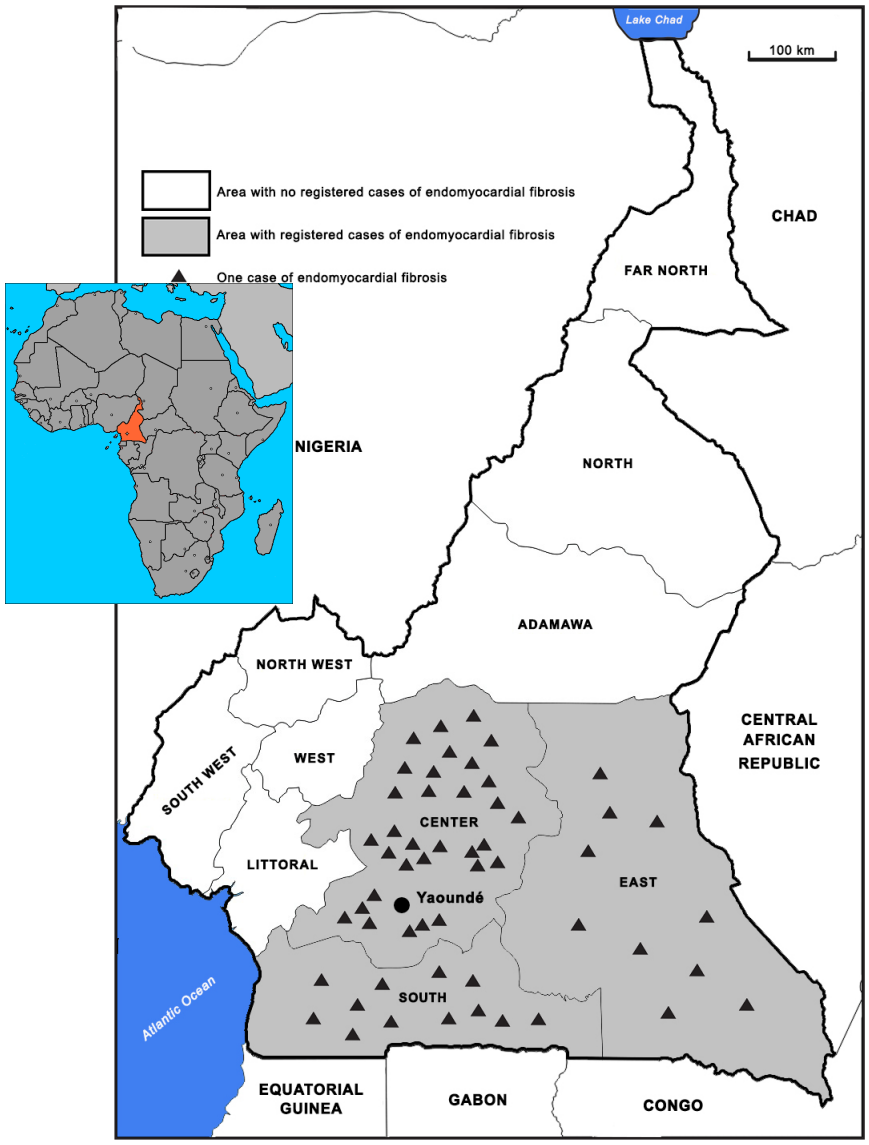

Figure 2: geographic origin of endomyocardial fibrosis patients

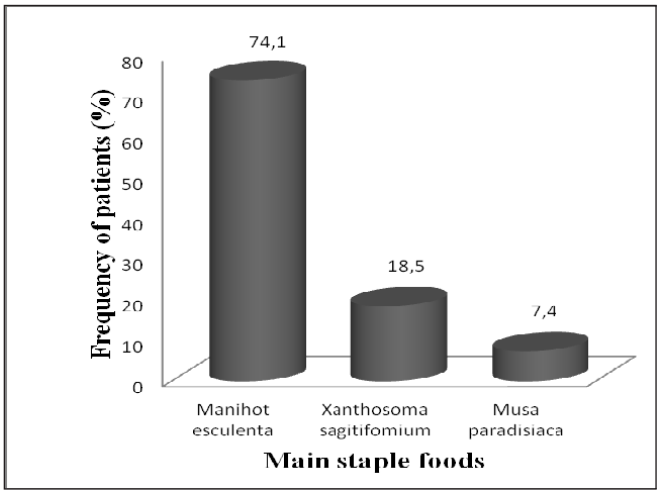

Figure 3: Main staple foods consumed by endomyocardial fibrosis patients

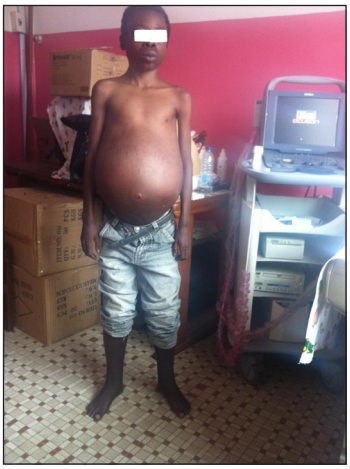

Figure 4: A 14-year-old boy with right ventricular endomyocardial fibrosis. The picture shows a stunted, dark lips, voluminous ascites contrasting with the absence of pedal edema

Echocardiographic profile

Exclusive right ventricular fibrosis [Figure 5] was seen in 43 patients (79.6%). The fibrosis involved the two ventricles in nine patients (16.7%). Only 2 patients (2.7%) had fibrosis solely located in the left ventricle. Thirty-one patients (57.4%) had pericardial effusion. Apart from the two patients who had left ventricular exclusive lesion, all patients had tricuspid regurgitation which varied from moderate to severe. In right side lesions, right atria were increased volume, sometimes severe. In biventricular lesions, the two atria were enlarged, while only the left atrium was in two patients with exclusive involvement of the left ventricle.

Other investigations

Cardiomegaly on plain chest X-rays were seen in all patients. The cardiothoracic index varied between 0.58 and 0.96 [Figure 6]. We found cardiac calcifications in three patients.

We equally found a low voltage QRS in 10 patients (58.8%) and a slow ventricular rhythm in 7 patients (41.2%) who did an electrocardiogram. Seven (41.2%) had atrial fibrillation, and two had atrial flutters. Auricular hypertrophy was frequent [Figure 7].

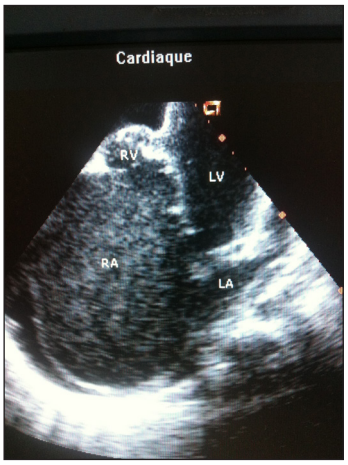

Figure 5: Echocardiograph picture of a patient with right ventricular (RV) endomyocardial fibrosis. Note RV tiny, almost filled by fibrosis. The right atrium (RA) is ectatic

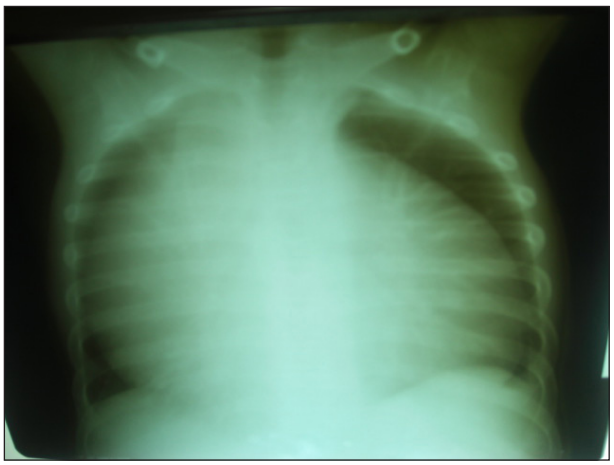

Figure 6: Chest X-ray of 13-year-old boy with right ventricular endomyocardial fibrosis. Note huge cardiomegaly

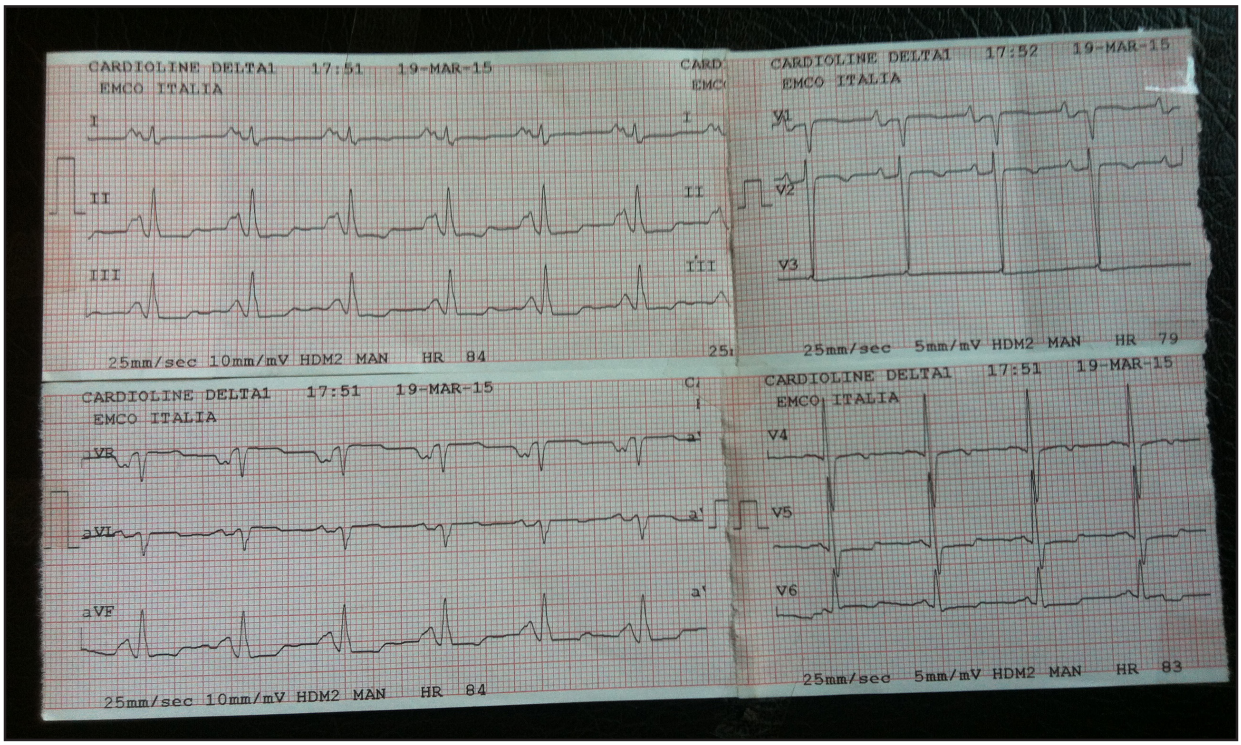

Figure 7: Electrocardiography of a 10 yrs old boy with endomyocardial fibrosis: bi-auricular hypertrophy

A full blood count was done in 44 patients. Hypereosinophilia was found in 27 subjects (61.4%), with values varying between 546-3780/μl. Hypereosinophilia were more frequent in subjects below 10 years of age ($P = 0.0000$). We did not note a meaningful link between hypereosinophilia and the main meal.

DISCUSSION

This study was intended to provide information about the geographical distribution of EMF in Cameroon and the sociocultural and economic characteristics of patients. The aim was not to review the whole subject of EMF. Our results show that the pathology exists in the pediatric setting of our country, which the patients originated from certain particular areas and that familial, cultural and nutritional links probably exist in patients.

In Cameroon like in many other countries, the prevalence of EMF is not well known. Mocumbi *et al.*[12] in a prospective study using echocardiography for active screening found 211 patients of EMF out of 1063 examined (19.8%). The prevalence of EMF obtained in the same population a couple of years prior to the study, using presumptuous clinical signs, was 8.9% before confirmation of diagnosis.[12,13] Only 22.5% of cases diagnosed in this population were symptomatic. Diagnosis is generally posed in the late stages of the disease. Studies based on data collected in hospital settings like ours, therefore, does not reflect the real prevalence of this pathology. No data on the pathology was found in the literature in our country. The data we showed do not arise from a rigorous health surveillance system. We believe that the number of patients in our hospital discovered in a short time while the disease was not described in the past was due to diagnostic facilities we currently have. Moreover, our diagnostic criteria were too specific and less sensitive. These probably made us lose some patients. Had it been we followed diagnostic criteria as established by Mocumbi *et al.*[12] which are more sensitive, we might have retained more patients.

The disease is known to be predominantly in hot and humid regions like in the equatorial zones of Asia and Africa.[7,12,14] The regions of origin of our patients are forest regions with hot and humid tropical climates. Even though, Yaounde is a cosmopolitan town with its inhabitants from different regions of the country; we discover that our patients all owe their origin to three regions. This is probably not just sheer coincidence or due solely to their geographic proximity to the Center region. Indeed, as shown in Figure 1. The patients suffering from other heart diseases come from all over the country, even the most remote areas. It is understandable that the geographical distance and financial difficulties can be a barrier to access to our hospital. But for other heart disease, all 10 regions of the country are represented. If there are patients of EMF in seven other regions, it would be difficult to explain that none of them could get to our level then we have received other heart disease patients from these areas. Bertrand *et al.* found in a series of 14 patients in Abidjan a significant predominance of certain specific cultural groups.[17] Other authors have had similar findings.[6,18,19] It is possible that there is a genetic predisposition component in the genesis of the disease, as is the case in acute rheumatic fever. In the three areas where the disease was discovered lived the same ethnic group or at least ethnic groups with family relationships. These are essentially the "fangs-betis" and "bassa". The first would be the ancestors of the latter. The main ethnic groups of the three regions cited mainly consume the tubers with as main cassava, which has a cultural aspect. However, we have no objective evidence to say that one or the other factor cited above is responsible or is more important than the other in the genesis of the disease. We believe that the occurrence of the disease is probably multifactorial, involving at least dietary factors (such as excessive consumption of cassava), possibly genetic considerations and infectious factors.

We did not find any difference in gender. This has been noticed by most authors in the pediatric population.[14,18,20]

Studies centered on children are rare although we are dealing with a disease of children and young adults.[21] Our work was done exclusively in a pediatric setting and usually, we receive children <16 years old. The 17-year-old patient in our study was followed in our services because of social reasons.

Endomyocardial fibrosis is known as a disease of poverty.[4,14,20] In our series, we note that 76% of patients live in rural areas while only 46% of the general population of Cameroon is rural.[22] In Cameroon, rural populations are known to be poorer than people who live in urban areas.[23]

Poverty, though, is not the only explanation. Cases of EMF have been described in well-nourished Caucasians living in tropical regions.[10,19] In Cameroon, poverty is not limited to the 3 incriminated regions in our series. On the contrary, the lowest poverty index in the country is found in the south.[23] The fact that the south is one of the most affected regions by EMF is proof that poverty alone does not suffice to justify the disease. No case of EMF was found in the North West region,[24] which has the highest index of poverty.[23] This region has the only cardiologic surgical reference center in the whole of Central Africa with health staff with skills to detect this kind of disease. In the West region, which is the most populated region of the country, out of the 1252 patients suffering from heart diseases over a period of 14 years, the cardiology services did not record any case of EMF.[25]

All the patients we registered in our series had very dark-bluish lips [Figure 4], but the pulse oximetry remained normal. This sign is rarely described in the literature. It is possible that in previous series this sign was mistaken for skin color. Besides the color of lips, cardiac insufficiency is equally a major clinical manifestation in our series.

Stunted growth along with malnutrition was a constant finding in our series. Patients had a rich carbohydrate and poor protein diet. The nutritional deficiency hypothesis as etiology of EMF is widespread. The excessive consumption of cassava is usually incriminated.[9,26] In our series, cassava was consumed by all the patients and was the basic diet of 74% of patients. Where it was not the first most consumed meal, it came at least second or third place. Meanwhile plantains which were initially incriminated in the mid-sixties, but not completely proven, constituted the second food source consumed by our patients. The consumption of cocoyam has rarely been evoked by authors. In our series, this was the third source of feeding. Cassava, cocoyams, and plantains are food sources widely consumed by the ethnic groups in the seven austral regions of Cameroon. People from the Centre, South and East regions consume these foods more due to cultural reasons than other people.[27] This contrasts with the populations of other areas of the country. In the northern region, in fact, the meals are made mainly of cereals. In western and coastal regions, there is a wide variety of menus including cereals, legumes, tubers mainly consist of potatoes, sweet potatoes, yams, cocoyam. Cassava is in any case rarely consumed in these regions. The patients of our series all came from poor backgrounds. The consumption of meat or fish was seldom. A poor protein diet and a rich carbohydrate diet were proven in Uganda to be a contributing factor of EMF.[20]

In our series, we observed a clear predominance of right ventricular cases (43/54). Predominance of right ventricle involment has been proven in many other studies.[28]

Actually the hypereosinophilia was significantly found in young children. The authors believe that the hypereosinophilia is an early discovery in the disease, and the likelihood of finding her is inversely proportional to the duration of the disease.[6] Young patients would probably early in the disease while older are at a later stage.

LIMITATIONS

Our study is a retrospective case series study done in a hospital setting. Moreover, a single hospital was the place of study. Therefore, it will be an illusion in this context to draw a conclusion as to the prevalence of EMF. We are convinced that the frequency of this pathology is underestimated in Cameroon. One of the explanations could be that, due to imposing cultural beliefs, patients with such affections usually don't seek for modern medical care.[29] Due to a number of constraints, we were forced to limit the subject to some specific aspects as mentioned on the title. Clinical aspects have not been developed because of the scope of the topic we made early. Due to budget constraints, a survey to compare socioeconomic situation of populations in a standardized way has not been made possible. We stuck some sociocultural cues to determine the comfort level of families.

CONCLUSIONS

Contrary to certain data in literature, EMF is frequent in Cameroon, and followed a specific geographic distribution. Patients were from three regions, all situated in the equatorial forest zone. They all had similar sociocultural characteristics, especially in their feeding habits. The affected patients came from a poor background. Their nutrition was very unbalanced, characterized by a high consumption of cassava, followed by plantains and cocoyams, and very low consumption of animal proteins. The dark-bluish color of lips was a constant finding with very little mentioned about it in recent studies. The right ventricle was the most affected cavity. Although our site deals principally with most cardiac pathologies in Cameroon, we cannot establish the prevalence of this disease based on a single-center study. Active research of cases in all the regions will help to demarcate zones affected by EMF to those regions incriminated in this study.

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