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Original Article

Endomyocardial fibrosis in children

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Abstract We describe 10 children with endomyocardial fibrosis who underwent surgical treatment between 1978 and 1999. Seven were male and 3 female, with an age range from 4 to 15 years, having a mean age of 11 years. All were in the final stage of heart failure. Three had biventricular disease, 6 had involvement of the right ventricle alone, and one had endomyocardial fibrosis confined to the left ventricle. There were 3 deaths (30%) in the postoperative period due to low cardiac output. The 7 survivors were followed up for a period ranging from 12 to 168 months, with a mean of 72 months. Two late deaths have occurred resulting from heart failure and infectious endocarditis. Five (50%) children are still alive. Two required 3 reoperations for dysfunction of the inserted valvar prosthesis. One patient is in functional Class IV, and 4 are in Class II to III, despite intensive medical treatment. It is concluded that surgery for endomyocardial fibrosis is an essentially palliative procedure and, especially in children, the results of surgical treatment leave much to be desired.

Keywords: cardiomyopathy, heart failure, endocardial fibrosis

ENDOMYOCARDIAL FIBROSIS IS A RESTRICTIVE cardiomyopathy, described by Davies in 1948,[1] in which the essential feature is the formation of fibrous tissue on the endocardium of one or both ventricles. It leads to progressive obliteration of the ventricular cavity, with increased resistance to filling of the ventricles, and atrioventricular valvar insufficiency due to papillary muscle dysfunction. The clinical and morphologic characteristics of endomyocardial fibrosis are well known,[2,3] and its surgical treatment is an established procedure.[4–9] The disease is more frequently seen in tropical countries, where little attention has been given to its occurrence in children.[10–12] In our institution, of 91 patients with endomyocardial fibrosis submitted to surgical treatment, 10 were children. It is this group which constitutes the basis for this report.

Patients

From March 1978 to December 1997, 10 children with endomyocardial fibrosis underwent surgical treatment in our institution. Seven were male and 3 female, with ages ranging from 4 to 15 years, and a mean age of 11 years.

According to the New York Heart Association Functional Classification, all patients were in Class III or IV. Three had biventricular disease, 6 had involvement of the right ventricle alone, and 1 had endomyocardial fibrosis confined to the left ventricle.

All patients were in poor general condition, with a low nutritional state, and none presented eosinophilia at the time of admission.

Nine patients with either biventricular or right ventricular disease presented a very similar clinical picture, characterized mainly by raised venous pressure, massive hepatomegaly, and ascites. Auscultatory findings were unremarkable. In the one child with left ventricular disease alone, the clinical picture was characterized by dyspnea on minimal exertion, as well as signs of mitral insufficiency and pulmonary hypertension.

The chest X-ray films showed enlargement of the heart in all cases (Figure 1). Marked pulmonary congestion was observed only in the children with endomyocardial fibrosis of the left ventricle alone. Two presented pleural effusion.

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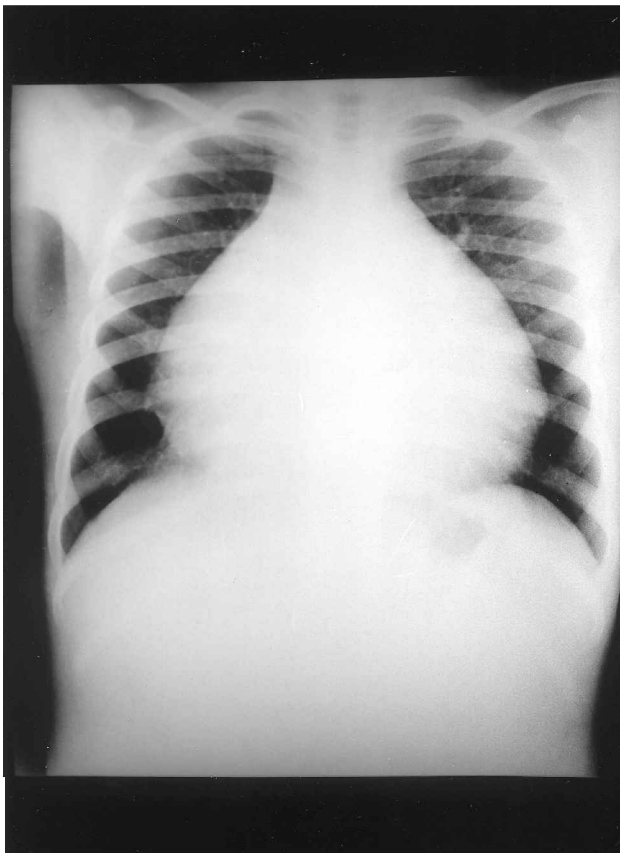

Figure 1.
Preoperative chest x-ray showing marked cardiomegaly.

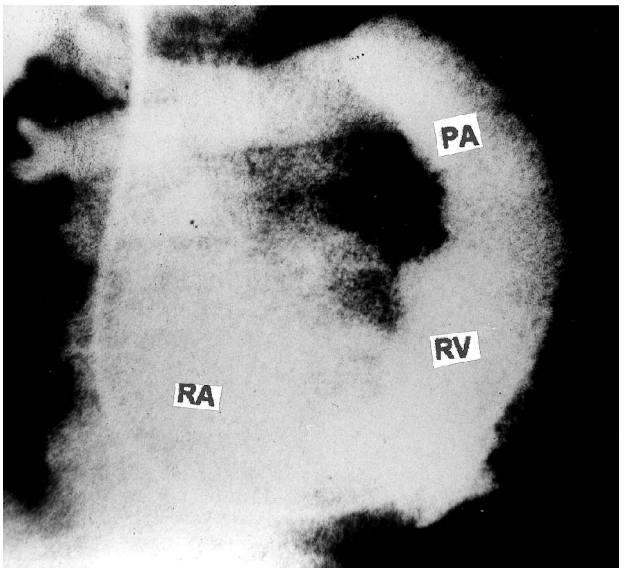

Figure 2.
Preoperative cineangiocardiogram showing a huge right atrium (RA), with amputation of the apical component of the right ventricle (RV), producing a tubelike configuration connecting the right atrium to the pulmonary trunk (PA).

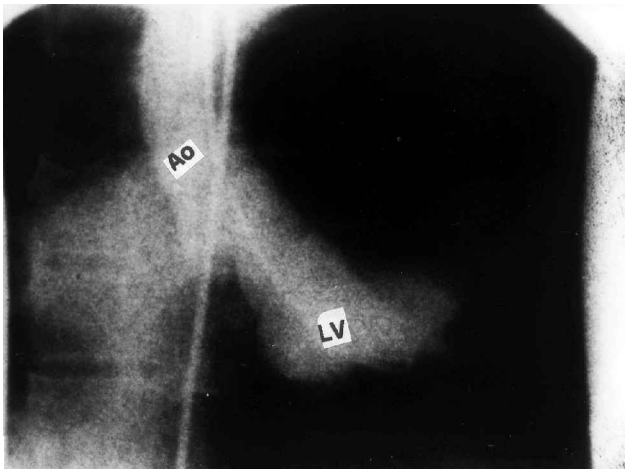

Figure 3.
Left ventriculogram showing a globular aspect of the left ventricle (LV).

The electrocardiogram disclosed no specific abnormalities. Eight patients had sinus rhythm, 2 had atrial fibrillation, one had right branch block, one had first-degree atrioventricular block, and 2 showed evidence of left ventricular hypertrophy.

Cross-sectional echocardiography performed in 7 patients demonstrated abnormal filling of the apex of the affected ventricle.

The definitive diagnosis in all cases was established by selective cineangiocardiography. Cineangiography of the right-sided cavities showed atrial dilation, tricuspid regurgitation, and an amputation of the apical component of the right ventricle with a characteristic tunnel-like formation that permits direct passage of the dye from the dilated atrium to the pulmonary trunk (Figure 2). Left ventricular cineangiography revealed a globular configuration of this chamber, again with obliteration of the apex (Figure 3).

Surgical techique

All patients underwent surgery through a median sternotomy, using conventional cardiopulmonary bypass lasting from 60 to 165 minutes, with a mean of 95 minutes. Myocardial protection was achieved by means of cold potassium cardioplegic solution, infused into the aortic root, and topical hypothermia of the heart. The intracardiac procedure was performed during a period of ischemic arrest. The duration of ischemic arrest ranged from 35 to 100 minutes, with a mean of 59 minutes.

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Endocardium decortication of the right ventricle was accomplished in 9 patients, of whom 3 had biventricular endomyocardial fibrosis and 6 the right-sided form of the disease. A large right atriotomy was performed in all cases. The remnants of the tricuspid valve were resected, and the fibrous tissue incised below the tricuspid annulus. The dissection was directed toward the ventricular apex into the posterior wall, allowing the resection of an entire fibrous shell. After the decortication had been completed, a valvar prosthesis was inserted in the tricuspid position.

Left ventricular endocardiectomy was performed in 4 patients, of whom 3 had biventricular disease and 1 had left-sided involvement alone. In all cases, a good exposure was obtained by a large vertical left atriotomy. The mitral valve could not be preserved in any patient, and the fibrosis of the left ventricular endocardium was resected using the same guidelines as those employed for the right side. A mitral valvar prosthesis was then inserted. In this series of 10 children, 8 received a bioprosthesis, and only 2 a mechanical valve.

Pathology

Histologic examination of the excised endocardium confirmed the diagnosis of endomyocardial fibrosis in all cases. The endocardial fibrous tissue consisted essentially of collagenous connective tissue with sparse elastic fibers and no infiltration of inflammatory cells (Figure 4). Areas of granulation were usually seen.

Results

There were 3 deaths during the postoperative period (30% hospital mortality), all resulting from low cardiac output, and all with biventricular disease.

The 7 survivors were followed up for a period ranging from 12 to 168 months (mean 72 months). There were two late deaths. One child did not improve clinically after the operation and died in severe heart failure 12 months later. The other one, the only patient with isolated left-sided disease, died 13 months after surgery as a result of bacterial endocarditis in the mechanical prosthesis implanted in mitral position.

Five (50%) patients are alive at the time of writing. Two required 3 reoperations for dysfunction of the inserted valvar prosthesis. One of these survivors is now functionally in Class IV, despite intensive medical treatment, and the other 4 are in Class II to III, all of them taking diuretics. Two patients showed echocardiographic evidence of progression of the disease to the non-operated ventricle.

Discussions

Endomyocardial fibrosis is a mysterious disease, its etiology and pathogenesis remaining obscure. The great majority of the reported cases have been from tropical countries, particularly in Africa, Southeast Asia and South America, but its occurrence in Europe and North America has also been described.

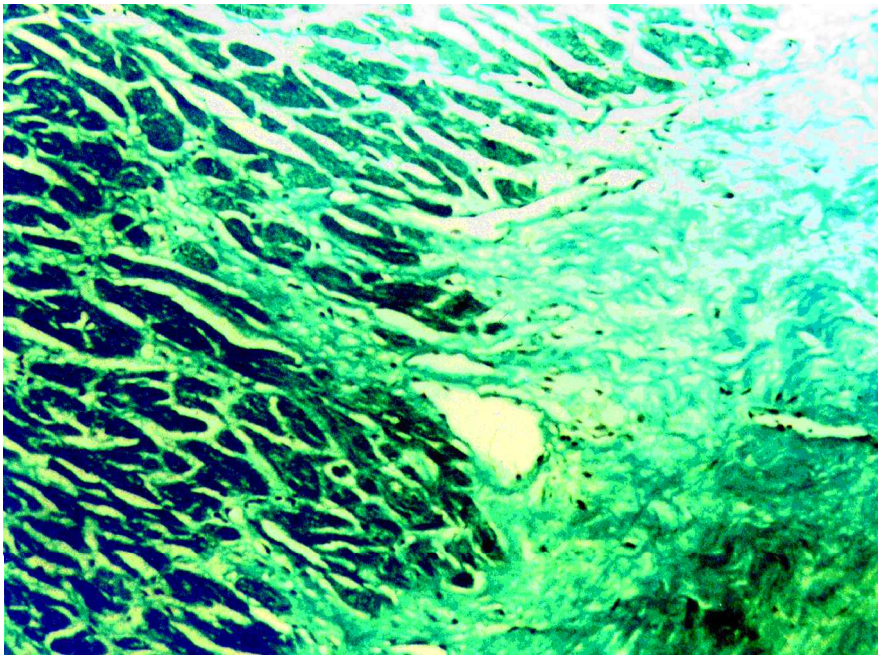

Figure 4.
Histological aspect of the removed endocardium, stained with hematoxylin and eosin. The endocardial fibrosis consisted essentially of collagenous connective tissue, with sparse elastic fibers in the absence of any infiltration of inflammatory cells. Note the extension of the fibrotic changes into the underlying ventricular myocardium.

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The incidence of the disease in children is difficult to establish. Reports from Africa usually mention that endomyocardial fibrosis is common among children and young adults, but exact figures are difficult to obtain.[13,14] Our experience in the last 22 years has shown an incidence of 10.9% in children under 15 years of age, accounting for 10 of our overall series of 91 patients undergoing surgery.

The clinical presentation of our children with endomyocardial fibrosis seems not to differ from that of adults. All were in the final stage of heart failure. Definitive preoperative diagnosis was made by selective cineangiocardiography, which demonstrated the typical ventricular deformities of the disease. Cross-sectional echocardiography is also a reliable method for diagnosis, showing obliteration of the apical components of the ventricular cavities by fibrosis.

The downhill course of patients with endomyocardial fibrosis, and the irreversible nature of the disease, are well known, most patients dying within 3 years of diagnosis.[15] This grave prognosis is no different in children. Medical therapy is usually ineffective. The only hope for survival is surgical treatment.

The operative technique is well-established,[7] and consists of resection of the fibrotic endocardium, a process which is facilitated by the presence of a plane of cleavage between the fibrosis and the underlying myocardium, and repair or replacement of the atrioventricular valve. Early and late results of surgical treatment are well described.[16–21] The high operative mortality, of up to one-fifth, observed in most of the reported series indicates the severity of the cardiac cachexia of most patients at the time of operation. It seems to be worse in children who, as shown by our experience, have always presented in the final stage of heart failure and in poor preoperative general condition.

The long-term outlook of patients submitted to surgical treatment of endomyocardial fibrosis has also been the subject of recent analysis.[19–21] Although it has been reported that surgery improves both functional capacity and the quality of life in the majority of surviving patients, it is recognised to be a palliative procedure.[21] Endomyocardial fibrosis seems to be an evolving disease, as demonstrated by the fact that some patients continued to present with endocardial fibrosis in the postoperative period, either as a recurrence in the operated ventricle or in the previously normal one[21]. Moreover, some patients do not improve clinically after the operation, as was observed in one of our children, probably as a result of severe infiltration of the myocardium by fibrous tissue.[21] Another fact that may adversely affect the postoperative course, especially in children, is dysfunction of the valvar bioprosthesis. Because of this, every effort should be made to preserve the native valve during the operation.[9,21] Unfortunately, in our experience with 10 children, despite recognising this imperative, it did not prove possible. We have preferred to insert bioprosthetic valves in the majority of patients because their low socioeconomic condiction precludes anticoagulation.

In conclusion, endomyocardial fibrosis, when encountered in childhood, is a serious disease. The results of surgical treatment are far less satisfactory than one would wish. It is obvious that children are at a disadvantage compared to adults in that they normally have a greater life expectancy. Despite the disappointing results, we continue to recommend surgery because of the ineffectiveness of medical therapy. Efforts should now be concentrated on elucidating the etiology of the disease, which might permit the establishment of preventive measures.

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