The Surgical Treatment of Constrictive Fibrous Endocarditis

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Constrictive fibrous endocarditis is a pathological entity described by Loëffler in 1936. Its etiology is unknown. The clinical course is characterized by an evolution towards cardiac insufficiency leading rapidly to a fatal outcome. Modern paraclinical investigations are necessary to assess the diagnostic. Cardiac catheterization brings the proof of adiastole and angiocardiography reveals the shape of amputation of the ventricle with auriculo-ventricular regurgitation. The operative procedure consists of resection of the ventricular fibrosis including the valves and auriculo-ventricular valve replacement by a prosthetic valve. The disease affects both Caucasians and Negros. Our experience includes 5 cases. The indications for operation and their results are discussed.

CONSTRICTIVE FIBROUS ENDOCARDITIS, first described by Loëffler[6] in 1936, consists of endocardial fibrosis several millimeters in thickness which is potentially constrictive and limited to the ventricular cavities.

The clinical course is characterized by an evolution towards cardiac insufficiency which may be predominantly right or left sided leading rapidly to a fatal outcome.

Modern paraclinical investigations such as cardiac catheterization and angiocardiography have defined the hemodynamic characteristics of the malady as being caused by adiastole which may be right or left sided or bilateral as well as asystole.

The etiology remains obscure and the pathologic changes are non specific.

The inevitable evolution towards death stimulated us to consider the possibility of a new operation which would consist of decortication of the fibrosed endocardium in a manner similar to resection of the constrictive pericardium of pericarditis.

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Endocardiectomy was first performed by us in 1971[5–7] and the patient was improved. Since then, four more cases have been done, either right or left sided or combined endocardiectomy and there is no doubt that a better knowledge of the clinical aspects of the disease will increase the number of cases referred for treatment.

Pathology

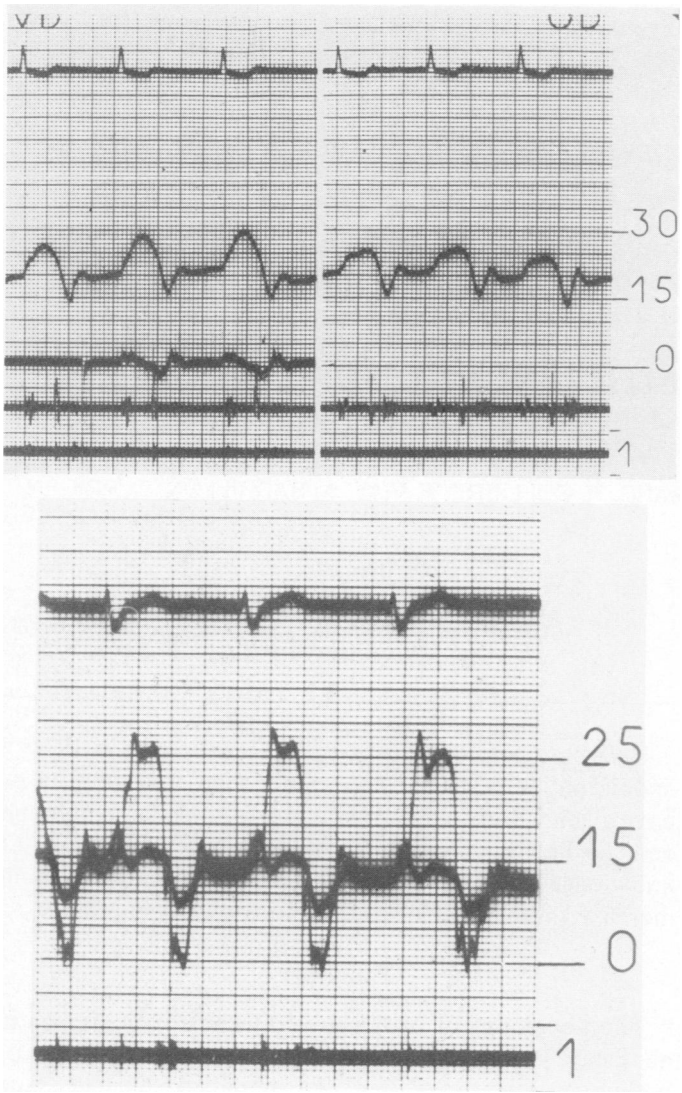
The development of fibrous endocarditis is similar in all cases: it involves the filling chambers of one or both ventricles including the papillary muscles and chordae tendinae of the mitral and tricuspid valves which themselves are included in the process in the majority of cases.[8]

It is of interest to note that in none of our cases did the fibrosis extend to the atria. The thickness of the fibrous process varies from 4 to 10 mm. The underlying myocardium presents fibrous lesions which are localized to the subendocardial layer.

Histologically, there is fibrous thickening of the endocardium which is made up of collagen without elastic fibers and few fibrocytes. This fibrous shell has on its surface small islets of hyalinized fibrosis deposits which are in the process of being incorporated. The papillary muscles are covered by a thick collagenous endocardium associated with a subendocardial sclerosis of the adjacent myocardium. The valvular and subvalvular apparatus are moderately thickened by a fibro-myxoid process.

Clinical Course

The presentating signs are usually severe right and left heart failure.[1,2]



FIGS. 1a and b. Right heart catehterization (Top) preoperative-equal pressure in the ventricle and atrium. Dip-plateau tracing. (Bottom): postoperative, return to normal tracing (Case 3).

The onset symptoms are most often recent but it is possible to observe periods of remission which are variable in duration and which are followed by relapses until end-stage irreversible heart failure. It is interesting that in 2 of our cases, cardiac transplantation was eventually suggested because of the gravity of the patient's illness.[4]

The right side forms are characterized by peripheral edema, frequently massive hepatomegaly and ascite.

The left side forms affect the small circulation and are characterized by repeated episodes of pulmonary edema.

Auscultation reveals the signs of mitral insufficiency with or without tricuspid insufficiency.

The electrocardiogramm is non specific but low voltage is noted.

The chest X-ray reveals the existence of a cardiomegaly which is often most marked at the atrial level.

Catheterization[5] of the right side of the heart shows equal pressure in the ventricle and the atrium. The tracing has a dip plateau which is characteristic of the constriction. In the left ventricle, the tracing confirms the adiastole with a plateau-like elevation of mid and telediastolic pressures. Postoperative studies show normal atrial and ventricular pressure curves with a disappearance of the plateau tracings.

Angiocardiography of the right sided cavities reveals atrial dilatation with stasis of the contrast medium. There is an amputation of the filling chamber of the ventricle with a direct passage between the atrium and the pulmonary artery. On the left side, the ventricular apex is deformed in a characteristic configuration resembling the "ace of hearts" and there is an associated marked mitral insufficiency with atrial dilatation.[2]

Postoperative studies demonstrate the opacification of the filling chamber of the right ventricle as well as the return of normal shape of the left ventricle.

Evolution

Medical treatment is capable of slowing down for a time the evolution of disease; after cardiotonic drugs and diuretics are no longer effective, cachexia leads to death.

Surgical Treatment

The operative procedure is through a median sternotomy. Extracorporeal circulation is established by cannulating the ascending aorta and both vena cava. Moderate hypothermia to 30° is obtained by perfusion.[7]

The left atrium is opened vertically and the mitral orifice is exposed. The lesions are characterized by a fibrosis which covers the entire filling chamber of the ventricle, encompassing the papillary muscles, the chordae tendinae and both mitral valve leaflets. The fibrosis never extends beyond the mitral annulus into the atrium and it never involves the aortic orifice.

The simplest way to perform the decortication is to resect the mitral apparatus and to incise the fibrous tissue at its junction with the mitral annulus as to find the plane of cleavage between it and the underlying myocardium. The dissection is pursued towards the ventricular apex at first on the posterior wall followed by the anterior wall. It is important to sharply dissect the fibrous bands which joins the endocardium shell to the cardiac muscle. Towards the ventricular apex, the adhesions are frequently dense and it is not unusual to find calcifications in this area.

At the level of the mitral papillary muscles, it is essential to divide these on their bases which are most often completely fribrotic.

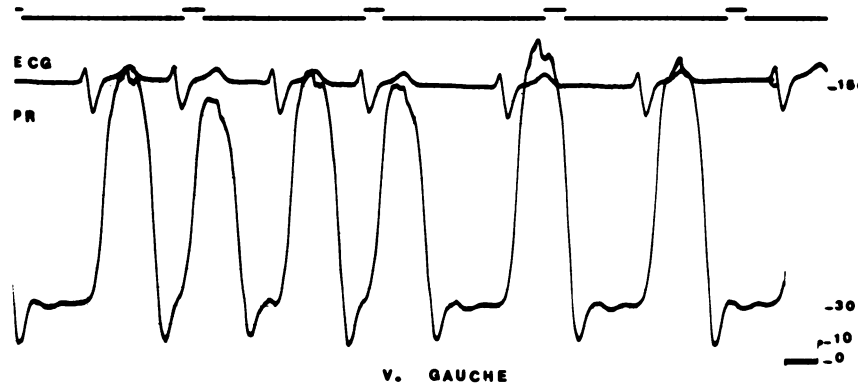


FIG. 2a. Left heart catheterization, preoperative: mid and telediastolic plateau (Case 1).

During the resection, it is indispensable to remain in contact with the fibrosis without cutting into the adjacent myocardium. In this way, it is possible to resect the entire fibrous shell which models the interior surface of the ventricle from the atrioventricular orifice to the apex. The mitral valve is then replaced by an artificial valve, either a Starr prosthesis with a silastic ball, a Björk valve, or a heterograft valve.

The right atrium is then incised vertically anterior to the vena caval cannulas. Right ventricular fibrosis is similar to that of the left ventricle: the filling chamber alone is involved, the outflow tract is free as is the atrioventricular annulus. There is one difference however: the right atrium is dilated and its wall is thickened and in 4 of our cases an organized thrombus was present in the auricular appendage and ajacent portion of the atrium. The resection of the ventricular fibrosis is performed using the same guidelines as for the left side and the tricuspid valve is replaced with an artificial valve, the heterograft valve being our primary choice. The thrombosed area of the atrial wall is resected. Both atriotomies are closed and the heart defibrillated when the temperature reaches 36°.

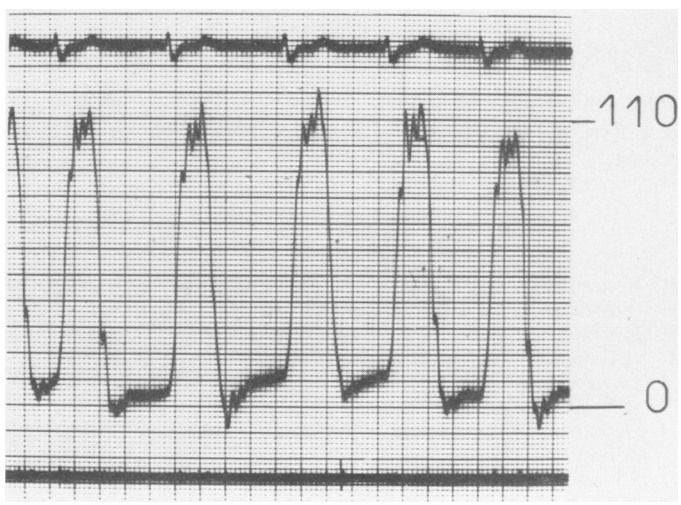


FIG. 2b. Postoperative: return to normal tracing (Case 1).

This combined form of endocardial fibrosis does not encompass all the aspects of the disease. There are isolated right sided forms, isolated left sided forms, left sided forms associated with functional tricuspid insuf-

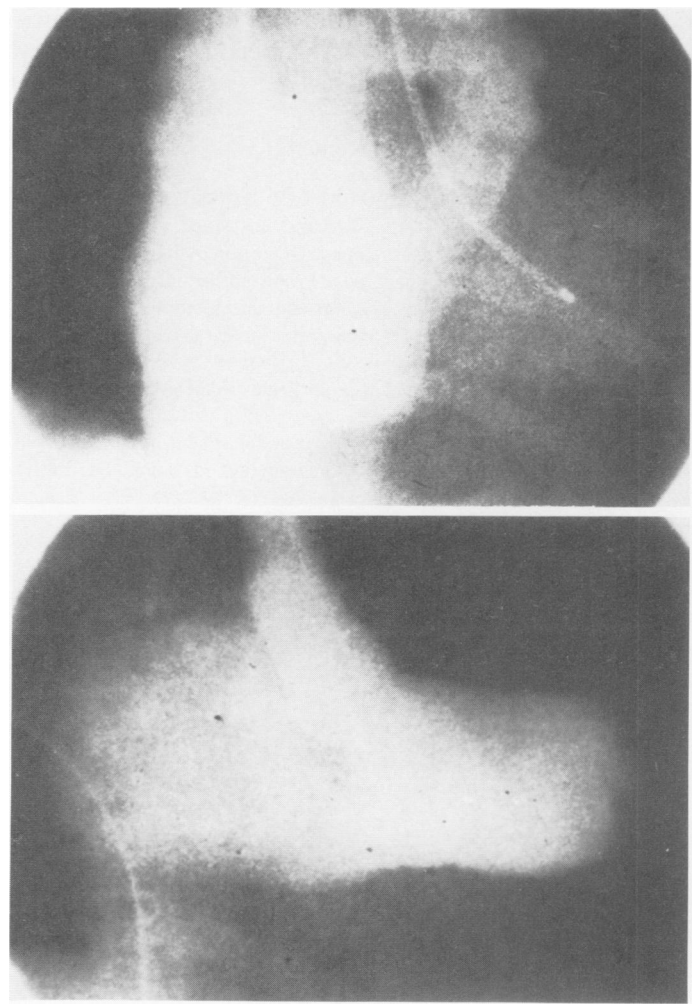


FIG. 3. Cineangiography of right cavities (Top) showing stasis of contrast medium in a large atrium associated with amputation of filling chamber of the right ventricle. Left cavities (Bottom): amputation of the left ventricular apex with deformity of the "ace of hearts." Marked mitral insufficiency with dilatation of the left atrium (Case 3).

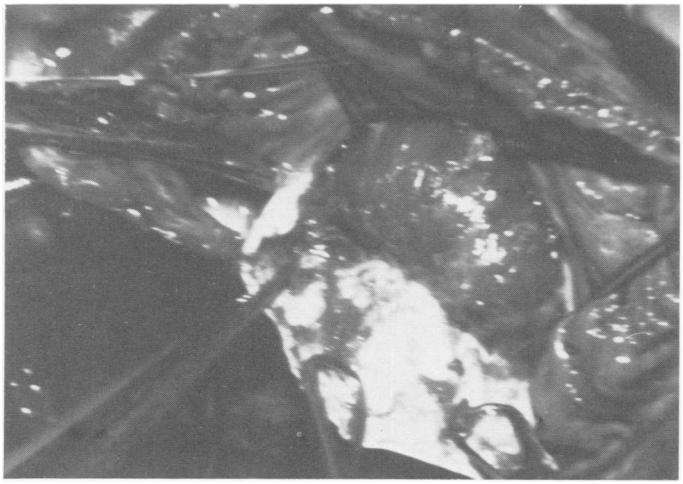


FIG. 4. Intra-operative photograph showing resection of left ventricular endocardial fibrosis (Case 1).

ficiency, without endocardial fibrosis of the right ventricle. In the latter instance, a Carpentier-ring valvuloplasty is to our eyes the best solution.

Case Reports

Our experience is composed of 5 operative cases.

Case 1. Caucasian man, age 58 years, left sided form associated with a functional tricuspid insufficiency. Evolution over a 9 year period. In the last stages of cardiac cachexia. Proposed for cardiac transplantation. Operation: September 1971: left endocardiectomy. Mitral valve replacement by Starr valve. Tricuspid annuloplasty. Good result after 4 years and 8 months.

Case 2. Negro boy, age 12 years. Right sided form with rapid evolution.

Operation: April 1974. Right sided endocardiectomy. Tricuspid heterograft. Atrio-ventricular block. Pace-maker insertion. Died after 20 months. Dysfunction of prosthesis.

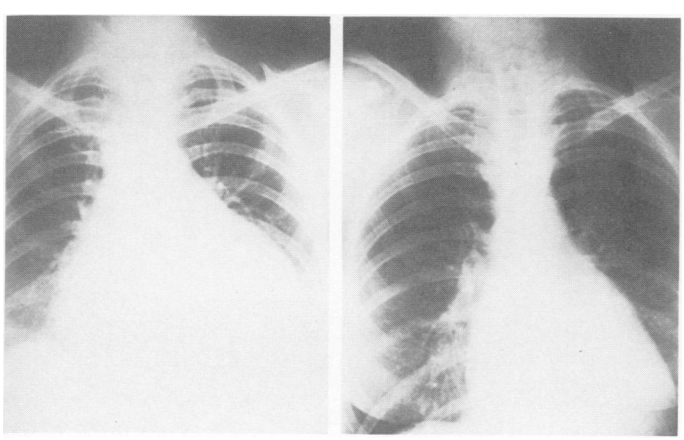


FIG. 5. Comparison between preoperative chest Xray and Xray taken 6 months following endocardiectomy (Case 3).

Case 3. Caucasian woman, age 40 years. Biventricular form in the last stages of cardiac cachexia. Cardiac transplantation discussed. Operation: July 1975, biventricular endocardiectomy. Two Björk prosthetic valves. Excellent result after 10 months.

Case 4. Caucasian woman, age 43 years, left sided form in poor condition. Operation: January 1976. Left endocardiectomy with valve replacement by a Björk prosthesis. Excellent result after 3 months.

Case 5. Negro woman, age 29 years. Biventricular form in advanced stage. Operation: February 1976. Right and left endocardiectomy. Mitral valve Björk prosthesis and tricuspid valve replacement by a heterograft valve. Excellent result after 2 months.

Conclusion

Löeffler's fibroplastic endocarditis and Davies[3] endocardiomyopathy are probably the same disease. From a clinical, pathological or surgical point of view there is no difference between them.

The constrictive character of the disorder is important: it explains the adiastole, the foremost clinical sign. The cause of the disease is unknown, it seems that at present time, the filarial etiology may be excluded. Constrictive fibrous endocarditis affects members of the Caucasian race who for the most part have never resided in Africa.

Neverthless, the frequency of the disease in Central Africa has stimulated one active epidemiological research, especially in the Ivory Coast.

In certain terminal cases the irreversible nature of the illness has brought up for discussion the admissibility of cardiac transplantation.

The operation which we have described in our 5 patients appears to offer a good chance for long term survival.

In the present status of our knowledge, it may be recommended as the therapeutic method of choice, even in the most advanced stages of the disease.

Research should be undertaken in the hope of learning the etiology of the syndrom and the reasons of the distribution, especially its frequency in certain regions of Africa.

Addendum

Since this paper was submitted for publication three more cases were operated on successfully, bringing our total number of cases up to eight.

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Discussion

Professor Åke Senning (Zurich, Switzerland): During the 15 years I have been in Switzerland, I have not recognized any cases of constrictive fibrous endocarditis until I heard Dr. Dubost's paper in Detroit last fall. Since then we have seen one case, which we operated upon—one month ago. It was a young woman who had a very typical left ventriculogram and a mitral insufficiency. We had the impression that the apex of the left ventricle couldn't dilate. The mitral valve was replaced, and an endocardiectomy done, as Dr. Dubost described, and the patient is in very good condition one month later.

I think we may find more of those cases, and I suspect that we have one case on the waiting list to be operated upon.

President Hardy: A colleague of mine called me in to look at a case a while back, and it was very much like this, with trabeculations and synechia, so I think it does occur in this country.

Dr. James V. Maloney, Jr. (Los Angeles, California): I recall that my pediatric cardiologist associate, Dr. Forest Adams, in 1958 spent the better part of a year attempting to get me to perform this operation on a series of young infants, in whom this condition is relatively common, intracardial fibroelastosis.

We had an epidemic at that time of a disease which we suspected was related to nutritional deficiencies caused by synthetic baby foods. At that time we found, as Dr. Dubost has, that the disease is extremely common in Africa, and is associated with kwashiorkor.

In retrospect, perhaps it is as well I did not operate on those patients at that time, because it was before the availability of prosthetic values, and simply removing the abnormal endocardium might not have resulted in a favorable result, because, I notice, each of these patients required valve replacement.

In view of the recent work of my associate, Dr. Buckford, on the demand-supply ratio as a cause of subendocardial necrosis, I think my present speculation would be that this disease is caused by an abnormal demand-supply ratio, probably less than 0.7, related, perhaps, to congenital heart disease, perhaps metabolic deficiency, and perhaps abnormal cardiac function in patients with nutritional disease. I think the fact that now and in the future it will be amenable to surgery is a great tribute to Professor Dubost.

Dr. Charles Dubost (Closing discussion): I would like to emphasize the fact that this disease occurs in both Caucasian and Negro people. The majority of cases occur, probably, in Africa, and for a long time it was believed that Filaria was the cause of the disease, but now it is admitted that this is not true. The disease occurs in patients with filariasis, but it is not due to Filaria. The first description by Löffler of the disease emphasized the importance of the presence of eosinophilia, but it is known that eosinophilia very often accompanies many diseases of Africa.

The most important notion is that this disease is of unknown cause, and that it occurs in wealthy Caucasians, and in poor Negros. It makes no difference. It is quite the same, and we had better try to operate on the people early in the evolution of the disease.

Now that we know that the disease can be operated, you will find it more often. It looks like a constrictive pericarditis but the angiogram makes the diagnosis different due to the lack of opacification of inflow chambers of the ventricles.

So when you see a constrictive pericarditis which is not constrictive pericarditis, it is possibly constrictive endocarditis.