Chronic Constrictive Pericarditis

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CHRONIC constrictive pericarditis, interestingly enough, was one of the very first pathologic conditions to be recognized. This is natural in view of the rather obvious gross lesion found at autopsy. Hence when the custom of postmortem examinations began to be established, pericarditis and its sequelae were quickly identified. Lower, in 1069, described the clinical effect of interference of cardiac diastole by a constricting fibrous pericardium. Chevers, however, in 1842, was the first to present clearly the clinical picture of chronic constrictive pericarditis. Wilks, in 1870, further emphasized this syndrome. Curiously enough, however, it was Friedel Pick whose name was attached many years later to the disease itself, which fact illustrates the neglect of the writings of our forebears.

An extraordinary delay supervened, following these early descriptions of the disease, before it was shown that the clinical diagnosis could be readily made and that surgical correction was feasible (White, 1935). Although much is now known about chronic constrictive pericarditis and its treatment, new advances are likely to lead eventually to its reduction and to its earlier recognition and proper treatment.

INCIDENCE

Chronic constrictive pericarditis is still a rare disease but is being recognized more frequently. In a large hospital such as the Massachusetts General in Boston, several new cases are encountered each year. Doubtless with a reduction of tuberculosis the disease will become more and more rare.

ETIOLOGY

For many years the cause of chronic constrictive pericarditis remained in doubt. Although it is still impossible in most cases to identify the etiology at operation or autopsy, it is quite certain in the opinion of many that the tubercle bacillus is the causative agent (Pickering, Paul and co-workers). Pickering has followed patients from the stage of acute tuberculosis of the pericardium to that of chronic constriction. Such an evolution, as a rule, takes many months or even a few years. The initial stage often is not observed. Observation of the initial stage will probably be possible more often in the future than in the past because of the institution of careful examination at the time of any acute illness in children and young adults. The active stage of tuberculous pericarditis may be readily overlooked. Malaise and slight fever may be the only symptoms, even in the presence of a large pericardial effusion which has developed so slowly that the body has accommodated itself. Children have actually been discovered at play with serious, acute, tuberculous involvement of the pericardium. This is quite different from the acute pericarditis of other cause, usually infectious, that is more likely to produce pain, rapid accumulation of fluid resulting in cardiac tamponade, and higher fever.

It is quite possible that other etiologic fac-
tors than the tubercle bacillus may infrequently result in chronic scarring of the pericardium with constriction. At one time pyogenic infections were blamed; they may continue to be factors in the future, especially with recovery from such serious illness made possible by antibiotic therapy. However, such etiologic factors exist almost certainly in but a small minority. The same is true of trauma which can produce hemopericardium; chronic constriction from such a cause is conceivable but must be very rare. Rheumatic fever, although frequently accompanied by pericarditis when there is pancarditis, does not result in chronic constrictive pericarditis. This can be stated quite definitely. Possibly there are rare exceptions but I myself have never encountered one. Rheumatic fever, to be sure, may leave pericardial adhesions, but these are not the massive constricting adhesions which we encounter after tuberculous infection. In only one case of some sixty with chronic constrictive pericarditis, have I noted a coincidental rheumatic valvular defect. In one other case was a congenital septal defect. These coincidental lesions, however, may be expected in the population at large in occasional cases. The pericarditis associated with uremia and with myocardial infarction is not of the constricting type.

Pathologic Anatomy

Chronic constrictive pericarditis consists of fibrous thickening of visceral pericardium, of parietal pericardium, or of both surfaces, which are usually welded together by organization of the exudate which originally had been present in the sac itself. On occasion there may be slight, relatively unimportant, constriction by the fibrous parietal pericardium with very little involvement of the visceral pericardium; the reverse may also be true. In the average case, however, the pericardial sac is obliterated and both surfaces are involved about equally.

The fibrosed pericardium varies in thickness from 1 to 2 mm. up to 0.5 cm. or more. It also varies in distribution over the heart and great vessels, sometimes more in front over the right heart chambers and sometimes more in back over the left. As a rule, there is involvement around the entire heart and base of the great vessels, but some of the pericardium is frequently left rather thin and nonconstricting; there may be actual pericardial pockets free from involvement or containing residual fluid despite extensive fibrosis elsewhere. Although separate fibrous bands have been reported around the great veins or great arteries, these are much less common and important than the general involvement of the heart chambers themselves. There are, however, two areas of heart surface which may be importantly constricted. One is the right or left auriculoventricular groove, particularly the left, which produces a mechanical obstruction resembling mitral stenosis; the other is the interventricular groove, either along the course of the descending branch of the left coronary artery or in back adjoining the posterior coronary vessels. There may be actual interference with the coronary blood flow as a result of such contiguous pressure.

The laying down of calcium, sometimes in the form of granules like sand which may be loose in the pericardium, is a common complication of chronic constrictive pericarditis. Calcification is found in about one-third of the cases and at times results in the formation of massive plaques of calcium like a tortoise shell over large areas, filling the auriculoventricular grooves or even penetrating into the myocardium itself. It should be added, however, that in a few cases calcification of the pericardium without appreciable constriction may be found by x-ray study or at autopsy.

The myocardium itself may be relatively normal throughout the heart although there are two difficulties that should be noted. In the first place, subpericardial myocarditis with some chronic scarring is usually present to a varying degree. Secondly, there is an atonic state of the myocardium when it is encased rigidly in calcium which prevents its proper function. The muscle is there but it tends to be atrophic and at times is interspersed with scar tissue.

The size of the heart varies greatly. Classic chronic constrictive pericarditis has been said to be accompanied by a heart smaller than normal and atrophic. Although this is possible, it is more common to find some cardiac enlarge-
ment. To be sure, a slight increase of the heart shadow seen by x-ray can be explained by the thick pericardium, which may add 0.5 cm. to each border of the heart shadow, but various strains on the heart and perhaps the diastolic fixation of some of the heart by the scarred pericardium explain the enlargement that is frequently found. Such enlargement is not great, as a rule, and the heart may be only slightly above normal in volume or weight. There are a few instances where the right ventricle is much larger than usual as a result of constriction of the left heart chambers, causing a disease picture much like that in mitral stenosis. The auricles may also be more capacious than normal, whether they are 'involved in the constricting process or not. The valves are usually normal, although there may be insufficiency resulting from dilatation of the ventricular cavities.

Chronic constrictive pericarditis is occasionally accompanied by chronic constricting pleuritis and even chronic fibrous peritonitis. Such concomitant lesions indicate the primary occurrence of polyserositis, also doubtless tuberculous in origin. The thickened peritoneum over the liver is due not to the engorgement of the liver itself in so-called Pick's disease but rather to the occurrence of acute peritonitis at the time of the acute pericarditis followed by residual fibrosis.

Pathologic Physiology

Little need be said about the pathologic physiology of chronic constrictive pericarditis. It was quite simply explained centuries ago by Lower and again over a hundred years ago by Chevers. The difficulty consists mainly in the inability of the heart to assume its proper size in diastole and to contract properly in systole, that is, there is a mechanical interference with the normal systole and diastole, especially the latter. Thus, the blood cannot enter the heart properly and is backed up in the great veins and liver and sometimes in the pulmonary blood vessels. There is also an insufficient output of blood, causing a certain amount of forward failure for which the muscle is not primarily responsible.

When the left heart chambers are preponderantly involved there is, as in mitral stenosis, an increase in pulmonary blood pressure which should be measured in every case by cardiac catheterization. The determination of the pulmonary blood pressure is important because it aids in the decision on surgical technic. If the pulmonary blood pressure is increased in chronic constrictive pericarditis, the surgeon should release the left heart chambers first; in such cases the surgical approach must be so arranged as to reach the back of the heart primarily.

If the right heart chambers are preponderantly constricted, there is naturally engorgement of the neck veins and liver, with relatively little pulsation evident. On the other hand, when the right heart fails because of constriction of the left heart chambers, then there tends to be a vigorous systolic jugular pulse and some pulsation of the liver, as in any case of total heart failure, except that in cases of chronic constrictive pericarditis the deep systolic pulsation becomes chronic and may continue for years, simulating the effect of tricuspid valve disease.

With diminution in the output of the heart, which is not uncommon, the blood pressure, especially the pulse pressure, tends to be low. It is not unusual to find the systemic blood pressure to be 100 mm. Hg systolic and 80 diastolic.

Both the irritation of the heart by the actual process involving the myocardium and the effect of constriction of the left heart chambers on the right ventricle and right auricle result not infrequently in auricular arrhythmias. Auricular fibrillation and less commonly auricular flutter occur as complications in chronic constrictive pericarditis. About a third of our cases have shown such arrhythmias, which add an increased burden to the heart; in such patients there must be control of the heart rate by digitalization.

Symptoms

The symptomatology of chronic constrictive pericarditis can be prophesied and simply explained when one understands the pathologic anatomy and physiology of this disease. It is, of course, of much interest to recognize the
initial acute pericarditis which is usually attended by malaise, some febrile reaction, and not infrequently some evidence of cardiac tamponade from a pericardial effusion.

The acute tuberculous pericarditis may be fatal per se or as a result of the complication of miliary tuberculosis. On the other hand, it may pass insidiously from the acute to the subacute and finally to the chronic stage. A year or two or three may elapse between the occurrence of the acute pericarditis and the beginning of evidence of chronic constriction. Sometimes the disease is discovered accidentally during routine examination for insurance or for military service, particularly when x-rays reveal calcification of the pericardium or when the astute observer notices a little increase in jugular pulse or enlargement of the liver or looks for the cause of an unexplained arrhythmia.

Actually there are very few symptoms of chronic constrictive pericarditis. The signs are more important. Dyspnea can occur if the left heart chambers are preponderantly affected, as in mitral stenosis, with engorgement of the lung vessels and decrease in vital capacity, but more commonly the liver enlarges and the abdomen increases in size causing discomfort to the individual affected. There may even be a little ascites before much attention is paid by the patient to his disease. Less commonly there is unexplained swelling of both legs early in the clinical course. The liver enlargement and ascites are more prominent than is edema of the legs. Fever occurs only if there is acute or subacute infection or recurrence thereof. Pain, as a rule, is absent and palpitation is not noted except in a few instances of tachycardia secondary to auricular fibrillation or flutter.

**Signs**

The signs of chronic constrictive pericarditis allow the diagnosis to be made readily in most instances. When, in the absence of clearcut etiology of the ordinary causes of heart disease such as congenital defects, rheumatism, hypertension, and coronary atherosclerosis, with little enlargement of the heart and usually normal rhythm, the neck veins are found to be full and the liver enlarged, one should think at once of the possibility of chronic constrictive pericarditis. The presence of ascites makes this even more likely. Heart murmurs are not the rule. On occasion there may be a systolic murmur at the apex associated with some mitral regurgitation or at the lower end of the sternum associated with tricuspid regurgitation, but important murmurs are invariably lacking. The pulmonary second sound may be increased if there is pulmonary hypertension. This is, however, found in the minority of the cases. In patients with dilated right ventricle a third sound may be heard along the left border of the sternum indicative of right ventricular enlargement. The lungs are generally clear, although in advanced cases there may be right hydrothorax, or pleural adhesions secondary to chronic concomitant tuberculous pleuritis. A complication that may be serious is that of engorgement and varicosities of the leg veins with possible thromboses. If arrhythmia is present, there may be a rapid regular or rapid irregular pulse rate and the blood pressure tends, as already stated, to be low with a small pulse pressure. The venous pressure, on the other hand, is very much elevated, frequently exceeding 200 mm. of water and on occasion even exceeding 300 mm. One other important sign in advanced cases is that of the paradoxical pulse, the blood pressure decreasing even to the point of disappearance during inspiration in some of the more advanced cases.

**X-ray Examination**

Usually the heart shadow on the x-ray film or seen with a fluoroscope is abnormal but there are a few instances in which little or nothing wrong can be found in the constricted heart. Cardiac pulsation is decreased as a rule, sometimes over one part of the heart shadow more than another. The heart varies in size and position. There may or may not be calcification. In about a third of the cases, bands of calcium can be seen, better in the oblique than in the anteroposterior view. The sheets of calcium are better seen end on, of course, than when viewed at right angles to the surface. Small areas of calcification may be undiscernible by x-ray examination.
ELECTROCARDIOGRAPHY

The electrocardiogram is usually very revealing; it shows a fairly distinct pattern of pericardial nature with lowering of voltage in most limb and precordial leads, and abnormality of the T waves without much change in the QRS complexes except for decreased voltage. The T waves tend to be flat or slightly inverted in all limb and precordial leads. The electrocardiographic pattern in chronic constrictive pericarditis is generally easily distinguishable from that produced by coronary heart disease. Arrhythmias are, of course, well shown.

One of the interesting evolutions of the electrocardiogram in cases that are observed for a long time is the development of evidence of right ventricular preponderance, sometimes accompanied by the onset of auricular fibrillation or flutter, when the constriction of the left heart chambers produces steadily increasing enlargement of the right ventricle. This is a telltale clue.

CARDIAC CATHETERIZATION

An important new technic which I would now consider essential to the study of any case of chronic constrictive pericarditis is determination of the pulmonary blood pressure by cardiac catheterization. Normally the systolic pressure in the right ventricle and pulmonary artery does not exceed 25 mm. Hg. If there is preponderant constriction of the left heart chambers, the pulmonary blood pressure may be twice or thrice the normal. As an aid to determining which part of the heart is most involved and, from this, the surgical approach, knowledge of the pulmonary blood pressure is of great help.

Course and Prognosis

Chronic constrictive pericarditis varies in its course from that of a fulminating disease, resulting in hopeless invalidism and death within a relatively few months or a year or two, to a chronic condition that causes relatively little disability and does not shorten life. In the average case the disease continues for ten to twenty years, unless some serious complication occurs to shorten life or unless, more optimistically, a cure is effected by surgical intervention. Before surgical therapy was introduced, I remember having followed several individuals for a decade or more who needed abdominal paracentesis perhaps once a month. Successful surgical treatment can apparently restore normal health and full length of life. The first case successfully treated by surgery in the United States, reported by Dr. Churchill and me14 over twenty years ago, has continued in excellent health so far as the circulatory apparatus is concerned. She is now 40 years old, a well and active woman who has raised a family.

One other patient may be cited to illustrate the occasional unimportance of the condition. A doctor practicing in Nova Scotia came to see Dr. Churchill and me in 1930; he showed evidence of chronic constrictive pericarditis of slight to moderate degree. He was then 57 years old. He gave a history of acute pericarditis of unknown nature while he was at Johns Hopkins Medical School in Baltimore in 1894. He began medical practice in Nova Scotia but after a few years, noticing some enlargement of the liver, he came to the Massachusetts General Hospital for study and there was diagnosed as having liver disease, probably cirrhosis, of unknown origin. When we examined him in 1930 we found him in fairly good health and, because of his age and relative well being, advised against surgical intervention. He continued to practice until 1939 and died in 1948 at the age of 76.

Treatment

Until about 20 years ago the treatment of chronic constrictive pericarditis was very discouraging. It consisted in efforts to keep the congestion under control. This congestion was often represented by general anasarca with preponderant liver enlargement and ascites. Not infrequently it was necessary to perform regular abdominal paracentesis. Although Finsen1 had shown that restriction of salt and fluid was effective in controlling the ascites, it was not customary years ago to advise a low sodium intake. Digitalis was known to be ineffective. Some diuretics were employed.

Since 20 years ago, both medical and
surgical treatments have improved greatly. Some individuals who are too old or too seriously involved to permit surgery are benefited by medical measures; any victim of the disease with tachycardia due to auricular flutter or to auricular fibrillation is better with a control of the heart rate by digitalis, whether or not surgical therapy is also employed. Since the advent of mercurial diuretics and strict low sodium diets, some patients can be kept reasonably comfortable without surgery, but the majority of patients are still in need of and are benefited by pericardial resection. A few mild cases, such as the Nova Scotian physician already referred to, can get along without surgery and with moderate medical measures.

The surgical treatment of chronic constrictive pericarditis was first recommended by Delorme in 1898, but the operation was not performed until some years later. A few cases were operated upon in Germany in the early part of the century, but only in the last twenty years has surgery become the routine method of choice. Pericardial resection, which is very different from Brauer's operation of freeing the heart from the thoracic cage, is a delicate, time-consuming procedure, somewhat hazardous, but under modern conditions of expert anesthesia and thoracic surgery no longer the dreaded ordeal that it was many years ago.

When surgery was first utilized for chronic constrictive pericarditis, it was the custom to approach the heart anteriorly and to free the right heart chambers. Even at that time, however, it was realized that if the whole heart were affected it would be wise to decorticate the left heart chambers first. Surprisingly, the majority of patients who were operated upon during the first few years of pericardial surgery were successfully treated through the anterior approach. As time went on, however, it became evident that occasional failures were due to the inadequate freeing of the heart over the anterior chambers only. Hence of late years decortication of both ventricles has been found advisable, either by a lateral approach or best by splitting the sternum and opening the thorax wide. Most observers now agree that it is wisest to free the left heart chambers first even if a second operation is necessary to release the anterior heart chambers. Cardiac catheterization with a determination of the pulmonary blood pressure is very helpful in indicating at once whether it is necessary to decorticate posteriorly first. With an elevated pulmonary blood pressure, the first attack should be on the left heart chambers.

Details of the pericardial resection can be found in the writings of Beck, Churchill, Sweet, and others.

Complications during or after operation can be disturbing. On several occasions the heart wall has actually been punctured, with resultant vigorous bleeding. However, rents in the auricle or ventricle can be and have been repaired, with survival and much improved health postoperatively. Rarely, especially if there is subacute activity of the process, there may be a draining fistula which tends to close with time. In a few instances there have been venous or intracardiac vascular thromboses with resultant pulmonary or peripheral embolism. There has been persistence of congestion in some cases as a result of either inadequate freeing of the heart or failure of the right ventricle after it has been freed or disturbing arrhythmia.

Undoubtedly, with still further improvement in preoperative preparation and surgical techniques, there will be a still greater percentage of clinical cure, complete or partial. In a series of cases of chronic constrictive pericarditis seen at the Massachusetts General Hospital, a recent report has given the following figures: In the period 1914 through May 1947, 53 patients with constrictive pericarditis had been seen. Surgical exploration with pericardiolysis and partial pericardectomy was undertaken on 42 of these patients. The results in 25 (60.9 per cent) have been excellent, good, or satisfactory. Six patients died as a result of the operative procedure itself, 5 died from complicating disease, and 4 died from the effects of their underlying disease. The cause of death of one patient is not known. Seven patients were too ill for surgery and died while receiving medical treatment. One patient died before the operative series began, 2 patients had such minimal symptoms that operation was felt to be unnecessary, and one patient has not been followed.
Differential Diagnosis

There are three conditions with which chronic constrictive pericarditis has been readily confused, although careful study should reduce very much such confusion in the future. These three conditions include: (1) cirrhosis of the liver in which, although there may be ascites, the venous pressure is normal and the neck veins are not engorged; (2) heart failure in which there is, as a rule, obvious disease of the heart itself by rheumatic involvement, hypertension, or coronary atherosclerosis; (3) myocarditis with failure of unknown cause in which there is, as a rule, much more cardiac enlargement, no evidence of pericarditis per se, and a different electrocardiographic pattern.

References

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Circulation. 1951;4:288-294
doi: 10.1161/01.CIR.4.2.288

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