ANNULAR CONstrictive Pericarditis

With an Account of a Patient with Functional Pulmonary, Mitral, and Aortic Stenosis

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Five patients with constrictive pericarditis are described, in whom the main constriction was found at operation to be concentrated in an annular band forming a hoop around the pericardial sac, at the level of the atrio-ventricular grooves. In four of them, although the constriction was predominantly at this annulus, the effect on the heart was similar to that resulting from more generalized constriction. In the fifth, signs of functional pulmonary, mitral, and aortic stenosis developed seven years after the original operation for constrictive pericarditis, due to a double annular constriction, encircling the heart at the atrio-ventricular groove and also the root of the aorta and pulmonary artery.

The five patients were under the care of Dr. William Evans and of Mr. Vernon Thompson, who operated upon them. Pre-operative investigation included postero-anterior and lateral radiograms, an electrocardiogram, and a phonocardiogram. In addition, cardiac catheterization was carried out before operation in two and in one it was repeated after operation.

Clinical Features

The patients were all male and their ages, at the time of operation, were 13, 46, 47, 54, and 66 years respectively. The boy, aged 13 years, who developed functional pulmonary, mitral, and aortic stenosis, will be discussed separately.

In two of these four, aged 46 and 54, the diagnosis was made on finding a calcified pericardium in the radiogram, the symptoms being effort dyspnea only, without edema. In addition, the older patient had a definite history of pericarditis at the age of 14 years; the younger had had a long and serious illness at the age of 17, which may have been complicated by pericarditis. In neither was there a family history of tuberculosis.

In spite of the absence of edema, both patients had a raised venous pressure. This measured 10 cm. above the sternal angle in the older and 5 cm. in the younger, when lying at 45° to the horizontal. The liver was enlarged one to two finger-breadths in both. In the jugular venous pulse, there was a clear early diastolic dip, and, on examination of the precordium, a diastolic rapid inflow beat was noted, accompanied by an early diastolic sound (Fig. 1) (Potain, 1856; Mounsey, 1955 and 1957). The radiogram showed annular calcification of the pericardium around the middle of the pericardial sac, the plane of the ring lying sagitally, the posterior portion being slightly to the left and the anterior slightly to the right of the mid-line (Fig. 2). In both patients pulsation of the apex of the heart appeared relatively free on screening. The general heart size was normal in the younger patient and the electrocardiogram showed low voltage T waves in

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FIG. 1.—Sounds and pulsations recorded over the mid-præcordium in a patient with annular constrictive pericarditis. The large diastolic rapid inflow (DRI) beat is replaced by a smaller one after successful operation: the early diastolic sound (EDS) is replaced by a third heart sound (3).

FIG. 2.—Right lateral, postero-anterior, and left oblique radiograms, showing annular pericardial calcification.
all leads with bifid P waves (Fig. 3). In the older patient the heart was enlarged and atrial fibrillation was present with right bundle-branch block. In addition, he had signs of associated aortic regurgitation with left ventricular enlargement.

The diagnosis was made on the history and symptoms in the other two patients, there being no certain evidence of pericardial calcification. Both had had considerable oedema and effort dyspnoea. The older, aged 66 years, had an attack of chest pain six years before, initially diagnosed as cardiac infarction, but later ascribed to pericarditis. The younger, aged 47 years, had developed generalized oedema seven years previously, without obvious cause; this subsided after a few months, only to recur four years later, when pleural effusions also developed. A diagnosis was made of constrictive pericarditis following an acute pericardial effusion four years previously. There was no past or family history of tuberculosis in either patient.

The signs of constriction were more marked in these two patients than in the first two described. The jugular venous pressure was raised to the level of the ears and the liver enlarged three to four finger-breadths below the costal margin. Both had a clear early diastolic dip in the jugular venous pulse, a diastolic rapid inflow beat, and an early diastolic sound. Radiograms showed slight generalized cardiac enlargement with active pulsation of the heart in both patients on screening. In the younger, this free pulsation was thought initially to weigh against the diagnosis of constrictive pericarditis. Both patients had chronic pleural effusions. In the older there was a small speck of calcium on the upper left border of the heart, suggestive, although not diagnostic, of pericardial calcification. The electrocardiograms were compatible with constrictive pericarditis, showing generalized low voltage T waves and atrial fibrillation. Cardiac catheterization was carried out in the older patient and revealed a pulse wave form in the right heart compatible with constrictive pericarditis. The mean right atrial pressure was 20 mm. Hg, with an early diastolic dip, the right ventricular systolic pressure 50 mm. Hg, and the cardiac output four litres a minute.

**FINDINGS AT OPERATION**

In all four patients a ring of greatly thickened pericardium was found around the middle of the pericardial sac. It passed across the base of the heart over the conus of the right ventricle, about 2.5 cm. below the pulmonary valve. It was attached laterally over both atrial appendages and then passed down into both atrioventricular grooves. It was not possible to trace its continuation posteriorly at operation. The ring was calcified in two of the patients, cartilaginous in one, and fibrous in the fourth.

The apical portion of the pericardium and that over the body of the ventricles showed evidence of old inflammation in all patients; it was moderately adherent and thickened in the two with a calcified ring, but was not adherent and hardly thickened in the other two. Inspection of the ventricular movement showed vigorous early diastolic filling, suddenly halted and abruptly checked by the restricting bands of thickened pericardium.

When the constricting ring was cut, the two divided ends sprang apart, and this was particularly striking in the two patients, whose ring was calcified. In one, the edges separated by as
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much as an inch, revealing a ridged furrow in the heart muscle beneath, with post-stenotic dilatation of the right ventricular outflow tract on the distal side of the ring, and proximal to the pulmonary valve. The heart beat appeared to improve in all four patients, as soon as the ring was divided. In two, no further pericardial stripping was thought necessary, while in the remaining two the thickened adherent pericardium was stripped from the ventricles, with further improvement of the heart's action.

Pericardial biopsy in every case showed evidence of chronic inflammation, but no clear signs of tuberculosis.

POST-OPERATIVE RESULTS

All four patients have improved following operation. The two patients presenting with calcification of the pericardium and effort dyspnoea, but no oedema, have now been followed for two years. In the 46-year-old man, with sinus rhythm and with a normal sized heart, the jugular venous pressure fell from 5 cm. above the sternal angle to near normal after operation: he is now free from symptoms. In the 54-year-old man, similar improvement in physical signs followed operation, but not to the same degree, the venous pressure falling from 10 to 3 cm. above the sternal angle: in this patient the signs of aortic regurgitation persisted.

Of the two patients without pericardial calcification, the younger, 47-year-old, has now been followed for ten years since operation and remains healthy. The 66-year-old patient has been considerably improved by operation; the jugular venous pressure has fallen from 12 to 5 cm. above the sternal angle and the diastolic rapid inflow beat and early diastolic sound have become smaller and later in time (Fig. 1). It is too early yet, however, to assess the success of operation.

FUNCTIONAL PULMONARY, MITRAL, AND AORTIC STENOSIS

The fifth patient differed from the other four, for at the time that the constricting annulus developed in his pericardium, he had already had a partial resection of his pericardium for constrictive pericarditis.

There was a strong family history of tuberculosis and, when the patient was three years old, the father died of open pulmonary tuberculosis. At the age of four, the child developed a tuberculous pericardial effusion, fluid from which gave a positive guinea-pig test. A year later he had a bilateral mastoid operation for chronic tuberculous otitis media. Two years after the original pericardial effusion, he had developed signs of established constriction, the jugular venous pressure being raised to the level of the ears and the liver enlarged to three to four finger-breadths below the costal margin. The radiogram showed considerable calcification over the posterior portion of the heart.

At the first operation the pericardium was cleared from the ventricles, as far as the atrio-ventricular groove. The visceral pericardium appeared raw red in colour, suggesting inflammation. The pericardium over the back of the heart and in the atrio-ventricular groove was thickened and calcified, and, on incising it, caseating matter was found, but no tubercle bacilli were seen in the specimen examined. No attempt was made to remove this portion of the pericardium, the ventricles having been satisfactorily freed.

The immediate result following this first operation was good and the patient eventually went to an open air school. During the next nine years, when he continued to attend the outpatient department at regular intervals, signs of inflow and outflow tract obstruction gradually developed. Before operation, no murmurs were heard, but within three months of operation, soft atrial systolic and mid-diastolic murmurs appeared at the apex, which gradually grew louder over the next five years (Fig. 4). These signs suggested mitral stenosis, but in view of the history of tuberculous pericarditis with residual pericardial thickening in the atrio-ventricular grooves, it was thought possible that the stenosis resulted from external compression of the thickened inflamed pericardium, rather than from a rheumatic process.

After five years the patient began to complain of increasing breathlessness and ankle oedema.
Fig. 4.—Serial phonocardiograms of patient R.L. from the mitral area (MA): 6.1.50, before operation: 11.12.50 and 25.2.53, after operation, showing the development of a mid-diastolic murmur (MDM) and an atrial systolic murmur (ASM). SM=systolic murmur; EDS=early diastolic sound; 1, 2, and 3=first, second, and third heart sounds; HF and LF=high and low frequency filters.

Fig. 5.—Serial phonocardiograms of patient R.L. (continued from Fig. 4): 25.2.53, showing loud murmurs (ASM, SM, and MDM) in the mitral area (MA), and in addition a basal systolic murmur (SM) with widely split second sound (2xx): 24.7.57, where the basal systolic murmur has increased in intensity: 30.10.57, after the second operation, showing great diminution in both mitral and basal murmurs.
It was now noted that he was developing signs of pulmonary stenosis, in addition to those of mitral stenosis, and since this lesion is hardly ever acquired as a result of rheumatic infection, it was thought likely that this also was due to external compression by pericardial bands. An alternative diagnosis was an argentafinoma, but the urinary 5-hydroxy-tryptamine test was within normal limits and excluded this diagnosis.

On examination, the jugular venous pressure was found to be raised to a mean level of 7 cm. above the sternal angle, with large a and c waves: the liver was enlarged two finger-breadths below the costal margin and pulsated. On auscultation, there was a loud basal systolic murmur, of rhomboid shape in the phonocardiogram, with wide splitting of the second heart sound (Fig. 5). The apical systolic and mid-diastolic murmurs were unchanged. Screening showed constriction of the pulmonary outflow tract, with concertina-like motion of the ventricles and see-saw movement of the heart about the point of constriction. The radiograms showed ballooning of the ventricles, particularly the right, beneath the point of constriction, and this was most apparent in the lateral views (Fig. 6). Residual calcification was noted over the back of the pericardium. The electrocardiogram was compatible with the diagnosis of acquired functional pulmonary stenosis, suggesting development of right ventricular preponderance, which had not been present previously (Fig. 7). In addition, besides the evidence of old pericarditis and inverted T waves in leads II, III, CR1 and CR4, it revealed a most unusual electrical position of the heart, thought to be due to herniation of the ventricles through the constricting pericardial annulus.

The patient became increasingly breathless on exertion and two years later, seven years after the original operation, developed right heart failure with functional tricuspid regurgitation. The systolic murmur at the base of the heart was now clearly conducted up into the neck and it was thought possible that he might also be developing functional aortic stenosis, in addition to the other functional valve lesions.

The patient was brought into hospital for investigation and for a second operation on the pericardium. Cardiac catheterization confirmed the diagnosis of functional pulmonary stenosis,
the right ventricular pressure being 125/5 mm. Hg and the pulmonary arterial pressure 25/12 mm. Hg. It was not possible to obtain a pulmonary capillary trace to confirm the presence of functional mitral stenosis. The right atrial pressure curve was compatible with functional tricuspid regurgitation.

A second operation was carried out, when the clinical diagnosis was confirmed. The ventricles bulged forward and to the left like a balloon and there was a tight constriction around both the aorta and the pulmonary artery at their origins (Fig. 8). Bands of thickened pericardium connected this constricting ring around the outflow tracts to the atrio-ventricular grooves on either side, and the mitral valve area was compressed and drawn upwards toward the outflow tracts. The aorta, pulmonary artery, atrio-ventricular grooves, and the mitral area were freed by careful dissection. Although the heart then assumed a more normal shape and relationship to the outflow tracts, a rather acute angle remained between the conus area of the right ventricle and the pulmonary artery.

After the second operation the patient was again examined by cardiac catheterization and it was found that the right ventricular systolic pressure had fallen from 125 mm. to 55 mm. Hg. It was not possible to enter the pulmonary artery at this investigation.

The patient is now restored to normal health, the signs of right heart failure having cleared. The basal systolic murmur and the systolic and diastolic murmurs in the mitral area persist, but are much softer (Fig. 5). Screening shows active cardiac pulsation as before, but no great constriction in the region of the outflow tract.
FIG. 8.—Sketch of film of patient R.L. at second operation, showing pericardial constriction, (1) in A-V groove, and (2) around base of aorta and pulmonary artery. Note ballooning of right ventricle below constriction.

**DISCUSSION**

In all five patients the site of the annular pericardial thickening was similar, the main ring of thickening encircling the base of the ventricles at their junction with the atria and the pulmonary outflow tract. In only one did it also encircle the two outflow tracts, passing behind the aorta and pulmonary artery through the transverse sinus.

Friedberg (1949) states that pericardial calcification in constrictive pericarditis is commonest in the coronary sulcus, over the diaphragmatic surface of the right ventricle, over the right atrium, and over the sternal surface of the right ventricle in the order mentioned. Evans and Jackson (1952) stress the finding of calcium over the left border of the heart and note that calcification sometimes appears as a cup holding the heart in the left oblique view, where an incomplete ring is seen. They mention that occasionally a completed ring of calcium is seen resulting from calcification in the atrio-ventricular groove. Plum et al. (1957) describe a similar distribution of calcification in constrictive pericarditis, emphasizing the cup or J shaped appearance, due to the continuation of the calcification in the coronary sulcus, lying between the left atrium and the left ventricle, with that on the diaphragmatic surface of the right ventricle.

Friedberg suggests that calcification occurs most frequently in these areas, since they are the zones where least movement of the heart takes place. Observations of the movement of the heart, however, during operations with the pericardium open, suggest that the reverse may be true. Although these are the areas of the cardiac silhouette that show least movement on screening of the heart, they are probably the areas of greatest friction between the visceral and parietal layers of
annular constrictive pericarditis

...When the heart is watched at operation, its action appears in some ways similar to a force-pump, the atrio-ventricular and semilunar valve rings moving toward the apex during systole and away from it during diastole. During systole the semilunar valve rings are drawn downward toward the ventricular cavities, the ascending aorta and pulmonary artery lengthening and the ventricles shortening. In diastole, the semilunar valve rings rise again, as blood flows forward out of the ascending aorta and main pulmonary artery. Similarly the movements of the atrio-ventricular rings are co-ordinated with the forward movement of the blood from atria to ventricles: the rings move backward toward the atrial cavities in diastole, when the atria are emptying into the ventricles, but forward into the ventricular cavities during ventricular systole, when the blood is being squeezed out of the ventricles. The areas of the exterior of the heart overlying the four valve rings are, for this reason, those at which the greatest movement takes place. In the normal heart, therefore, movement of the visceral upon the parietal layers of pericardium might be expected to be greatest in these areas. Hence in the presence of pericarditis, friction of the inflamed surfaces of the visceral and parietal pericardium will be greatest here also, thus giving rise to a more intense inflammatory reaction.

In the patients described in this paper, the predilection for increased inflammatory reaction, with subsequent fibrosis and calcification, around the base of the ventricles was unusually great, with the result that a ring of rigid pericardium completely surrounded the heart at this level. Although the pericardium of the more distal portions of the ventricles was relatively little involved in two of the patients and only to a lesser degree in another two, generalized effects of constrictive pericarditis resulted. This is probably explained by the fact that for free, normal filling to take place, free movement of the atrio-ventricular valve rings must be possible, since these movements are an essential part of the smooth force-pump action of the heart.

 Patients with annular constrictive pericarditis may prove difficult to diagnose, especially in the absence of calcification, if too much emphasis is placed on the presence of a still heart on screening. In fact, many patients with constrictive pericarditis have very active pulsation during systole and early diastole. McKusick (1952) has shown that in constrictive pericarditis the electrokymogram shows a distinctive pattern, which is of a V shape followed by a plateau. The downward stroke of the V is due to ventricular contraction in systole, the upward stroke to sudden early ventricular filling in diastole, after which, no further filling being possible as a result of the pericardial constriction, a plateau is inscribed during the remainder of diastole. This abnormal form of cardiac pulsation is especially well seen in patients with localized constrictive pericarditis and especially in three of the patients in this series. For the same reason all five patients had a striking diastolic rapid inflow beat and also a loud early diastolic sound, which are the clinical counterparts in the chest examination of the early abrupt arrest of ventricular filling in constrictive pericarditis.

 The fifth patient differed from the other four in that the residual thickened pericardium, after the body of the ventricles had been stripped at a first operation, caused obstruction to ventricular ejection and also inflow obstruction to ventricular filling. In this patient the main constricting annulus was in the same place as in the other patients, but this was connected to a second smaller ring, passing through the transverse sinus of the pericardium and encircling both the aorta and the pulmonary artery. It is likely that the successful freeing of the body of the two ventricles at the first operation tended to increase the relative constriction in the region of the pulmonary, aortic, and mitral valves.

 Functional mitral stenosis resulting from constrictive pericarditis was described by Paul et al. (1948). They reported a patient in whom a mid-diastolic murmur was present at the apex, and at autopsy slight mitral stenosis was found to have been caused by penetration of the calcified inflammatory process into the left ventricle, with impingement upon the mitral valve. Eliasch et al. (1950) described a similar case. It seems almost certain that the mid-diastolic murmur in the patient in this series was due to functional mitral stenosis from invasion of the mitral valve area by the fibrosing and calcifying process in the pericardium, since dense pericardial thickening was found in this area at the second operation.
I have been unable to trace a report of another patient developing functional pulmonary and aortic stenosis as a result of constrictive pericarditis. The rarity of this syndrome probably indicates that the transverse sinus is infrequently involved to any extent in the fibrosis consequent on pericarditis and so the secondary ring around the root of the great vessels does not form to constrict the aorta and the pulmonary artery.

The excellent result following the second operation in this patient, when the functional stenosis at the mitral, pulmonary, and aortic areas was relieved by stripping off the thickened pericardium in these areas, emphasizes the importance of careful examination and full dissection of these sites in every operation for constrictive pericarditis.

**Summary**

Five patients with constrictive pericarditis are described, in whom the main constriction was found at operation to be concentrated in an annular band forming a hoop around the middle of the pericardial sac. This band was related to the atrio-ventricular grooves and traversed the conus area of the right ventricle anteriorly.

Symptoms of generalized constrictive pericarditis resulted in four of the patients, although the rest of the pericardium was relatively little involved. Removal of the band alone in two patients, and of more extensive areas of pericardium also in the other two, relieved the symptoms of constriction.

It is suggested that the atrio-ventricular grooves and the conus region of the ventricles are those areas where most movement of the heart takes place and hence the areas where pericardial friction is greatest: as a result dense thickening might also be expected in these sites in the presence of pericarditis. Free movement of the atrio-ventricular rings and the conus area of the ventricles is necessary for the normal free pumping action of the heart and tethering of these areas is sufficient to produce symptoms of constrictive pericarditis.

The fifth patient developed signs of functional pulmonary, mitral, and aortic stenosis from external compression of the heart by two bands of thickened pericardium, which had been left at the first operation for constrictive pericarditis. These were divided at a second operation with relief of the functional stenosis. The importance of inspection and, if necessary, full dissection of the atrio-ventricular grooves, the conus area, and the transverse sinus of the pericardium in operations for constrictive pericarditis is stressed, in order to avoid the possibility of this serious complication.

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