Mitral regurgitation in congestive cardiomyopathy

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CARDIOMYOPATHIES have been defined as 'A sub-acute or chronic disorder of heart muscle of unknown or obscure aetiology often with associated endocardial or pericardial involvement but not due to occlusive coronary atherosclerosis' (Goodwin *et al.*, 1961; Goodwin, 1966).

Primary cardiomyopathy is a cardiomyopathy without generalized disease elsewhere in the body. This group includes cryptogenic congestive cardiomyopathy, endomyocardial fibrosis, endocardial fibroelastosis and the cardiomyopathies of pregnancy, the puerperium and alcohol. Löeffler's eosinophilic cardiomyopathy (Löeffler, 1936), Fiedler's 'myocarditis' (Fiedler, 1899), giant cell 'myocarditis' and Becker's cardiomyopathy (Becker, Chatgidakis & Van Lingen, 1953) also come into this group. Finally, there is hypertrophic obstructive cardiomyopathy, otherwise termed idiopathic hypertrophic sub-aortic stenosis (Goodwin *et al.*, 1960; Morrow & Braunwald, 1959; Braunwald *et al.*, 1960; Cohen *et al.*, 1964).

Secondary cardiomyopathy is a cardiomyopathy resulting from, or associated with, a generalized process involving other organs.

Cardiomyopathies commonly present clinically in one of three ways: with cardiomegaly and often congestive heart failure (congestive cardiomyopathy); with a syndrome suggesting cardiac restriction and mimicking constrictive pericarditis (constrictive or restrictive cardiomyopathy) or with gross hypertrophy of the heart, abnormal contractile function and often systolic gradients across the outflow tracts of the ventricles and signs suggesting outflow tract obstruction (hypertrophic obstructive cardiomyopathy) (Goodwin *et al.*, 1961).

Abnormal mitral valve function is common to many of these conditions and this paper describes some of the abnormalities of the mitral valve seen in association with the congestive group of cardiomyopathies. Mitral regurgitation is also an important factor in the other types of cardiomyopathy and a fuller report including these types will be published subsequently.

Normal mitral valve function

Closure of the mitral valve is accomplished by the interposition of the anterior and posterior leaflets lying between the left ventricle and the left atrium. Efficient closure is dependent upon the chordae and papillary muscles allowing sufficient latitude for the leaflets to come into apposition and also to supply a check to them in the ideal position. An adequate amount of cusp tissue must be present to provide continuity and there must be freedom of commissural movement (Levy & Edwards, 1962).

The papillary muscles tense and seal the mitral valve cusps in apposition during isovolumetric contraction of the left ventricle. During ejection the size of the cavity is reduced by the constrictor muscles of the ventricle and the long axis is shortened by contraction of the spiral muscles. Thus full or almost complete emptying is obtained without mitral reflux (Rushmer, 1961). The position of the papillary muscles relative to the inflow and outflow tracts of the left ventricle is shown in Fig. 1.

FIG. 1. Diagram of the left ventricle to show the position of the inflow and outflow tract and of the papillary muscle of the mitral valve. LV = left ventricle; RV = right ventricle; TV = tricuspid valve; LA = left atrium; RA = right atrium. (Arrows show direction taken by blood flowing from left atrium to left ventricle.)

Disordered mitral valve function

Mitral regurgitation may be due to loss of integrity of the papillary muscle from rupture or damage from other causes, disease of the chordae which fail to exercise the correct amount of restraint to the cusps, dilatation of the valve ring secondary to left ventricular enlargement, or destruction of the valve cusps or congenital abnormalities in them. Finally, fusion of the commissures is common in many patients with rheumatic mitral valve disease who have a rigid narrowed valve which is also regurgitant.

Types of mitral regurgitation

Mitral regurgitation may be divided into *valvar*, *annular* and *sub-valvar* types.

Valvar mitral regurgitation is nearly always rheumatic or congenital in origin and will not further be dealt with here.

Annular mitral regurgitation may be used to describe mitral regurgitation due to dilatation of the valve ring in association with enlargement of the left ventricle and ventricular cavity, usually as a result of heart failure. It is often transitory although it may become established and is common in congestive cardiomyopathy with left ventricular failure.

Patients with congestive cardiomyopathy have a serious derangement of the ventricular muscle and impairment of contractility. The disease often presents with an episode of left ventricular failure, sometimes following an upper respiratory tract infection. Left ventricular failure commonly proceeds relentlessly to right ventricular failure and the condition may be complicated by pulmonary or systemic emboli. Many of the patients are young adults who have a low cardiac output, cool extremities, peripheral cyanosis and small pulse volume. The jugular venous pressure is commonly raised and when right ventricular failure has occurred tricuspid regurgitation is common. The heart is considerably enlarged, the cardiac impulse diffuse and the left ventricular thrust of poor quality. A gallop rhythm is the rule, usually arising from the left ventricle and due to a third heart sound (Goodwin, 1964). Mitral regurgitation is indicated by the presence of a pansystolic murmur at the cardiac apex conducted to the axilla and followed by a third heart sound. The short decrescendo mid-diastolic murmur which is so common in severe rheumatic mitral regurgitation is almost invariably absent.

Cardiac catheterization commonly reveals a modest degree of pulmonary hypertension, or normal right ventricular and pulmonary artery pressures. Some patients develop severe pul-

monary hypertension, but this is not the rule. When right ventricular failure has become established, there is frequently a high end-diastolic pressure in the ventricles, introducing a restrictive factor which tends to mimic constrictive pericarditis (Figs. 2 and 3). In such circumstances the ventricles have dilated against a tight pericardium, so that some element of constriction as well as restriction may exist. The left ventricular end-diastolic pressure is also raised,

FIG. 2. Right atrial and right ventricular pressure pulses in a patient aged 27 with congestive cardiomyopathy with restrictive features. The right atrial pulse (RA) shows large 'a' and 'v' waves, and the end-diastolic pressure in the right ventricle (RV) is elevated. Note that the 'a' wave is transmitted to the right ventricle. The early diastolic pressure in the ventricle is above zero, distinguishing the disorder from constrictive pericarditis in which the early diastolic pressure falls below.

FIG. 3. Left atrial, left ventricular and aortic pressure pulses in the same patient as in Fig. 2 with congestive cardiomyopathy showing the high left atrial pressure due to the high end-diastolic pressure in the left ventricle. There is no systolic gradient between left ventricle and aorta (pressures in mmHg).

which with the low systolic pressure gives a lower pulse pressure than normal (Fig. 3).

Left ventricular angiocardigraphy by the trans-septal route with injection into the left ventricle shows a dilated ventricular cavity with little muscular hypertrophy and poor contractile function manifested by a small ejection fraction and a large residual fraction (Raphael, 1966; Goodwin, 1967; Oakley *et al.*, 1968). Stroke volume is commonly low and minute volume also reduced.

Absolute evidence of mitral regurgitation is difficult to obtain for we do not usually perform left ventricular angiography by the retrograde route because of the irritability of the chamber, which often makes studies of ventricular volumes impossible because of frequent ventricular ectopic beats. Furthermore, ectopic contractions tend to produce false mitral regurgitation. The presence of mitral regurgitation in these patients may be strongly inferred from the character of the systolic murmur and the appearance of the left atrial pressure pulse, with tall early 'v' waves typical of mitral regurgitation, although they may occur in left ventricular failure (Fig. 2).

The variable nature of the systolic murmur, which tends to wane with clinical improvement in left ventricular failure and wax with deterioration is also consistent with mitral regurgitation due to stretching of the valve ring in a dilated left ventricle. Furthermore, necropsy studies show a normal mitral valve and subvalvar apparatus with some increase in circumference of the mitral valve ring, although this is often not striking. Dilatation of the tricuspid valve ring may be more impressive in right ventricular congestive failure.

It seems improbable that annular mitral regurgitation in congestive cardiomyopathy exerts an important effect upon the course of the disease and it should be regarded as part of the pattern of severe left ventricular failure and myocardial disturbance rather than a fundamental part of the disease. Its presence, however, is likely to aggravate the left ventricular failure. Mitral regurgitation occurs only in patients with considerable cardiac enlargement and is usually absent in constrictive cardiomyopathy such as primary amyloid disease, in which the heart is not usually greatly enlarged. Diastolic excursion is restricted, which tends to limit the dilatation of the valve ring unless there is extensive endocardial involvement.

Sub-valvar mitral regurgitation

My colleagues and I (Raftery, Oakley & Goodwin, 1966) coined the term *sub-valvar mitral*

incompetence (regurgitation) to describe lesions of the mitral valve chordae and papillary muscles. The papillary muscles may be involved by infarction due to ischaemic heart disease as described by Burch, de Pasquale & Phillips (1963). Papillary muscle infarction or insufficiency can also occur in severe aortic valve stenosis (Moller, Nakib & Edwards, 1966), in anomalous coronary arterial supply (Noren *et al.*, 1964) and in discrete sub-valvar aortic stenosis (Goodwin, 1967). Chordal rupture may occur spontaneously or in bacterial endocarditis.

Sub-valvar mitral regurgitation is an important feature in endomyocardial fibrosis and in hypertrophic obstructive cardiomyopathy. These lesions, however, will not be dealt with in this paper.

In sub-valvar mitral regurgitation the mitral valve leaflets fail to maintain apposition during ventricular systole because of failure of the papillary muscles or chordal function. The reflux steadily increases throughout systole as the valve leaflets prolapse back into the left atrium (Sleeper, Orgain & McIntosh, 1962; Burch *et al.*, 1963; Raftery *et al.*, 1966). The normal mobility of the valve allows the posterior cusp to prolapse back into the left atrium and to direct a stream of blood over the anterior leaflet and against the base of the aorta (Sleeper *et al.*, 1962; Raftery *et al.*, 1966).

The papillary muscles may be infiltrated or damaged in secondary cardiomyopathy as in two of our patients, one of whom had dermatomyositis and the other sarcoidosis. The latter patient, who has previously been described by Raftery *et al.* (1966), presented symptoms and signs closely similar to those produced by papillary muscle infarction and it is likely that the mechanism was also similar. Thus acute dyspnoea due to pulmonary oedema occurred in the presence of only moderate cardiac enlargement and there was an ejection systolic murmur heard at the apex and radiating to the base of the heart. The ejection quality and distribution of the murmur have been ascribed by Sleeper *et al.* (1962) and by Burch *et al.* (1963) to progressive regurgitation during systole and direction of the stream of blood over the anterior cusp of the mitral valve against the base of the aorta as the posterior leaflet prolapses back into the left atrium. Under these circumstances, the left atrial 'v' wave may assume very considerable proportions. The peak of the left atrial pressure pulse occupies the same position in the cardiac cycle as the normal 'v' wave (Fig. 4), rather than occurring earlier in systole as with valvar and

annular mitral regurgitation, when the reflux tends to occur throughout the whole of systole.

The presence of persistent pulmonary oedema with only moderate cardiac enlargement reflects the mechanical nature of the mitral regurgitation. Extreme pressure is generated in the normal-sized left atrium leading to very high pulmonary venous pressure as a result of poor compliance of the atrium. In our patient with myocardial sarcoidosis, the remainder of the myocardium was presumably only slightly involved in view of the absence of conduction defects and the striking response to mitral valve replacement. At operation the valve and papillary muscles were excised, and replaced by a Melrose prosthesis (Melrose *et al.*, 1964). The patient has made a striking improvement since the operation and is on treatment with steroids.

FIG. 4. Indirect left atrial pressure (PC) and left ventricular (LV) pressures in a patient (V.W., age 25) with papillary muscle insufficiency due to sarcoidosis showing the tall 'v' wave with peak in mid-systole in the left atrial pulse (electrocardiogram below, pressures in mmHg).

This result of infiltration of the heart must be rare and, to my knowledge, has not previously been reported. It illustrates well one of the mechanisms by which the mitral valve may become regurgitant in congestive cardiomyopathy. This patient also underlines the important principle that when persistent pulmonary oedema and signs of mitral regurgitation are associated with but modest left ventricular enlargement, a mechanical cause should be suspected and the possibility of operative treatment considered.

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