FAILURE OF THE RIGHT VENTRICLE

A CASE REPORT

BY

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Within recent years the belief that one ventricle may fail without coincident failure of the other has been steadily gaining ground against the older hypothesis of Mackenzie that the heart must always fail as a whole. Much recent work, in America and elsewhere, recently reviewed by Bedford (1939), has shown that hypertension and aortic valvular lesions can occasion a failure of the left ventricle with little or no failure of the right ventricle. The finding of pulmonary congestion and delay in the arm-to-tongue circulation time in patients showing a dilated failing heart in the absence of a rise of venous pressure is convincing support for this theory.

Although separate failure of the right ventricle is theoretically possible, it has been discussed less and its application to the general problems of cardiology has often been too little emphasized. McGinn and White (1935) and White (1935) have shown that an acute failure of the right heart may be rapidly produced by pulmonary embolism. A dilated pulsating conus and gallop rhythm localized to the left second and third interspaces, with a rise in venous pressure, speak eloquently for a failure and dilatation of the right ventricle; the left remaining normal. In the reports on the "cor pulmonale" there are many instances of chronic enlargement and failure of the right heart in the presence of a normal left ventricle. Rogers (1908–1909) and Clarke *et al.* (1927) report cases in which the right ventricle was larger than the left, which was itself normal.

The following case is presented as one that falls into this group and shows that the right ventricle may fail for a long time without any appreciable failure of the left side.

CLINICAL COURSE

A woman of 73 was quite well until four years before her death, when she noticed increasing dyspnoea on exertion and swelling of the legs and abdomen. At her first admission to Addenbrooke's Hospital she was cyanosed and showed gross œdema of the legs and sacral area, and a large abdomen which was distended with ascitic fluid. The cervical veins were engorged and the liver

was enlarged to three finger-breadths below the costal margin. Her heart, at this time, was not greatly enlarged, but there was some dullness to the right of the sternum. The pulmonary second sound was loud and slapping and there was a long, harsh systolic murmur in the mitral area. The rhythm was regular.

She was re-admitted on nine occasions, on each of which the abdomen was tapped, sometimes several times. In between she was well enough to do a little light housework, such as sewing, peeling potatoes, or cooking. Although short of breath on slight exertion, the relative absence of dyspnœa in comparison with the gross congestive failure was very striking ; on one occasion she was able to walk into the ward with three gallons of fluid in her abdomen. Although she preferred the sitting posture, she was not in a true sense orthopnœic, and, except when the ascites was extreme, was able to lie flat without dyspnœa. During the whole time of observation, a period of some two years, she was deeply cyanosed and showed gross engorgement of the cervical veins. Although under continual treatment with mercurial diuretics, her œdema and ascites persisted, and the ascites was only temporarily relieved by tapping of the abdomen.

In the past she had been healthy and had never had rheumatic fever or chorea.

On examination on many occasions during the last two years of her life she showed the following signs.

She was emaciated and cyanosed, with many telangiectatic venules over the face, which was not unlike that of mitral stenosis. The cervical veins were engorged up to the level of the angles of the jaw and pulsated feebly ; they stood out as thick, knotted cords almost as thick as the little finger. Œdema and ascites were always present.

The pulse was regular and of small volume : the blood pressure, 110/85 mm. On her first admission the left border of the heart seemed normal and there was some dullness to the right of the sternum. Later the apex moved out a little, but the dullness to the right of the sternum disappeared. The heart was never greatly enlarged to clinical examination. In the mitral area there was a loud, rough systolic murmur ; the pulmonary second sound was increased ; the aortic sounds were normal ; and there was a systolic murmur in the tricuspid area. There was no pulsus paradoxus, no systolic retraction, no Broadbent's sign, nor any of the other signs attributable to an adherent pericardium.

Throughout her illness the chest appeared to be clear except for occasional rales at the bases. There was no pleural effusion.

On all her admissions the abdomen was tightly distended with fluid. The liver was greatly enlarged and was not tender to pressure apart from her first admission early in the disease ; it appeared to be hard and smooth. The spleen could not be felt. The nervous system showed no abnormality.

Radioscopy four months before her death showed some enlargement of all the chambers of the heart, most marked in the right ventricle and left auricle. Pulsation was rather feeble and pulmonary congestion was said to be slight.

The electrocardiogram showed no abnormality apart from right ventricular preponderance. The circulation time (arm to tongue with suprachol) was

35 seconds. The Kahn reaction was negative. The lævulose tolerance test showed moderate liver deficiency.

Throughout the four years of her illness there was neither deterioration nor improvement ; she died suddenly for no apparent reason just after admission to hospital for tapping.

In view of the constant engorgement of the neck veins and the anasarca, a clinical diagnosis of chronic right heart failure of unknown cause was made.

AUTOPSY

An autopsy showed acute failure supervening on chronic failure of a hypertrophied right heart, and emphysema of the lungs.

Heart.—This showed great hypertrophy of the right ventricle and right auricle. The left ventricle did not look hypertrophied. Before the fat was removed the total heart weight was 15 oz. (425 g.). Differential weighing after separation of the ventricles and removal of all fat (Herrmann and Wilson, method B, 1921–22) showed that the right ventricle weighed 107 g., the left 127 g. This gave a left to right ventricular ratio of 1·19. Lewis (1913–1914) gives the normal average L/R ratio as 1·8, the limits of variation lying between 1·47 and 2·06. Herrmann and Wilson consider anything below 1·50 or above 2·20 as abnormal.

The normal figures for the weights of left and right ventricles are shown in the table (after Lewis and Herrmann and Wilson). As both these authors weighed the septum separately in their series, the figures below have been obtained by adding one third of the septal weight to the weight of the right ventricle and two thirds to that of the left.

				Weight of Ventricles in Grammes	
				Right	Left
Average (Herrmann and Wilson)	..			51	91
Average (Lewis)	..	..	..	56	100
Range (Lewis)	..	..	..	36 to 80	74 to 141
Present case	..	..	..	107	127

It will be seen that while the left ventricle may, at the most, have been slightly hypertrophied, its weight fell within the range of normal figures.

The right ventricle was extremely hypertrophied and was responsible for most of the increase in heart weight. The relationship between the size of the two ventricles is well seen in the coronal section of the heart shown in the figure on the next page.

Microscopy of the myocardium showed that the fibres in the right ventricle were greatly thickened, while those in the left were normal. In spite of the patient's age, atheroma was inconspicuous in the coronary arteries and aorta. On the other hand, there was severe atheroma in the pulmonary artery and its branches. Microscopy of the pulmonary arterioles showed no abnormality and no obstruction. The valves and pericardium were normal.

FIG. 1.—Coronal section of the heart. Natural size. R: right ventricle. L: left ventricle. Diameter of coin is one inch.

Lungs.—These were small, generally congested, and very œdematous; but this was probably terminal. There were emphysematous bullæ along both anterior borders. Microscopically, the lungs showed a very fine, even emphysema, with œdema, congestion, and phagocytosis of blood pigment.

Abdominal Organs.—The liver showed fibrous thickening of the capsule, mainly over the antero-superior surface and the anterior border, with plaques of hyaline “sugar icing” which could be stripped from the underlying organ with moderate ease. Microscopically and macroscopically there was fibrous thickening both of the portal systems and of the tissues around the central veins. The latter areas showed congestion. The hepatic fibrosis, well marked in all regions, showed its greatest intensity just beneath the capsule. The larger hepatic veins were widely patent and of quite abnormal cross-section. All were surrounded by a rim of fibrous thickening.

The sugar icing was composed of hyaline fibrous tissue and lay outside the elastic fibres of the real capsule. It had evidently been laid down from outside. It is interesting that the changes in this liver were similar to those found in eight cases of constrictive pericarditis that came to autopsy (Day and Armstrong, 1940).

The peritoneal cavity contained several pints of clear yellowish-brown fluid. There was fibrous thickening with sugar icing all over the parietal peritoneum,

most marked anteriorly and less pronounced over the under-surface of the diaphragm. The spleen was congested and œdematous, with plaques of sugar icing on its surface. There was general fibrous thickening of the intestines, stomach, pancreas, and mesenteric and retro-peritoneal fatty tissues.

There was gross induration and œdema of the subcutaneous tissues. The remaining organs were normal.

DISCUSSION

The view that this patient suffered from a failure of the heart restricted to the right ventricle is based on a variety of clinical and pathological observations. In the first place, she suffered from right ventricular failure for at least two years, during which time personal observation revealed gross engorgement of the neck veins. Her relative lack of dyspnœa and orthopnœa, often in the face of gross ascites and œdema, is strong evidence against any material degree of pulmonary congestion caused by left ventricular failure.

The pathological findings present even stronger evidence. The left ventricle was normal in weight and in the thickness of its walls. Practically all the increase in weight of the heart was due to the right ventricle, which weighed nearly as much as the left. The muscle fibres of the right ventricle were greatly hypertrophied; those of the left ventricle were normal in thickness. The slight degree of atheroma in the thoracic aorta was in keeping with the clinical finding of a low systemic blood pressure; the conspicuous amount of atheroma in the larger pulmonary arteries suggested that the pulmonary arterial pressure must have been greatly above the normal. The chronic œdematous and fibrous changes in the lower part of the body and the hepatic congestion and fibrosis of "cardiac" type are proof that, in addition to compensating hypertrophy, there had also been a long-continued failure of the right side of the heart.

As in the cases reported by East (1940), the ultimate cause of the right ventricular failure remains obscure. There were no valvular lesions and no obstruction or narrowing of the pulmonary arterioles. The atheroma in the larger branches of the pulmonary circuit was evidently the result and not the cause of the hypertension. This is in accordance with the views of Rosenthal (1930), who states that primary sclerosis of the pulmonary arteries affects the arterioles most and the larger branches least; secondary atherosclerosis, as in mitral stenosis (and as in this case), affects principally the large branches and leaves the arterioles unimpaired. It is possible that the emphysema was the cause of the obstruction, but it is unlikely in view of the relative mildness of the dyspnœa during life. Moreover, according to Parkinson (1937), right heart failure, when caused by emphysema uncomplicated by cardiovascular disease, is present only in the terminal phase a short while before death. Persistent or recurrent heart failure in emphysema is nearly always the result of coincident cardiac disease. As the heart of this patient had been failing for at least two years, it is unlikely that emphysema was the cause.

It is impossible to state whether pulmonary hypertension was the cause or the result of the right ventricular hypertrophy. Perhaps the most likely

hypothesis is that a local rise of pressure occurred in the pulmonary arteries as a result of spasm or some other unknown factor that left no mark upon the final pathology of the lungs, and that such isolated pulmonary hypertension occasioned hypertrophy and later failure of the right ventricle.

A survey of the papers on pulmonary arteriosclerosis, *cor pulmonale*, and the condition known as "Ayerza's syndrome" leads one to the conclusion that in a proportion of these cases no pathological explanation has yet been found for the right ventricular failure. Many cases (Arrilaga, 1913 ; Ribierre and Geroux, 1921 ; Warthin, 1919 ; and Clarke *et al.*, 1927) failed to show any arteriolar obstruction that could impede the pulmonary blood flow. Atheroma and medial degeneration of the larger arteries will not obstruct the flow owing to the relatively large size of the cross-section of these vessels : it is only when the lumen is relatively small that projections from its wall will cause obstruction. These cases showed atheroma and dilatation of the pulmonary arteries with normal arterioles, and in Warthin's case (a syphilitic) the smallest pulmonary vessels and even the pulmonary veins were dilated. This observation would appear to exclude a primary obstruction in the vessels themselves. These cases are mentioned to emphasize that there are some on record that show right ventricular hypertrophy without any adequate pathological explanation ; the clinical picture is, of course, quite different, for this patient was in no sense a "black cardiac." In the other cases of *cor pulmonale* a true endarteritis of the smaller vessels often accompanied by thrombosis (Rosenthal *et al.*, 1930) constitutes a real obstruction to the pulmonary circuit and would be expected to produce hypertension and hypertrophy of the right chambers of the heart. In a proportion of cases of *cor pulmonale*, however, it would seem that the pulmonary atheroma, as in the present case, was the result rather than the cause of the condition ; the exact ætiology of the cardiac hypertrophy remaining unknown.

Whatever the cause, it appears that in the case here reported the right heart failed alone, or at least greatly in excess of the left. Many other examples of preponderating right heart failure, but with a different symptomatology, are to be found in the papers mentioned.

The interest in this case lies, in particular, in the chronicity of the disease and the long-continued failure of the heart with a rise of the venous pressure. It would seem that the absence of left ventricular failure and pulmonary congestion was the reason for the extreme chronicity, and that this was the deciding factor in prolonging life. In most cases the left ventricle precedes the right in failure, and when the right heart fails it is a more transitory condition, relieved by treatment unless the patient dies. A series of attacks of right heart failure is common, but in the intervals the right heart remains efficient and the venous pressure is not above normal. It is suggested that in the presence of a failing left ventricle, right ventricular failure is not compatible with life for more than a limited period of time. It is only when the burden of pulmonary congestion is absent, that is when the right ventricle fails alone or at least greatly in excess of the left, that congestive cardiac failure can continue almost indefinitely. This hypothesis is in keeping with the observation that many patients with rheumatic tricuspid

disease live for a quite disproportionate time if the severity of the valvular lesions and the degree of failure are taken into account. The extra valvular lesion protects the lungs from flooding and, in rationing the left ventricle with blood, wards off the failure of this chamber. The same syndrome of long-continued right heart failure with persistent ascites is seen occasionally when an enormous enlargement of the right heart has taken place ; I have seen several such cases, but no autopsy was obtainable and the exact anatomical diagnosis of the cardiac lesions remained in doubt. The same result is seen in constrictive pericarditis—perhaps the most chronic of all forms of congestive failure—although here the heart itself does not fail, but is prevented from doing its work by outside factors. It is, none the less, a failure of right-sided type without pulmonary congestion. In conformity with the above hypothesis there is an unremitting heart failure of long duration. The similarity between constrictive pericarditis and the case recorded here hardly needs emphasis.

CONCLUSION

It is suggested, therefore, that the duration of the failure and its unremitting nature in this case were due to the right ventricle failing alone. The unusual pathological findings, the cardiac “cirrhosis” of the liver, and the chronic indurative changes in the peritoneum, were probably an expression of the extreme chronicity of the disease, and are strictly comparable with the findings in constrictive pericarditis.

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