is not nearly so high as in cases of double uterus (uterus bicornis). In association with transverse lie premature rupture of the membranes, prolapse of the cord, premature labour, abnormal uterine action, and obstructed labour may occur. In all cases the placenta is more liable to be retained, atonic post-partum haemorrhage is fairly frequent, and lochia may be excessive or prolonged in the puerperium.

Double Uterine Cavity

In uterus simplex septus bicornis and uterus bicornis bicornis—unicollis or bicollis—the uterine cavity is completely divided at least as far as the internal cervical os. Two separate compartments which do not communicate with one another are formed and the uterus is therefore a true double uterus (Fig. 3). Loculation of

FIG. 3

the ovum results in a high rate of abortion and premature labour. There may be a high incidence of hyperemesis and toxæmia, although my personal findings on these points have been inconclusive. Bleeding may occur, especially during the early months of pregnancy, from the non-pregnant horn; but this does not occur in the majority of cases, and the amount of loss is usually small. Accidental ante-partum haemorrhage is apparently more frequent than in normal cases. The non-pregnant horn of the bicornuate uterus cleft externally to the level of the internal cervical os is liable to rotate into and become impacted in the pouch of Douglas, where it may subsequently cause obstructed labour. If the uterine cleft does not reach the internal os it will nearly always be raised during pregnancy, or occasionally during labour, above the level of the pelvic brim and will not then be apt to obstruct labour. The foetal lie is almost invariably longitudinal, but the incomplete breech commonly occupies the fusiform lower segment and constitutes the presenting part. Twin pregnancy is commoner in double than in single uteri.

The lower uterine segment may resist expansion in late pregnancy and during labour. This may lead to non-engagement of the presenting part in the pelvis and failure of approximation of the presenting part to the cervix. Incoordinate uterine action or hypotonic inertia with subsequent prolonged labour, and possibly arrest of progress and foetal and maternal distress, is particularly apt to arise. If labour is obstructed by the non-pregnant uterine horn uterine rupture may occur, especially through the scar of a previous caesarean section or the thin medial wall of a bicornuate uterus. A cervical or vaginal septum may obstruct labour or may, on being lacerated during the birth, bleed profusely. On occasion the presence of a recto-vesical septum may add to the

difficulties of caesarean section (Hunter, 1960). Retention of the placenta and atonic post-partum haemorrhage, which may be severe, often occur. On rare occasions, when manually removing a retained placenta from a septate uterus, the oedematous, bruised, and possibly lacerated uterine septum may be mistaken for a subendometrial fibromyoma. Loss of blood during the puerperium may be excessive and prolonged, and the decidua of the non-pregnant horn may be discharged piecemeal or as a complete cast towards the end of the first week after delivery.

Summary

Although in the majority of cases of uterine deformity pregnancy and labour are normal, the incidence of complications is much higher than would be expected if the uterus was normal. If the risks are known and anticipated, earlier recognition and prompt treatment will undoubtedly reduce the hazards of pregnancy and labour.

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IDIOPATHIC CARDIOMEGALY

BY

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"Heart muscle," says Cameron (1952), "hypertrophies for many causes, all of which increase the amount of work thrown on the muscle." This conception of hypertrophy involves an increase in the mass of each muscle cell, without an increase in the total number of muscle cells. It also limits the conception to an increase brought about by a work stimulus such as, for example, valvular disease or increased peripheral resistance from hypertension. So far as we are aware, the steps by which work stimulus is translated to increase in cell mass await elucidation. In the meantime it becomes increasingly clear, as more and more cases are reported, that not all abnormal increases in total muscle mass are due to work stimulus. A group of cases variously described as idiopathic hypertrophy, familial cardiomegaly, asymmetrical hypertrophy in young adults, or hypertrophy of unknown aetiology have in common the absence of a work stimulus to account for the increase in muscle mass.

The two cases here reported illustrate the condition of the heart in the latter category. Both cases came to necropsy at the instance of the coroner. Commonly, in coroners' cases there is little or nothing in the way of clinical history to aid the pathologist in diagnosis. If he is confronted by a case of sudden death in which the sole finding is a heart with an obviously hypertrophied left ventricle showing marked excess of weight over the average for the sex, age, and development, the tendency is to attribute death to hypertensive heart

failure unless there is some special feature, such as its occurrence in a child or young adult or an obvious explanation for the hypertrophy. In the second case here reported, which occurred in a middle-aged man, probably such a cause of death would have been declared, but for the fact that hypertension was denied by the clinical history. We feel, therefore, that this condition may be commoner in older age-groups than is generally appreciated.

Case 1

The deceased was a youth of 16 who worked as a clerk. He had never had any serious illness. The only recent medical attention he had had was six weeks before his death, when he had an attack of influenza, from which he appeared to have fully recovered. On the afternoon of his death, he had gone for a ride on his cycle—one of his usual activities—when he was seen to fall off for no apparent reason. A near-by policeman, who was summoned, stated that he found the youth in a coma, breathing very deeply, with purple cheeks, lips, and ears. He died while being conveyed to hospital.

Necropsy Findings

The body was that of a tall youth of normal physique. There were some trivial injuries, sustained by his fall, of no significance as regards the cause of death. There was froth in the trachea. The pleura and lungs appeared healthy. The heart weighed 550 g. It was grossly hypertrophied on the left side, and the cavity of the left ventricle was dilated. The valves were healthy. There was no congenital defect in the heart, its valves, or vessels, and there was no congenital narrowing of the aorta. The muscle of the left ventricle was pale. The papillary muscles were abnormally thick. The left coronary ostium was smaller in diameter than the right, and opened into a main trunk, which divided almost immediately into descending and transverse branches. The latter was very small, and indeed served only a negligible fraction of the usual field of supply of this branch. The descending branch also appeared small, and certainly so in relation to the large muscle mass, which lay in its field of supply. The lumen of the right artery was larger than that of the left and carried a blood supply to most of the area usually serviced by the transverse branch of the left artery. No atheroma

was noted in the coronary arteries. There were no other findings of note except some congestion of the abdominal viscera.

Histological Findings

The heart muscle presented an appearance of bizarre dishevelment (Fig. 1). The fibres were coarse and at many points showed loose ends, as if broken across. Small foci of necrosis were present throughout the muscle. Here and there the muscle was replaced by collagen of loose texture, infiltrated in some cases diffusely with a small number of lymphocytes. Generally there was increased cellularity of the interstitial tissue. The muscle fibres generally were two or three times as thick as is seen in sections of normal heart. The fibrils were discernible as entities, separated by clear spaces, and in both longitudinal and transverse sections there were numerous large clear vacuolar spaces within the fibre (Fig. 2). Many of the nuclei were enlarged to about four times normal size. Many were hyperchromatic, in many the chromatin was finely granular, and some showed disruption of the nuclear membrane. Occasional twin nuclei were to be seen. At the poles of the nuclei were moderate collections of yellowish-brown pigment granules, usually described as lipofuscin. Appropriate stains for fats, amyloid, and glycogen gave negative results. There was no endocardial fibrosis.

Microscopical examination of the left coronary artery showed appreciable subintimal thickening of the main trunk and of the descending branch during its course in the subepicardial tissue (Fig. 3). The intimal thickening was entirely fibrous in character. In parts it was equal to or slightly greater than the thickness of the muscularis. No atheromatous degeneration was seen. In many places opposite to the subintimal thickening the internal elastic lamina showed degenerative change.

Case 2

The deceased was a colliery worker, aged 57, who, until seven years before his death, had been engaged in heavy work at the coal-face, and was in good health. At that time he noticed swelling of the ankles, and experienced discomfort in the right hypochondrium, and breathlessness. He thereupon consulted his doctor, who found oedema of the ankles and feet, an enlarged and tender liver, raised venous pressure, cyanosis, and irregular pulse and heart

FIG. 1.—Case 1. Heart muscle, showing coarse dishevelled appearance with grossly thickened fibres, separation of fibrils, giant hyperchromatic nuclei, and increased cellularity of interstitial tissue. (Haematoxylin and eosin. $\times 240$.)

FIG. 2.—Case 1. Transverse section of heart muscle, showing clear vacuolar spaces within the fibres. (Masson's trichrome. $\times 240$.)

sounds, the rate being uncountable. His doctor found no evidence of mitral disease, hypertension, or thyrotoxicosis. He treated the patient with digitalis for five weeks, with a response which he assessed as fair, and then referred him to hospital, where on examination there was no evidence of congestive failure.

His heart appeared to be of normal size. Blood-pressure was 150/78. There was atrial fibrillation at a rate varying during his stay in hospital from 100 to 124 a minute. The electrocardiogram showed also some ventricular extrasystoles with flattening of T waves. It was thought that the findings indicated coronary ischaemia. There was no suggestion of a recent coronary occlusion. There were no heart murmurs; but numerous widely scattered skin petechiae (which the patient said came and went intermittently) and a palpable spleen prompted an investigation for bacterial endocarditis. He had no rise in temperature during his stay in hospital, and five successive blood cultures were negative for organisms. His haemoglobin level was 14.8 g./100 ml., and the red blood count 5,100,000/c.mm. Sedimentation rate was normal. White blood count totalled 9,500/c.mm., with normal differential. Radiiodine tests disclosed results consistent with normal thyroid function in all respects.

On discharge the spleen had receded in size. His heart was still fibrillating at an uncontrolled rate, but he appeared reasonably well. He was kept on digitalis and was under regular medical supervision until his death. He resumed work as a conveyor-belt attendant, which involved merely the control of switches. Thus he continued with an occasional short absence from work, and with no substantial change in his condition. At no time did his doctor find any rise in his blood-pressure. On October 29, 1958, he was found dead while at work. He had been seen about an hour before, apparently well and cheerful.

Necropsy Findings

The body was that of a well-nourished middle-aged man. The main findings related to the heart, which weighed 675 g. It was dilated and hypertrophied on both sides,

FIG. 3.—Case 1. Coronary artery, showing subintimal thickening. The adjacent muscle shows the appearance of breakage of the fibres. (Masson's trichrome. $\times 60$.)

but particularly on the left. There was no congenital defect in the heart, its valves, or its vessels. The heart valves were normal. There was some narrowing of the coronary arteries, but this was assessed as mild. There was no occlusion. The heart muscle appeared healthy, and no fibrosis was evident. All the peripheral vessels were healthy, and showed no congenital defect. In particular, there was no coarctation of the aorta. There was congestion of the spleen, and the liver was rather fibrous.

Histological Findings

The main findings related to the heart. The changes were substantially the same as those in Case 1, though in this heart the patches of collagen replacement of muscle were generally larger, more condensed, and more numerous than in Case 1. Apart from the changes in the heart, the lungs showed slight thickening and increased cellularity of the alveolar septa. There was some alveolar oedema. The arteriolar walls were slightly thickened. The liver was congested and oedematous. There was slight fibrosis of the portal tracts, and a mild fibrotic change surrounding the central veins of the lobules. The sinuses of the splenic pulp were greatly distended and literally stuffed with red cells. The lining cells of the sinusoids were clearly picked out by reason of their heavy uptake of iron-containing pigment. The smaller vessels of the kidneys were markedly congested, and the pyramidal interstitial tissue was oedematous. There was no arterial or arteriolar thickening in the kidneys. The thyroid gland was normal.

Discussion

The two cases reported belong to a group which is coming to be recognized as one with a characteristic clinical and pathological pattern. As happens when an entity is in process of recognition, there is often dispute and confusion concerning its truly fundamental features. It appears to us that the fundamental features of this group are a marked increase in cardiac muscle mass, an absence of any work stimulus to account for this increase, and a marked liability to sudden death from heart failure. The cause or causes have so far not been defined, though, as is usual in any so-called "idiopathic" entity, there is no lack of suggestion regarding possible causes. Among the commoner causes canvassed have been vitamin deficiency (notably of the B complex), potassium deficiency, strenuous exercise, arrhythmias, anoxia, coronary arteriosclerosis, thyrotoxic heart disease, specific and non-specific myocarditis, subendocardial fibrosis, von Giercke's disease, and amyloid disease. With the exception of anoxia, all of these causes have been excluded from many undoubted cases of this entity, and we agree with Elster *et al.* (1955) that there is little point in reciting a list of suggested causes without any adequate evidence to support their claim.

The question whether deficiency of oxygen to the heart muscle can produce increase in myocardial mass has been debated in the literature. Experimental work in animals and observations on human beings exposed to high altitudes have been held to negative such a hypothesis. However, Serbin and Chojnacki (1955) have suggested (we think rightly) that the exposures in these cases to deficiency of oxygen were too short for a valid conclusion to be drawn. A wide spectrum of oxygen deficiency, both quantitative and as regards duration, can be envisaged in the human. The needs of the heart vary widely with the patterns of activity. There are compensatory mechanisms such as polycythaemia, whose limits we do not know. All of these

features combine to make the dynamic picture so complicated that it is difficult to conceive the possibility of designing suitable experimental conditions.

We think there is clear evidence in human pathology for the thesis that increase in cardiac muscle mass results from anoxia. When the coronary arteries originate in the pulmonary artery the myocardium inevitably suffers from anoxia over a long period. Jurishica (1957), in reviewing 12 adult cases of this anomaly in the left coronary artery, showed that increase in the mass of the left ventricle and liability to sudden death from heart failure were characteristic. Indeed, no one can read the reports of cases of this anomaly without being struck by the similarity of the clinicopathological features to those in so-called idiopathic hypertrophy. Again, it would appear to be clearly established that chronic anaemia causes increase in myocardial mass (Paplanus *et al.*, 1958), and chronic exposure to high altitude has been found to produce increase in cardiac muscle mass (Van Liere, 1936; Kerwin, 1944; Van Liere *et al.*, 1955). The common factor in all of these induced increases in muscle mass is chronic deficiency of oxygen.

In our view the thesis that chronic oxygen deficiency to the heart muscle is the fundamental cause of the clinicopathological entity under discussion is by far the most convincing. Inevitably, this raises the question of coronary insufficiency in these cases. It must be admitted that in many of the cases no mention is made of any defect in the coronary blood supply, but we do not think that is surprising. The assessment of the sufficiency of the coronary blood supply is, in practice, at best a crude one, and we doubt whether it normally involves more than a naked-eye inspection of parts of the coronary vessels by the pathologist. It appears to us that a much more detailed examination of the coronary vessels is required for a valid assessment, and some such injection and radiographic technique as was carried out by Fulton (1956) is called for, particularly in cases where the insufficiency is not concentrated in the major branches of the coronary arterial tree. Weight is lent to the theory of chronic anoxia and coronary insufficiency by the persistent reports of anomalies or defects in the coronary circulation in cases of unexplained cardiomegaly.

In our second case the naked-eye assessment of the coronary arteries was that they were narrow, the degree of narrowing being assessed as mild. Unfortunately only small portions of the heart muscle were retained for histology in this case. In Case 1 the naked-eye investigation of the coronary tree suggested insufficiency, and this was reinforced by the finding of extensive intimal fibrosis of the descending branch of the left artery on microscopy. Small coronary arteries as compared with the grossly hypertrophied ventricles have been noted before (Campbell and Turner-Warwick, 1956; Evans, 1957). Case 1 described by Fulton (1956) was one of cardiomegaly in which hypertension or other work stimulus was ruled out, and the other finding was pronounced coronary atheroma with narrowing. Master (1954), in a study of 600 cases of coronary artery disease, found that increase in cardiac muscle mass occurred "simply and definitely in the presence of coronary sclerosis alone." He held the cause to be obscure, but pointed out that in arteriosclerosis of these arteries there is anoxaemia of the myocardium. We agree with Campbell (1960) that there is too much

tendency to say, when you find a large heart, that there must have been hypertension in the past.

Many of the reports make a feature of the familial character of the unexplained cardiomegaly with the implication that there is some inherited defect intrinsic to the cardiac muscle. It is quite feasible that the familial incidence noted in a number of the reports of this condition may be due to the inheritance of anomalies in the coronary tree, which produce coronary insufficiency and anoxia. The asymmetrical character of the cardiomegaly noted by Teare (1958) is consistent with the theory of anoxic cardiomegaly. It is only to be expected that, if anoxia is produced by mechanical insufficiency of the coronary arteries, then in many cases the condition would have a distribution in the local field of supply of branches of the arterial tree.

Summary

Two cases of cardiomegaly, not due to work stimulus, are described. It is suggested that, since it is clear that cardiomegaly can be a result of chronic deficiency of oxygen, a much more careful assessment of the sufficiency of the coronary blood supply should be carried out in cases of unexplained cardiomegaly than is usual in present pathological practice.

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"In many countries in recent years the numbers of reported cases of gonorrhoea have remained virtually static or have increased—in some countries substantially. Comparative data were presented to the Committee of the number of gonorrhoea cases reported in 22 countries and territories. In no less than 15 there has been a rise in numbers, and in four the situation appears static. The annual incidence ranges from 10 to 50 per 10,000 inhabitants. The magnitude of the reservoir of infection was also demonstrated by the morbidity statistics of gonorrhoea published in a recent 10-year review by W.H.O. The limitations of figures of reported cases are well known, as many unreported cases of gonorrhoea are treated by private practitioners. For example, in the U.S.A. it is estimated that the incidence of gonorrhoea is five to ten times higher than is actually reported. Available information indicates that gonorrhoea is widespread, remains uncontrolled, and is one of the most challenging health problems in many parts of the world." (*Expert Committee on Venereal Infections and Treponematoses*, W.H.O. Technical Report Series, No. 190. Available from H.M.S.O., P.O. Box 569, London S.E.1, price 5s.)