Mucinous Tumours of the Ovary

Cystadenomata of the mucinous and serous types are the commonest varieties of ovarian tumour, probably accounting for 60% of them. The serous type is probably commoner than the mucinous, which is characterized by cystic spaces full of mucus. These cystic spaces are lined by a single layer of tall cells with a clear, refractile cytoplasm and darkly-staining nuclei close to the basement membrane.

Ovarian cystadenomata are benign, but in both varieties a malignant cystadenocarcinoma occurs. This kills by peritoneal implantation and distant metastases. Figures for the incidence of malignancy vary considerably, owing to the existence of a borderline lesion that shares some of the microscopical features of carcinoma but seldom metastasizes. If such intermediate tumours are included in the category of carcinoma, a confusingly good prognosis may be found in a series of cases of ovarian cancer. It is therefore important to study the course history of borderline serous and mucinous ovarian tumours and to see how they differ from the frankly malignant cystadenocarcinoma.

To this end W. R. Hart and H. J. Norris have recently reviewed 688 mucinous tumours examined at the Armed Forces Institute of Pathology, Washington. In 552 cases (80%) the tumour was a typical cystadenoma, while in a further 97 cases (14%) the lesion was more proliferative. The epithelium lining the cystic spaces showed a more exuberant pattern and was stratified into two or three layers. The overall pattern resembled that of an adenomatous polypl of the colon. The epithelial cells had atypical nuclei with hyperchromatism and enlarged nucleoli. Mitotic figures were frequent, but there was no severe degree of anaplasia. In no case was there any stromal invasion. These tumours were classified as borderline. In the remaining 39 tumours (6%) there was undoubted carcinomatous change characterized by stromal invasion and overgrowth of atypical epithelial cells. These stratified cells were more anaplastic than those of the borderline lesion and always exceeded three layers in thickness. Even when stromal invasion was difficult to assess, this epithelial proliferation served to delineate the malignancy of the tumour.

Hart and Norris found that only 3% of the 87 patients followed up with a borderline tumour died of cancer as compared with 33% of the 27 patients followed up with a cystadenocarcinoma. The rupture of the tumour with a spillage of its contents into the peritoneal cavity did not cause adverse effects in either the benign or the borderline lesion.

A rare result of rupture of a mucinous ovarian or appendical tumour into the peritoneal cavity is pseudomyxoma peritonei. The tumour cells take root in the peritoneal mesothelium and secrete large amounts of mucus, which leads to great abdominal distension. The condition recurs when the mucus is removed, but a radical excision of the primary tumour together with cytotoxic agents introduced into the peritoneal cavity may result in a cure. In this series of cases there was no instance of pseudomyxoma peritonei, which the authors suggest arises from a special aggressive borderline mucinous ovarian tumour.

The existence of a borderline variant of ovarian cystadenoma, both mucinous and serous, is now well established. Its prognosis is far better than that of the cystadenocarcinoma. It should be removed by unilateral salpingo-oophorectomy, and the other ovary can be retained after careful inspection. Since many of these tumours occur in young women below the age of 35, their prognostic significance is considerable in relation not only to the patient's life but also her fertility.

Transposition of Great Arteries

Some 300-400 babies are born annually in the United Kingdom with transposition of the great arteries. Their prospects were admirably reviewed in a recent St. Cyres lecture by R. E. Bonham-Carter from the extensive experience at the Hospital for Sick Children, Great Ormond Street. About the same time a report appeared, a model of its kind, of a symposium on heart disease in infancy held in Auckland, New Zealand, in February 1972. These important sources show remarkable unanimity on the treatment of the disorder and enable us to summarize the position.

If they are given no treatment, 85% of infants with transposition of the great arteries will die before their first birthday, either from inadequate mixing of the separate pulmonary and systemic circulations or from harmful effects of a high pulmonary blood-flow. Most of them present with cyanosis and dyspnoea in the first few weeks of life, and accurate diagnosis depends on cardiac catheterization. Though the range of anatomical and haemodynamic abnormalities is wide, there are three main groups of cases.

Transposition with atrial septal defect alone—"simple transposition"—forms the largest group. The extent to which mixing of the pulmonary and systemic circulations is adequate depends on the size of the septal defect. The second group is of transposition with ventricular septal defect. In these cases, if the defect is sizeable, mixing may be adequate but the risk of
pulmonary vascular disease from high pulmonary blood-flow is greater than in simple transposition. The third group comprises transposition with a large ventricular septal defect plus obstruction of left ventricular outflow. The addition of what is in effect pulmonary stenosis removes the risk of pulmonary vascular disease but adds to the difficulties of correction. In a surgical series of 200 cases reported from Great Ormond Street the percentages of these three groups were 69, 22, and 7, though many factors must have influenced their selection.

While the treatment of each baby must be tailored to his individual haemodynamic requirements, in general most of them will benefit from increased mixing of the circulations at atrial level. The Blalock-Hanlon operation for atrial septectomy has been largely superseded by the Rashkind atrial septostomy, performed during the initial diagnostic investigation. Though the mortality of septostomy is lower than that of septectomy, particularly in the first month of life, 35%, of infants still die before 1 year of age even after adequate septostomy. To reduce this wastage, mainly due to thromboembolism and pulmonary vascular disease, cardiologists now generally favour reinvestigation of all cases between 3 and 6 months of age, advising a corrective operation for those found to be particularly at risk. The best age for elective correction of simple transposition without pulmonary vascular disease is probably 6-12 months, though the Auckland group have results to justify their choice of 3 months. Children with the lesions comprising group 3 should wait considerably longer for correction.

One of the few areas of controversy in the recent reports concerns the management of seriously ill infants with transposition and a large ventricular septal defect. The choice lies between correction and a palliative constriction of the pulmonary artery, which reduces the risk of pulmonary vascular disease by obstructing the left ventricular outflow. Probably a staged approach is still preferable, because the mortality of correction is around 50%. Less controversial is the problem of the "failed septostomy." Rather than surgical septectomy, most centres would now offer correction, with a mortality in the best hands of well below 10%.

By "correction" is meant physiological rather than anatomical correction. The temptingly obvious operation of transsecting and "switching" the aorta and pulmonary artery remains impracticable, despite pioneering attempts, largely because of the problem of transposing the coronary arteries with the aorta. Only in group 3 cases is a type of anatomical correction at present feasible in the form of the Rastelli operation. At the Mayo Clinic this operation has carried a 25% mortality, though only 7% in children over 5 years of age. In all other groups of cases correction means the ingenious procedure popularized by W. T. Mustard in which systemic and pulmonary venous flow are rearranged within the atria by means of a baffle of pericardium or prosthetic material.

In cases of simple transposition the Mustard operation gives satisfactory results, with a mortality of less than 10%. The presence of a small ventricular septal defect adds little to the risk, but the concomitant closure of a large defect, likely to be associated with pulmonary vascular disease, raises operative mortality to 20-40%. For group 3 cases correction based on the Mustard operation carried a prohibitive mortality of 50%. However, the Rastelli operation has proved a superior alternative. Though many children are leading normal lives up to seven years after Mustard operations, there are also disquieting reports from more than one centre of second operations to relieve vena caval obstruction. Postoperative arrhythmias have virtually disappeared owing to modifications in operative technique, while anxiety about the long-term competence of the tricuspid valve in the systemic ventricle so far appears groundless.

The advances made in recent years are epitomized by some Auckland data. Before 1967 only 38%, of patients with transposition of the great arteries and poor mixing survived surgical septectomy followed by correction at an average age of 37 months. Since then atrial septostomy has been followed by correction at an average age of 13 months, and 77%, of children have survived. Thus the reports from the experts might be summarized in the schoolmaster's words, "Considerable progress, but still room for improvement."


---

End of Industrial Action

The small size of recent B.M.J.s has been due to an industrial dispute at our printing works. This dispute has now been settled and the journal should soon be back to normal.