The ten best recent articles in the medical literature relating to endocrine malignancies are reviewed here.

The poor prognosis of adrenal cortical carcinoma may be improved by early diagnosis and complete resection. Radical surgery is the sole effective therapy, particularly in early stages. Surgical treatment of recurrence seems to improve survival and should be attempted systematically.

The author presents a well-written review that emphasizes the biology of the multiple endocrine neoplasia syndromes as the basis for good care.

Mutations in the ret proto-oncogene are associated with some familial forms of Hirschsprung's disease. In the case of multiple endocrine neoplasia type 2, the ret mutations are activating (ie, they enhance the function of the encoded protein), whereas in Hirschsprung's disease, the mutations are inactivating and lead to loss of function. In rare families, Hirschsprung's disease and multiple endocrine neoplasia type 2 cosegregate.

There is general agreement that the primary operation for medullary carcinomas of the thyroid should include total thyroidectomy and central neck lymph node clearance. The role of microdissection for recurrent disease awaits longitudinal evaluation. External radiotherapy, radionuclide therapy, and chemotherapy may have roles in palliation but have not definitively shown a curative value.

An algorithm was developed for diagnosis of adrenal lesions that uses the density reading on noncontrast computed tomography scan as the first step, with chemical-shift magnetic resonance imaging for CT-indeterminate lesions. The algorithm is cost-effective and reduces the number of biopsies required without reducing the sensitivity of detecting malignant lesions.

This small pilot study underscores the need for more effective systemic approaches for the treatment of advanced adrenocortical cancer.

Surgery continues to be the primary treatment of endocrine neoplasia. The surgical management of endocrine tumors of the pancreas, the adrenal glands, and the gastrointestinal tract (specifically carcinoid tumors) are reviewed.

This consortium analysis suggests that genotype-phenotype correlations do exist and, if made reliably absolute, could not only prove useful in the future in clinical management with respect to screening, surveillance, and prophylaxis, but also provide insight into the genetic effects of particular mutations.

Biochemical screening indicated that the penetrance of multiple endocrine neoplasia type 1 by the ages of 20, 35, and 50 years was 43%, 85%, and 94%, respectively, and that the development of MEN 1 was confined to first-degree relatives in 91% of patients and to second-degree relatives in 9% of patients. These findings have helped to define a proposed screening program for MEN 1.

Although there is an effect of hypercortisolism on wound healing, infection, diabetes, hypertension, coronary artery disease, and pulmonary embolism, it is possible to perform adrenalectomy surgically with acceptable morbidity and mortality. These results may serve as a standard against which laparoscopic adrenalectomy may be compared.