Problem Management in Endocrine Surgery

GLENN W. GEELHOED

PROBLEM MANAGEMENT IN ENDOCRINE SURGERY

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For
DONALD WILLIAM
and
MICHAEL ALAN

Foreword

IN THIS VOLUME Dr. Geelhoed has concentrated on one aspect of the surgical management of a whole series of diseases and disorders. This is not a book on "diagnosis" or "pre- and postoperative care" or "operative technique." Instead, it concentrates on the events immediately surrounding the operation and, in certain instances, some of the newer aspects of diagnosis that might be carried out by quasioperative means (e.g., transhepatic sampling of the portal vein). Several of the procedures should properly be carried out either in the operating room or close to the time and place of operation: they are "perioperative" diagnostic methods.

As in any book dealing with operations performed by highly specialized surgeons, the object is not to guide the inexperienced by cookbook procedures, taking the surgeon by the hand through procedures he may never see or do again. If a book on surgical management in specialized areas ever has such an object, it is being published 30 years too late.

Instead, this book recognizes that in every major center of the country there will be 3 or 4 people who concentrate on some of the rare endocrine neoplasms and possibly 40 or 50 who deal with the other, commoner disorders. It is to those surgeons, and to those students of surgery, interns, or residents who are pointing in that direction, that this book speaks.

Its usefulness will be found not only in guidance to better and safer surgical care, but also in the stimulus it may give to others to provide alternative opinions, or record in the literature the outcome of experiences conducted by alternative means.

Dr. Geelhoed is to be congratulated on completing this remarkable volume. May it have a long and useful life, passing through future editions as this rapidly changing field further evolves.

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Preface

SURGICAL ENDOCRINOLOGY is a dynamic, expanding new discipline. Understanding of neuroendocrine control systems has developed rapidly with the integration of genetic, embryologic, and biochemical information into meaningful patterns. Refined techniques of radioimmunoassay and radiographic imaging have made diagnosis and localization of endocrine hyperfunction much more precise. As a result, surgical treatment can now be guided toward restoring patients to the normal balance of physiologic autoregulation as well as curing them of endocrinologic excess.

Endocrine surgical patients usually seek help for the resolution of problems that are functional; rarely is the mere presence of an endocrine tumor the patient's complaint. In only one anatomical area discussed in this book—the thyroid—is the tumor mass likely to be a problem noted by the patient. More often, *finding* a hidden tumor will be a problem for the physician. However, increasing application of sophisticated imaging techniques on asymptomatic patients has increased the incidental discovery of nonfunctioning masses in endocrine organs. With the exception of these "incidentalomas" that are localized before diagnosis, the increasing sensitivity and specificity of endocrinologic testing can confirm the diagnosis of preclinical disease. When cautiously applied—particularly in stimulation tests for screening family members with hereditable endocrinopathy—endocrine surgery can *prevent* some diseases before they cause patient problems.

This book is designed to deal with endocrine surgical problems: those presented by the patient in the clinical syndromes of their disease, and those posed to the physician in management of the diagnostic and therapeutic options among a sometimes bewildering array of new methods. It is hoped that the text will be useful to the student who is taught by patients, whether he be at an undergraduate, postgraduate, or continuing education level in surgery, medicine, or radiology.

Each chapter begins with the patient's problems as a starting point for the management process by the physician. The tumors, hormones, and syndromes are reviewed as background for a diagnostic approach in screening, confirmation, and localization. Discussion of treatment includes preoperative preparation, anesthesia and operative technique, postoperative

care, and follow-up for special consideration of each of the endocrine surgical problems.

Any book dealing with the challenging problems and myriad unanswered questions in a rapidly advancing field will be biased by the prejudices and limited by the experiences of the author at the time it is written. However, the conceptual framework of neuroendocrinology as an integrated physiologic control system can be helpful to the reader in assimilating new information in endocrinology as it develops. It is easier to assay than to understand the abundance of hormones and candidate hormones that are now emerging; but the clinical significance of a patient problem without a known mediator remains more compelling than that of a candidate hormone without a known effect. By observing and listening to our patients' problems we will learn to further modify our concepts and to initiate new procedures in the management of neuroendocrine disorders.

GLENN W. GEELHOED

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1 / Introduction: Neuroendocrinology and Endocrine Surgery

Endocrine surgery is physiologic surgery. The endocrine surgeon must recognize the often subtle deviations from normal physiology in the effects of abnormal metabolism in the target end-organ. The metabolic derangements that are the primary problem in surgical endocrinology make this discipline distinctive from most other fields of surgical specialty interest, which often begin with anatomical derangement and "mass lesion" effects. The anatomical abnormality is often inapparent as the endocrine surgeon follows the physiologic clues through the identification of a humoral mediator. The rational inference of an abnormality in this humoral mediator based on the observed clinical findings may lead further to a search for the source of hormonal hyper- or hyposecretion. Observing the effects of the mediator in the clinical syndrome evident through the target organ's reaction leads the physician back primarily to the source of the endocrine secretion. Only then, with diligence in the use of radiologic and operative techniques for localization, does the surgeon encounter the anatomical defect that gave rise to the hormonal imbalance and the clinical syndrome. This discovery of a morphological abnormality in the endocrine surgical patient often requires the pathologist's techniques to confirm subtle variants from normal structure. Endocrine surgery, then, is not so much the surgery of mass lesions and the correction of anatomical defects as the restoration of normal endocrine physiologic balance in functions. Looking forward along the same inferential pathway, this book will discuss each of several endocrine surgical abnormalities from the starting point of the tumor, its hormones, and the clinical syndrome that results.

The myriad and often confusing presentations of patients with hormonal disorders would require a large capacity for rote memory from the clinical observation of these syndromes. A long experience would be required before the difficult integration of much of this clinical information into meaningful patterns might be possible. Fortunately, the details of clinical observation cluster—not randomly, but within recognizable patterns—in the clinical syndromes. Study of hormone origin and function makes the organization of the details of clinical experience much more rational and easier

not only in remembering the clinical patterns but also in assimilating new information in endocrinology. The precision of many endocrinologic diagnostic tests, enhanced by radioimmunoassay techniques, has sharpened the clinician's focus on clinical syndromes. New radiologic techniques contribute to a very refined capability to localize subtle morphological abnormalities preoperatively. The organization of diagnostic methods used in this book will proceed from screening methods through confirmation of the endocrine diagnosis to localization techniques (Table 1–1). It is the surgeon's responsibility, then, to correct the disorders of surgical endocrinology by intervening directly and by reducing or ablating endocrine tissue, correcting hyperfunction, removing neoplasm, and restoring the patient, ideally, toward the balance of physiologic autoregulation.

This pattern of behavior in endocrine surgery is generally applicable, but becomes critical in some specific endocrine surgical abnormalities that may be life-threatening. Recognition of these high-risk endocrine abnormalities requires that the surgeon be a capable endocrinologist who can manage the patient problems that result from such life-threatening syndromes. Safety in patient management and judgment in intervention are critically important in endocrine surgery. Preoperative preparation of the patient and details of postoperative management parallel the technical aspects of opera-

TABLE 1–1.—OUTLINE FOR DISCUSSION OF EACH CLINICAL SYNDROME IN THIS BOOK

Management problems
For the patient
For the physician
Introduction to the endocrinologic abnormality
The tumor
The hormone
The syndrome
Diagnosis
Screening
Confirmation
Localization
Preoperative preparation
Anesthesia management
Operative technique
Postoperative care and drug management
Follow-up and special considerations

tive dissection in importance. Kocher's technique of thyroidectomy awaited Plummer's use of iodine before the operation could be safely performed to the benefit of patients with hyperthyroidism. Repletion of contracted blood volume and restoration of electrolyte deficits are major features of the endocrine surgery of several syndromes, rivaling the significance of operative technique. The careful use of blocking agents and proper anesthetic technique are critically important in the management of the patient with pheochromocytoma. A major focus of this book will be on the management of these perioperative problems, including preoperative preparation and anesthesia management (Table 1–1), as well as on operative technique.

Endocrine tumors rarely present solely as mass lesions producing signs and symptoms by impinging upon or growing into adjacent viscera. Endocrine tumors rarely disrupt function by their presence alone. In two instances in this book we will discuss primary lesions simply because they occupy space; one such tumor is in the neck and the other is hidden in the silent suprarenal retroperitoneum. A more common problem in endocrine surgery is locating the source of hormonal hypersecretion. The reverse of symptoms caused by a tumor merely by its mass is the problem in localizing a source of known hypersecretion that may remain obscure even on the operating table with the gland exposed for inspection, palpation, and biopsy examination.

BIOLOGIC CONTROL SYSTEMS AND THEIR IMBALANCE

Biologic control systems are servomechanisms that delicately seek a balance in homeostasis. In the most simple example, a gland secretion, a hormone, exerts an effect upon a target organ, which in turn results in diminished hormonal secretion. The source of such a hormone may not be a single or paired gland: the chromaffin system consists of many nests of cells located throughout the body, and this more diffuse system has broken down the concept of "a ductless gland" as a focal distinctive organ. It is now recognized that a "gland" may not be a group of cells located in a single anatomical site; similar cells may act from widely scattered anatomical locations. For example, different species of pancreatic islets are scattered within an exocrine gastrointestinal (GI) tract gland, and similarly functioning cells may be found outside the pancreas itself.

The body's biologic control systems can be approached either through endocrinology or through the study of a different discipline—neurology. It could be postulated that the CNS is primarily in control of all the activity of the organism's component parts. The CNS transfers information by electric impulse and by neurotransmitters. The belief that the CNS and the

endocrine system were independent in exerting control has been confounded by the later discovery that some of these CNS neurotransmitters circulate systemically with hormonal effects, and hormones controlling GI secretion have been found in the CNS acting as neurotransmitters!

Integration of the CNS and peripheral hormonal secretion is accomplished by the ductless messenger system of the hypophysis–pituitary portal venous system. Releasing factors elaborated by the hypophysis transfer information from the CNS to the pituitary gland. The pituitary releases a hormone (ACTH, TSH, and so on) which influences another endocrine gland in the periphery. The peripherally secreted hormone is perceived by the CNS and hypophyseal secretion is shut down, completing the servomechanism loop (Fig 1–1). Further examples of interesting collaboration between these biologic control systems have become more apparent whenever one chooses to study a patient problem as one of purely endocrine or neurologic abnormality.

When interruption of the loop in this neurohumoral servosystem occurs, clinical syndromes of hormonal excess or deficiency result. The precise site of interruption may not be apparent. Cushing's *disease* is based upon excess ACTH production by the CNS; Cushing's *syndrome* results from ex-

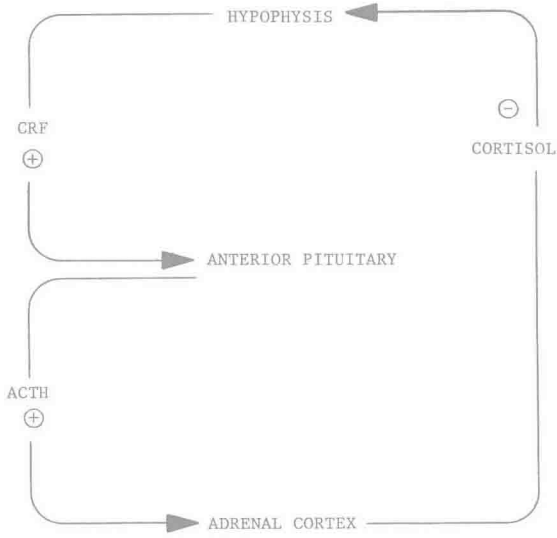

Fig 1–1.—The hypophysis-pituitary-adrenal axis offers a near-perfect example of the integration of neuroendocrine control systems, with neurotransmitters and classic hormones in feedback control of a vital homeostatic system. CRF, corticotropin-releasing factor.

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