

### INTRODUCTION

Certain endocrine disorders are managed surgically: the most common are disorders of the thyroid gland (see Chapter *Thyroid Gland*) and less commonly are those of the parathyroids, adrenal gland and certain rare islet cell lesions of the pancreas. There are fundamental processes that all have in common: Clinical recognition, biochemical confirmation, anatomical localisation, selection for surgery, surgical operation and the sequelae. Close co-operation between the surgeon and endocrinologist or physician provide optimal patient care.

### HYPERPARATHYROIDISM

#### *Types of hyperparathyroidism*

Hyperparathyroidism (HPT) is a condition of excess secretion of parathyroid hormone.

The most common form is *primary* hyperparathyroidism where the disease is primarily that of the parathyroid glands, most commonly due to a single adenoma, or less commonly due to 4 gland hyperplasia. The excess secretion of parathyroid hormone (PTH) results in the consequent features of the disease.

#### Types of hyperparathyroidism

- Primary  
single adenoma (95%)  
4 gland hyperplasia (5%)
- Secondary / tertiary  
renal failure, dialysis  
↓  
(P04 retention, less  
hydroxylation of vit D,  
reduced calcium absorption)  
↓  
Parathyroid stimulation  
↓  
Bone loss

*Secondary* and *tertiary* hyperparathyroidism may arise in patients with chronic renal failure where phosphate retention and calcium deficiency secondarily stimulates the glands to secrete PTH in order to restore low normal calcium levels; in some patients the process may become autonomous - *tertiary* hyper-parathyroidism.

### PRIMARY HYPERPARATHYROIDISM

The aetiology is not clear. In small proportion of cases associated with multiple endocrine neoplasia (MEN), where 4 gland hyperplasia is usually found, there is a genetic abnormality, the abnormal locus having been identified on chromosome 11. In most cases, however, the cause of the adenoma is unknown.

PTH stimulates the osteoclast and promotes bone re-absorption, producing the characteristic bone changes, and releasing calcium, phosphate and alkaline phosphatase. The calcium and phosphate are presented in excess to the kidney and into the urine, which may result in stone formation. Although PTH promotes calcium absorption and phosphate loss in the renal tubule, the excess calcium "spills over". The hypercalcaemia may affect many organs including the brain.

The most common cause of hypercalcaemia in a non-hospital population is hyperparathyroidism; in a hospital population, however, malignant disease makes up half the cases. In the past, certain calcium containing ulcer medications taken with milk ("milk alkali syndrome"), and excessive administration of cod liver oil and vitamin D preparations (hypervitaminosis D) produced hypercalcaemia: These are no longer encountered. Hypercalcaemia (>2.6 mmol/L) and its implications is

frequently missed in clinical practice. Every case of hypercalcaemia requires a clinical explanation.

### Clinical Features

The diagnosis of primary hyperparathyroidism depends on the possibility coming to mind in certain common clinical situations. *Bone, stones, groans and moans* is a convenient memory aid to the effects of the disease. In addition to these well known modes of presentation, *hypercalcaemic crisis* (dysequilibrium hypercalcaemia) may present as a fulminating emergency with rapidly rising calcium levels, polyuria and dehydration, and subsequent confusion, coma and death. This emergency requires major rehydration, hypocalcaemic drugs and urgent surgery.

#### **Bones:**

This form of presentation is rare (von Recklinghaus disease) although radiological and densitometric abnormalities are common.

#### **Stones:**

The most common clinical presentation.

#### **Groans:**

Abdominal pain from renal stones is common; association with pancreatitis and ulcer are questioned.

#### **Moans:**

Psychiatric abnormality (eg: depression) is common.

#### **Asymptomatic:**

A significant number present with incidental hypercalcaemia.

### Diagnosis

The diagnosis is made from the clinical features (eg: renal stones) and the biochemical confirmation. Many cases

are asymptomatic, and the only clue to the diagnosis is unexplained hypercalcaemia. The diagnosis usually rests on two observations: elevations of the serum calcium and parathyroid hormone. Anatomical localisation of enlarged glands by ultrasonography is not part of the diagnostic process as it is rather inaccurate, and may be misleading. When, however, the biochemical diagnosis has been made and surgery is indicated, special localising studies may be performed (ultrasonography, HIDA isotope scanning, PTH venous sampling) to aid surgery

### Biochemical Features

ELECTROLYTES	elevated: Ca, Cl, Alk Phos depressed: PO <sub>4</sub> , HCO <sub>3</sub>
URINE	elevated: Ca, n CAMP
HORMONE	elevated PTH

### Treatment

Surgery is undertaken in most cases: the single adenoma is removed, or in the case of 4 gland hyperplasia, 3 glands are removed. Asymptomatic patients are submitted to surgery unless they have a severe medical contra-indication as they may proceed to renal damage, osteopaenia or psychological abnormality in later life.

### SECONDARY/TERTIARY HYPERPARATHYROIDISM

Calcium deficiency may develop during the course of chronic renal failure, particularly in patients on haemodialysis. Many mechanisms are responsible for this: decreased gut absorption (which is vitamin D mediated), decreased levels of vitamin D, due to absent renal hydroxylation of calciferol, and perhaps PTH resistance in bone. Phosphate retention and the falling calcium levels stimulate the parathyroid glands (hyperplasia), and the excessive levels of PTH then stimulate osteoclast activity causing osteopaenia.

The clinical picture is that of a patient in chronic renal failure who develops osteopaenia with elevated alkaline phosphatase and PTH levels. If the serum calcium is normal or low, the condition is called *secondary* hyperparathyroidism, and if elevated *tertiary* hyperparathyroidism. The surgical management is to remove 3 of the hyperplastic glands. The fourth gland may be partially removed or transplanted into the forearm for easy access should it become overactive.

### ISLET TUMOURS OF THE PANCREAS

The pancreas consists of *exocrine* cells which secrete pancreatic enzymes into the duodenum and *endocrine* cells which produce hormones which are secreted into the blood. The latter are located in the pancreatic islets and may undergo neoplastic change becoming adenomas or carcinomas: the islet tumours of the pancreas. The most common tumours are gastrinomas and insulinomas; glucagonomas and vipomas are excessively rare.

B cell - insulin	insulinoma
A cell - glucagon	glucagonoma
D cell - gastrin	gastrinoma

#### Gastrinoma

The gastrin producing cells of the pancreas and duodenum - but not the antrum - may undergo neoplastic change and produce adenomas and carcinomas which secrete gastrin, with excessive acid production and the features of the disease. The triad of recurrent peptic ulceration, marked acid secretion and an islet tumour defines the Zollinger Ellison syndrome. The diagnosis rests with the clinical suspicion of the condition and demonstration of elevated serum gastrin levels, together with raised acid secretion, particularly the basal (fasting) secretion. The lesions are localised and staged by CT scanning, but surgical exploration is regarded as

the most accurate method of localisation. Many lesions are located in the duodenal wall. Localised lesions without metastases are best treated by surgical removal; more advanced lesions may be managed by a proton pump inhibitor (if the tumour has been shown to be responsive and the patient compliant); in other cases a total gastrectomy, to abolish acid secretion, is undertaken.

#### Suggestive features

- recurrent ulcer
- refractory ulcer
- multiple ulcers
- ulcers in distal sites
- ulcer and diarrhea
- ulcer and MEA

#### Insulinoma

An adenoma of the islet cells may secrete insulin autonomously and continuously. In normal people, after exercise or fasting, insulin secretion reduces; with insulinoma, however, the secretion continues, the blood levels of insulin being *inappropriate* for the glucose level (usually after an overnight fast) and the demonstration of an adenoma of the pancreas on CT scanning.

#### Causes of hypoglycaemia

- Anti-diabetic drugs
- Liver disease
- Alcohol
- Late dumping
- Insulinoma

Portal venous insulin sampling may be done to locate the lesion for surgical excision.

#### Suggestive features of insulinoma

- *Hypoglycaemia* (usually on exercise or fasting)
- *Neuroglycopenia* (weakness, visual disturbance, amnesia, confusion, abnormal behaviour, convulsions, coma.)
- *Catecholamine release* (sweating, palpitations)

## **MEN1 and MEN2**

Certain endocrine neoplasms may occur concurrently or sequentially in the same individual, and the trait may be inherited. These groupings are called the MEN (multiple endocrine neoplasia) syndrome. The location of the insulin, calcitonin and Harvey ras oncogene on chromosome 11 might suggest a hereditary abnormal regulation at this site. Individuals with any component should be tested for the others (eg: patients with gastrinoma should have growth hormone, prolactin and PTH measured; patients with phaeochromocytoma should be investigated for medullary thyroid carcinoma). Relatives of patients with the MEN syndrome should be screened for the disease.

## **COMPONENTS**

### **MEN 1** (Wermers syndrome)

*Pituitary* (acromegaly, prolactinoma)

*Pancreas* (insulinoma, gastrinoma, etc.)

*Parathyroid* (Hyperparathyroidism)

### **MEN 2** (Sipple syndrome)

Medullary thyroid cancer

Phaeochromocytoma

Hyperparathyroidism

### **MEN 3** (2b, mucosal neuroma syndrome)

Medullary thyroid cancer

Phaeochromocytoma

Marfanoid habitus

Mucosal neuromas

## **ADRENAL GLANDS**

### ***Cortical function***

The outer *zona glomerulosa* secretes the mineralo-corticoid, aldosterone. The inner *zona fascicularis* and *zona reticularis* together secrete glucocorticoids (cortisol and corticosterone), androgenic steroids and certain steroid precursors.

The hormones are secreted in response to external demand. Cortisol secretion is controlled by pituitary ACTH, the levels being maintained by a feed-back loop, which acts on both the pituitary and hypothalamus.

Aldosterone secretion is influenced by angiotensin (which is controlled by renin production), and by the concentration of plasma sodium and potassium. The sex hormones have a minimal role, the major role being taken by gonadal secretion.

### ***Medullary function***

The adrenal medulla secretes adrenaline, noradrenaline and dopamine, the noradrenaline precursor. Control is through the splanchnic nervous system. The urine may contain small amounts of these catecholamines, as well as their meta-metabolites, and vanillylmandelic acid (VMA). Medullary function may be assessed by urinary measurement of these.

Adrenaline stimulates alpha and beta adrenergic receptors, redistributing bloodflow from the skin and splanchnic vessels to the heart, brain and muscles. Noradrenaline stimulates alpha receptors, causing generalised vasoconstriction and elevation of blood pressure.

### **Imaging the adrenal gland**

The best images of the adrenal gland are obtained with a CT scan or its latest modification, the spiral CT scan. Excellent images are also obtained with nuclear magnetic resonance scanning, where the T2 weighted images show detailed adrenal anatomy. Unfortunately, these apparatus are expensive and not readily available. Complex adrenal problems are best referred to tertiary centres where CT scanning facilities are available.

Ultrasonography is less satisfactory, but may be helpful in many cases. Intravenous pyelography may demonstrate displacement of the kidney, and the presence of an adrenal mass. A plain film of the abdomen may demonstrate adrenal calcification.

Arteriography may be used to demonstrate adrenal tumours, but is contra-indicated when a pheochromocytoma is suspected, as it may precipitate an attack. Venography and retroperitoneal air insufflation are no longer performed.

Certain important adrenal lesions are only a few centimeters in diameter, and require detailed imaging.

## Diseases

### ***Cushing's<sup>1</sup> Syndrome***

Cushing's syndrome refers to the clinical features found with an excess of adrenal cortical hormones. The pituitary-based variant was first described by the American neurosurgeon Harvey Cushing. Most body systems may be affected, and the presentation may be subtle. The diagnosis is frequently missed by many doctors: the obese patient with depression, hyper-tension, amenorrhoea and diabetes may well have Cushing's Syndrome.

There are various causes of the syndrome, the most common being the administration of steroid therapy. In patients not on therapy, the most common cause in women is Cushing's Disease (the problem being a pituitary adenoma), in men carcinoma of the bronchus (the problem being an ACTH secreting bronchial tumour), and in children the most frequent cause is an adrenal adenoma.

### **Screening for Cushing's syndrome**

The differential diagnosis includes obesity and alcoholism. Three 24 hour urinary samples should be taken and tested for cortisol and perhaps other steroid hormones. A 24 hour urinary cortisol level of more than 100µg is diagnostic of Cushing's syndrome. The episodic nature of cortisol secretion makes random blood cortisol levels quite valueless. A low dose dexamethasone test (1mg orally at 11pm, and measure plasma cortisol at 8am). Normal individuals will have suppressed levels below 3µgms/100 ml.). In rural areas the eosinophil (Thorn) test may be used: an injection of ACTH is given and a fall in the total eosinophil count obtained, indicating a normal functioning adrenal gland. Once the diagnosis of Cushing's syndrome has been made, one then proceeds to locate the cause.

#### CLINICAL FEATURES OF CUSHING'S SYNDROME

- **Central obesity 90%**  
round face  
buffalo hump  
supraclavicular fat pads  
truncal obesity  
thin arms and legs
- **Weakness, proximal myopathy 80%**
- **Hypertension 80%**
- **Skin changes 70%**  
thin skin, bruising  
acne, greasy skin  
hirsutism  
plethora  
abdominal striae  
infection  
pigmentation
- **Psychiatric changes**  
mental slowing  
depression
- **Oligo / amenorrhoea / impotence**
- **Osteoporosis**
- **Thirst / polyuria**
- **Glucose intolerance**

### **Causes of Cushing's Syndrome**

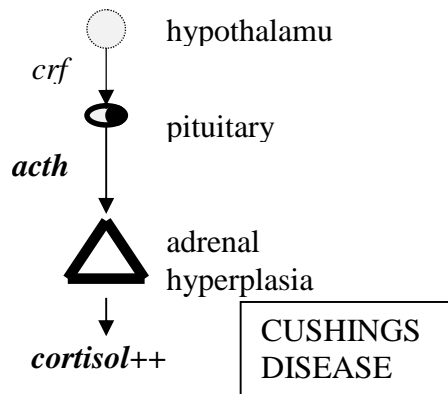
- Steroid administration
- Cushing's Disease 70%
- Ectopic ACTH 10%

<sup>1</sup> Harvey Cushing, Boston. 1869-1939.

- Adrenal adenoma 10%
- Adrenal carcinoma 10%

### Localizing the cause of Cushing's syndrome.

A good screening test is the serum ACTH level: suppressed levels



indicate an adrenal level neoplasm, normal or slightly raised levels indicate a pituitary level adenoma, and markedly elevated levels indicate a non-endocrine ACTH secreting neoplasm. The high dose dexamethasone suppression test (2mg 6hourly for 2 days) relies on the fact that autonomous neoplasms (cortical adenomas and non-endocrine tumours) will not respond to the external feed-back stimulus of dexamethasone, but that pituitary level Cushing's disease will.

Anatomical localization is best achieved by CT scanning of the pituitary and the adrenal glands. A skull radiograph may show an enlarged sella in 20% of cases of Cushing's syndrome. Iodocholesterol scanning of the adrenals may show a "hot" adenoma. Ultrasonography is less sensitive in localization.

### Treatment

**Cushing's disease** is a disorder that mainly affects women between 20 and 50 years. The lesion is located at pituitary level and is usually a corticotroph adenoma, driven by CRF from the hypothalamus. It is managed by transphenoidal resection of the tumour, which is successful in 80% of

cases. Radiotherapy and medical therapy are adjuncts to this. Where pituitary surgery has failed, bilateral adrenalectomy with replacement therapy may be considered. *Nelson's syndrome* refers to the development of a pituitary adenoma due to lack of feed back in the absence of adrenal tissue.

**Adrenal adenomas** are managed by adrenalectomy, and a gradually reducing replacement therapy.

#### Inappropriate hypertension

- hypertension in the young
- rapidly progressive hypertension
- poorly controlled hypertension
- paroxysmal attacks

**Adrenal carcinomas** are resected if possible, and the tumour bed irradiated.

**Ectopic ACTH** producing tumours (usually of the bronchus, but also the pancreas, thymus and at other sites) require control of the primary lesion. If this is not resectable, medical adrenalectomy with metyropone, aminoglutethamide and mitotone may be undertaken. The occurrence of Cushing's syndrome in a patient with an ACTH secreting malignancy is usually a terminal event in a patient dying from that malignancy. Frequently no medical action is taken.

### Phaeochromocytoma

*Phaeochromocytomas* are tumours of the adrenal medulla, or more rarely (10%) the sympathetic tissue adjacent to the vertebral column, where they are called *paragangliomas*. The name comes from the Greek: *phaios* (dark), *chroma* (colour). They characteristically secrete adrenalin and noradrenaline, and untreated are invariably fatal. Approximately 10% are malignant, and 10% multifocal.

Phaeochromocytomas may be associated with the MEN11 (Multiple endocrine neoplasia, type 2) syndrome: medullary carcinoma of the

thyroid, hyperparathyroidism and neurofibromatosis.

### Clinical features

These are analogous to a constant or intermittent infusion of adrenaline or noradrenaline. Some may be missed in life, and the patient undergo a fatal cardiovascular illness, sometimes precipitated by surgery for other indications.

PRESENTATION OF PHAEOCHROMOCYTOMA	
<ul style="list-style-type: none"> <li>Sustained hypertension</li> <li>Paroxysmal hypertension</li> <li>"Attacks" <ul style="list-style-type: none"> <li>headache</li> <li>palpitations</li> <li>blurring of vision</li> <li>sweating</li> <li>abdominal pain</li> <li>anxiety</li> </ul> </li> <li>Major cardiovascular illness (pulmonary oedema, stroke, infarct)</li> </ul>	

### Investigations

Patients with *inappropriate* hypertension should be screened for surgically correctable diseases, including phaeochromocytoma. The most widely used biochemical test is urinary VMA; the urine is best collected after an "attack". Where laboratory facilities are available, other urinary metabolites may also be measured, as well as serum catecholamines.

Localization of the tumour is best done with a CT scan. Radio-iodine labeled MIBG (meta-iodobenzylguanidine) will show up the tumour location. Other imaging modalities are less successful. Paragangliomas along the abdominal vertebral column, as well as a lesion at the bifurcation of the abdominal aorta (organ of Zuckerkandl) may be sought.

### Treatment

Patients with these tumours are best managed in major centres. An essential preparation is alpha-

blockade using the drug phenoxybenzamine (starting at 10mg twice a day, and gradually increasing the dose); such blockade may take 10 days or more, and is judged adequate when there is postural hypotension, a slow pulse and nasal stuffiness. The progressive loss of vasoconstriction leads to normovolaemia, and avoids dangerous hypotension due to loss of vasoconstriction after removal of the tumour. Intra-operative sodium nitroprusside or magnesium infusion may be required to avoid blood pressure swings during tumour handling.

### Conn's<sup>2</sup> Syndrome

*Conn's syndrome* refers to the presence of an aldosterone secreting adenoma of the adrenal cortex ("aldosteronoma"), which has the exaggerated effect of an excess of aldosterone action and causes hypertension and hypokalaemia. The condition is extremely rare, and should be suspected when hypertensive patients present with muscular weakness from hypokalaemia ( $K^+ < 3$  mg). Thiazide diuretics may also cause hypokalaemia in hypertensive patients, and should be stopped for two weeks before the potassium level is re-checked, and the diagnosis reconsidered. Patients with suspected Conn's syndrome should be referred to major centres for further investigation.

### Investigation of Conn's syndrome

This requires the following investigative steps:

- Confirmation of hypokalaemia, and excessive urinary potassium secretion.
- Confirmation of excessive aldosterone secretion by measuring blood and urinary levels. (ie *hyper-aldosteronism*)
- Confirmation of *primary hyperaldosteronism*, and exclusion

<sup>2</sup> Jerome Conn, Ann Arbor. 1907-

of secondary variants by demonstrating depressed renin levels.

- Localization of the adenoma by CT scanning.

Surgical removal of the adenoma leads to normotension in approximately 70% of cases, and reduction in antihypertensive therapy in the remainder.

Conn's syndrome constitutes one of several causes of hypertension, which may be corrected by surgical means. While hypertension is common, certain subsets of patients merit investigation, as the surgical correction of their problem is rewarding, leading to cure, and reduction in drug cost.

Surgically correctable hypertension
<ul style="list-style-type: none"> <li>• Coarctation of the aorta</li> <li>• Renovascular hypertension</li> <li>• Pheochromocytoma</li> <li>• Conn's syndrome</li> <li>• Cushing's syndrome</li> </ul>

### Adrenal carcinoma

Adrenal carcinoma is a rare condition, where most patients present with advanced disease. Syndromes of hormone overproduction occur in about half the patients (Cushing's syndrome, hyperaldosteronism, virilisation), a palpable mass is present in 50% and 25% have hepatomegaly. Local invasion and metastases are common.

These patients have a poor prognosis, and the only hope of cure is resection of earlier, smaller, non-metastasizing lesions. Radiotherapy and chemotherapy yield disappointing results.

### Adrenogenital Syndrome

There are various genetically determined enzyme defects, which impair adrenal steroid synthesis. This

causes an increase in pituitary ACTH production and resultant adrenocortical hyperplasia, with inappropriate adrenal androgen secretion. The consequences depend on the sex and age of the patient.

Infant girls have an enlarged clitoris, and varying fusion of the labial folds. Virilism supervenes with the development of pubic and facial hair; temporal alopecia, deepening of the voice, amenorrhoea and minimal breast development.

Young boys undergo precocious puberty, with excess muscle growth, but short stature ("infant Hercules").

The condition is treated with cortisol to suppress ACTH. Occasionally plastic surgical correction of genital abnormalities is required.

### Adrenal feminization

Certain adrenal tumours secrete oestrogen and cause precocious puberty in females and feminization (gynaecomastia, testicular atrophy, loss of facial hair) in males. These tumours are usually malignant, and surgical removal is attempted. Recurrence and metastases are common.

### Incidentaloma

Diseases of the adrenal gland may be discovered incidentally at autopsy, surgery, or during imaging procedures (usually CT scanning) for other problems.

### Adrenal lesions found at autopsy

Malignancy <1%

nodular hyperplasia, adenoma, adenolipoma, cyst, myolipoma, metastases pheochromocytoma, cortical carcinoma, ganglion neuroma.

Such lesions have been called "incidentalomas". The vast majority are benign, silent and do not affect the patient's life. The discovery of such a lesion requires a balanced approach: if



here are features that may suggest a pheochromocytoma, appropriate investigations should be done. If the lesion is more than 5cms in diameter, an adrenal carcinoma should be excluded.

### **Adrenal insufficiency**

Addison's<sup>3</sup> disease bears the name of the British physician who described it in 1855. The causes include destruction of the gland by Tuberculosis, auto-immune adrenalitis, and metastatic disease. Tuberculosis is the most common cause where this disease is prevalent. The increasing prevalence of AIDS has seen an increase of this condition. Metastatic disease from breast and bronchial cancer, as well as melanoma find adrenal insufficiency a terminal event in these diseases.

### **Clinical and investigative features**

Features are variable and result from mineralocorticoid and glucocorticoid insufficiency. These are fever, nausea, vomiting, severe hypotension and lethargy. Chronic symptoms are more subtle and include fatigue, weight loss, anorexia, nausea, vomiting, abdominal pain and diarrhoea.

Laboratory findings include hyponatraemia, hyperkalaemia, hypoglycaemia and renal failure. There may be eosinophilia, and adrenal calcification may be visible on a radiograph. Low serum levels of cortisol, together with high ACTH levels are diagnostic.

### **Management**

The management of an adrenal crisis may be necessary before laboratory confirmation is made. Immediate administration of 4mg dexamethasone, together with intravenous administration of normal saline may be necessary.

Chronic states require glucocorticoid and mineralocorticoid supplementation. (See: *Replacement therapy*.)

### **Other diseases**

#### **Ganglioneuroma**

These are well encapsulated benign tumours, which arise from ganglion cells. Their growth is progressive and slow, reaching a large size. Surgical removal is indicated.

#### **Neuroblastoma**

This is one of the most common malignant tumours found in infancy and childhood. Most arise in the adrenal gland, but some may arise at other sympathetic nervous system sites, such as the mediastinum and retroperitoneum. About 75% of these tumours secrete catecholamines. Haematogenous spread occurs to the brain, bone, lung and liver.

Half of the cases occur before the age of two years and most before the age of 10 years. The presenting feature may be a mass in the loin, or in some cases, with metastatic disease.

*Investigation* includes CT scanning, and estimation of urinary VMA. A metastatic screen of investigations would examine the lung, brain and liver. The treatment is a combined modality one: after surgical removal of the primary, the tumour bed is irradiated, and adjuvant chemotherapy administered (cyclophosphamide, vincristine).

### **Surgical approaches to the adrenal glands**

The investigation and management of diseases of the adrenal gland is complex, difficult and requires special facilities and expertise. Patients with suspected adrenal disorders which require surgery should be referred to specialist centres.

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<sup>3</sup> Thomas Addison, London. 1793-1860

The confirmation of diagnosis requires many special investigations, and relies on biochemical facilities and CT scanning. Where a definite diagnosis has been made, certain preparations must be made prior to surgery: patients with a pheochromocytoma must have mandatory therapeutic alpha-blockade (phenoxybenzamine) for approximately 10 days, and patients with Conn's and Cushing's syndromes require potassium supplementation.

There are various surgical approaches to the adrenal gland, which are dictated by the disease and the preference of the surgeon. Adrenalectomy for Cushing's syndrome is characterized by the difficulty of excess truncal fat, and extra-peritoneal approaches are preferred: an extraperitoneal, extrapleural approach in the line of the 10th rib interspace provides good access, as does an approach posteriorly through the bed of the 10th rib. CT scanning has made the intra-operative search for contra-lateral and extra-adrenal Pheochromocytomas unnecessary, and these tumours may be removed extra-peritoneally, or trans-peritoneally.

Recent approaches have been laparoscopic, but it has become clear that such approaches are only suitable for smaller tumours, and that the surgeon should be familiar with the open route, should he have to convert to it.

### **Replacement therapy**

Replacement therapy is required after bilateral adrenalectomy, or unilateral adrenalectomy for an adrenocortical adenoma. Post-operative hydrocortisone sodium succinate (100mg 8hourly intramuscularly) is given until the patient is able to take oral medication. Thereafter the oral dose is tapered down to hydrocortisone 100mg daily in divided doses and fludrocortisone 0.1mg daily.

Correct dosage is monitored by serum electrolytes, blood pressure and patient well being.



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