GUIDELINES FOR THE SURGICAL MANAGEMENT OF ENDOCRINE DISEASE AND TRAINING REQUIREMENTS FOR ENDOCRINE SURGERY

THE BRITISH ASSOCIATION OF ENDOCRINE SURGEONS

CONTENTS

GUIDELINES

The British Association of Endocrine Surgeons (BAES) is the representative body of British surgeons who have a special interest in the surgery of the endocrine glands other than the pituitary. The Association is recognised by the Department of Health, The Association of Surgeons of Great Britain and Ireland and the British Association of Surgical Oncology. The BAES advises the SAC and the Intercollegiate Board on the Curriculum for Training in Endocrine Surgery.

In 1998, Members of the Association at the Annual Meeting mandated the Executive to form a working party for the purpose of drafting Guidelines for recommended practice in endocrine surgery. The general membership of the BAES had the opportunity to review and comment on the Guidelines which were first published in 1999.

The Guidelines represent core standards for clinical practice and it is expected that they will evolve and be subjected to regular revision. The BAES has contributed to the National Thyroid Cancer Guidelines Group which has produced an evidence based document on the surgical and non surgical management of thyroid cancer. In recognising that much of surgery is not based on randomised controlled trials, especially for rarer conditions such as endocrine disease, the guidelines are a consensus on what might be considered "best practice".

The Guidelines do not define an endocrine surgeon or specify who should practice endocrine surgery. There will be surgeons who, with the help of Guidelines and their existing clinical experience will be able to manage some thyroid and parathyroid diseases to the highest standard. Elective endocrine surgery may not be in the portfolio of every District Hospital but, where it is, based on experience and caseload, it should be in the hands of a nominated surgeon with an endocrine interest. Those patients requiring more complex investigation and care as detailed in these guidelines should be referred to an appropriate centre. These rare and complex diseases will only be managed effectively by multidisciplinary teams in Units familiar with these disorders. We consider that this category includes patients with endocrine pancreatic tumours, adrenal tumours, thyroid malignancy, familial syndromes and those requiring reoperative thyroid and parathyroid surgery.

Audit with outcome analysis is critical to the credibility of any surgical team. Such audit should produce data to allow meaningful comparison between units. Participation in the BAES Audit is a requirement for recognition as an Endocrine Unit and for endocrine training and is also a condition of membership of the BAES. It is intended that analysis of audit data will inform future modifications of these guidelines.

The BAES supports the principles of structured information for patients and documented informed consent and therefore examples of suitable forms are enclosed.

In future, all surgeons undertaking endocrine surgery should be able to demonstrate that they have been specifically trained in the subject. The current SAC requirements for endocrine surgical training are therefore appended to this document together with a curriculum for endocrine surgery previously developed and approved by the BAES.

These revised Guidelines for the Surgical Management of Endocrine Disease and the Training Requirements for Endocrine Surgery were produced by the BAES Guidelines and Training Sub-group; and revised in 2003, they will be further revised in 2006.

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SURGICAL TREATMENT OF DISEASES OF THE THYROID GLAND

2.1 MANAGEMENT OF PATIENTS WITH NODULAR GOITRE

Introduction

Nodular goitre is the commonest surgical endocrine disease and may be managed by the general as well as the specialist surgeon.

Clinical Assessment

Clinically identified thyroid nodular disease should be categorised as:

* Solitary nodule
* Dominant nodule within a multinodular goitre
* Multinodular goitre

The clinical assessment of all patients should assess the existence of obstructive problems or tracheal deviation, should elicit any features of malignant disease, should identify if there is a family history of thyroid disease and should determine whether there is a history of external beam radiotherapy to the thyroid, mantle or head and neck.

Investigations

These should include:

* Blood tests - thyroid function tests - TSH (Free T4/T3, if the TSH is abnormal) and Calcium/Albumin prior to any neck exploration.

* Thyroid Auto-antibody estimation may predict post lobectomy hypothryroidism and assist in the interpretation of thyroid function tests.

* Fine Needle Aspiration biopsy is indicated for all solitary nodules and for all dominant nodules in multi-nodular goitres. Cytology is unable to differentiate follicular adenomas from carcinomas and has difficulty in differentiating lymphoma from thyroiditis and anaplastic carcinoma. Ultrasound guided core biopsy may be of value when freehand biopsy is difficult.

* Ultrasound may differentiate solid from cystic nodules show solid components in a cystic nodule, identify multinodularity, identify associated lymphadenopathy BUT cannot reliably distinguish between benign and malignant lesions.

* Istotope Scan. This is only indicated in a patient with a nodule who is thyrotoxic and considered for surgery.

* CT/MRI/CXR These are indicated in patients with retrosternal goitre, for assessment of tracheal narrowing or deviation or, as part of preoperative assessment according to local protocols.

Other tests

* Pulmonary Function tests may assist the assessment of a patient with clinical evidence of airway embarrassment.

* Laryngoscopy. This is indicated where the patient has a history of voice change, previous thyroid surgery or is to be operated on for thyroid malignancy.

Indications for Surgery in Nodular Disease

* Cytological features of papillary or medullary cancer or a follicular lesion (adenoma or carcinoma)

* Clinical suspicion of malignancy

* Pressure signs or symptoms

* Patient preference – cosmesis/anxiety

* Toxicity (toxic adenoma or toxic multinodular goitre may be appropriate for radioactive iodine treatment)

* Progressive enlargement in a retrosternal goitre

* Discomfort in a goitre or associated with its presence

Preparation for Surgery

Patients undergoing surgery for thyroid nodular disease must be informed of the risks of thyroid surgery – see Section 6.1

Surgery for Benign Nodular Disease

Subtotal lobectomy for nodular goitre results in a 15% recurrence rate ultimately requiring re-operation. Recurrent nerve palsy rates are higher at re-operation.

Solitary Nodule

* Total lobectomy or isthmusectomy is recommended.

* No firm recommendation can be made on the value of frozen section for routine use in patients with benign cytology. Its use and value will be determined by local skills and protocols.

* The recurrent laryngeal nerve must be clearly identified and safeguarded in all patients. The superior laryngeal nerve should be safeguarded by careful ligation of the superior thyroid vessels at the margin of the capsule.

* Parathyroid glands should be identified and safeguarded. We recommend visualisation and preservation of all normal parathyroid glands.

* It is recommended that the branches of the inferior thyroid artery are ligated adjacent to the thyroid capsule to preserve the blood supply to the parathyroid glands in preference to continuity ligation of the trunk of the vessel laterally.

* There is no evidence that routine drainage is essential.

* Routine thromboembolic chemoprophylaxis is not recommended because of the risk of haematoma formation in the neck, but other DVT precautions are advised.

Multinodular Goitre

* Biochemical toxicity must be controlled prior to surgery.

* Unilateral surgery may be indicated when only one lobe is severely affected but, when there is significant involvement of both lobes, then total/near total thyroidectomy is recommended.

* Recurrent laryngeal nerves must be clearly identified and safeguarded. The superior laryngeal nerve on each side should be safeguarded by careful ligation of the superior thyroid vessels.

* Every effort must be made to preserve parathyroid glands.

* All parathyroid glands excised with the thyroid should be re-implanted, after dicing, into adjacent muscle. Details should be recorded in the operation notes.

* Retrosternal extension of a multinodular goitre should be sought. Most retrosternal goitres can be removed via a cervical incision.

* Risk factors that may identify patients who require sternotomy include – previous thyroid surgery, extension to a level below the aortic arch, significant posterior mediastinal extension.

Postoperative Management

* We recommend that serum calcium should be checked within the first 24 hours of surgery and subsequently if abnormal. Calcium replacement may be indicated when the corrected calcium falls below 2mM/1.
  Intravenous calcium gluconate is normally indicated for a corrected calcium below 1.8mM/1 or if the patient is symptomatic. Patients must not be discharged until the calcium level is stable without intravenous calcium administration in the preceding 24 hours.

* Facilities and a written protocol must be available for the management of airway obstruction or wound haemorrhage.

* Accurate assessment of vocal cord function can only be obtained by formal laryngoscopy in the conscious patient. It is best performed by a practitioner with expertise such as an ENT surgeon. It is not essential postoperatively in every case but is recommended in patients with persisting postoperative voice change.

* All patients undergoing thyroidectomy should have TSH levels checked at follow-up. Patients undergoing total thyroidectomy should commence Thyroxine prior to discharge from hospital.

2.2 THYROID MALIGNANCY

The evidence base relating to the management of thyroid carcinoma is predominantly in the form of retrospective reviews of single centres' management of the condition. There are no prospective randomised studies. However the following guidelines are consistently supported by the available evidence.

Surgery for thyroid cancer should be performed by endocrine surgeons or surgeons with an interest in endocrine surgery with appropriate training. This surgery should be carried out in a unit with access to appropriate cytology, pathology, endocrinology and oncology support.

PAPILLARY THYROID CANCER

Diagnosis

The diagnosis may be made or suspected on clinical grounds (usually presenting with a dominant nodule) on preoperative aspiration cytology or a previous operation (e.g. lymph node biopsy, thyroid operation or lung resection). It may be made on intraoperative frozen section or postoperative definitive histology.

Investigation

* Clinical examination of the neck is required to identify palpable abnormalities in the thyroid, adjacent structures and related lymph nodes.

* Fine needle aspiration biopsy is mandatory for dominant or isolated thyroid swellings. It should be performed at the first clinic visit.

* A chest radiograph is required.

* Routine preoperative imaging with scintigraphy or ultrasonography is not recommended, but imaging with CT or MRI may be indicated in patients with extensive or recurrent disease.

* Patients undergoing thyroid surgery should have thyroid function and serum calcium recorded prior to operation.

* Preoperative laryngoscopy is indicated in the presence of voice change, clinically suspected or proven malignant disease.

Preparation for Surgery

Patients undergoing surgery for papillary thyroid cancer must be informed of the risks of thyroid surgery and the increased risks associated with more radical surgery for malignant disease (viz hypoparathyroidism and recurrent laryngeal nerve injury). See Section 6.1.

Surgical Treatment

* The objective of surgery is to remove all macroscopic malignant disease in the thyroid, draining lymph nodes and involved adjacent structures.

*A tumour of 1 cm diameter or less with no palpable lymphadenopathy or clinically overt disease in the contralateral lobe or evidence of distant metastases can safely be managed by total lobectomy alone.

* Patients with macroscopic nodal disease and/or distant metastases, patients with clinical evidence of bilateral or multifocal unilateral disease should undergo total thyroidectomy.

* Patients treated by unilateral lobectomy who are shown on histology to have multifocal disease should be considered for complete thyroidectomy.

* Patients with macroscopic lymphadenopathy should be managed by lymph node excision which conserves non lymphatic structures. Radical forms of block dissection of the neck which sacrifice non lymphatic structures are not indicated unless those structures are infiltrated by tumour.

* There is controversy concerning the extent of thyroid surgery with solitary tumours greater than l cm in diameter without evidence of lymph node or distant metastases. The published evidence does not clarify whether lobectomy near total or total thyroidectomy is most likely to improve outcome.

* Extensive extra thyroidal disease in the neck is usually associated with a poor prognosis and surgery which is more radical than total thyroidectomy and lymph node dissection is not routinely recommended.

* Surgery should minimise the risk of avoidable injury to the recurrent and superior laryngeal nerves and attempt to conserve functioning parathyroid tissue.

Tumour Staging

We advise that pathological TNM staging is recorded for all patients with papillary thyroid cancer to facilitate comparative studies of treatment and outcome (local/regional/systemic relapse and death). Description of the resected tumour should be according to Royal College of Pathologists minimum data set.

Post operative management is as for benign disease, with the exception of thyroid replacement therapy.

* If the patient has undergone total thyroidectomy they are placed initially onto Tri-iodothyronine 20 µg tds and considered for radioactive iodine scanning and therapy. TSH suppression with T4 is recommended in the long term following surgery for papillary thyroid cancer. Patients who have undergone thyroidectomy for frankly invasive lesions will require surveillance in a specialist clinic with monitoring of TSH and thyroglobulin. Rising serum thyroglobulin levels are a marker of recurrent disease; levels should always be interpreted with care due to variation between laboratories and the confounding effects of anti-thyroglobulin antibodies.

* All patients diagnosed with papillary thyroid cancer should be reviewed by the Cancer Centre designated specialist multidisciplinary team.

Recurrent Disease

Recurrence may be detected clinically by detection of rising thyroglobulin levels or, on follow-up radioiodine scanning. Palpable disease should be assessed by Fine Needle Biopsy and CT/MRI/USS.

Options for the treatment of recurrent disease include further surgery, radioiodine or external radiotherapy. Management decisions in these circumstances should be made by a multidisciplinary team.

(See also the British Thyroid Association Guidelines on the treatment of Thyroid Cancer: http://www.british-thyroid-association.org/)

FOLLICULAR THYROID CANCER

Diagnosis

Most patients will be referred with a solitary thyroid nodule perhaps recently increasing in size. Pressure symptoms, hoarseness of voice and pain often radiating into the jaw and face are particularly relevant.

Investigation

* Fine needle aspiration biopsy is mandatory for dominant or isolated thyroid swellings. It should be performed at the first clinic visit, or by ultrasound guided technique.

* A chest radiograph is required.

* Routine preoperative imaging with scintigraphy or ultrasound is not recommended prior to a first operation, but imaging with CT or MRI may be indicated in patients with extensive or recurrent disease.

* Patients undergoing thyroid surgery should have thyroid function and serum calcium recorded prior to operation.

* Preoperative laryngoscopy is indicated in the presence of voice change, clinically suspected or proven malignant disease.

Preparation for Surgery

Patients undergoing surgery for follicular cancer must be informed of the risks of thyroid surgery and the increased risks associated with more radical surgery for malignant disease (viz hypoparathyroidism and recurrent laryngeal nerve injury). See Section 6.1.

Surgical Treatment

* All follicular neoplasms should be treated by surgical excision. i.e. thyroid lobectomy and isthmectomy.

* Definitive histology may indicate the need for completion thyroidectomy.

* Lesions which after adequate pathological evaluation are found to have no capsular or vascular invasion are not considered malignant and lobectomy is sufficient treatment.

* Total thyroidectomy is recommended where there is clear evidence of vascular or full thickness capsular invasion.

* Patients with lesions showing minimal capsular invasion alone may be treated by unilateral lobectomy.

* From the evidence available it is difficult to make firm recommendations for the treatment of follicular thyroid cancer with full thickness capsular invasion and no evidence of vascular invasion. Total thyroidectomy and radioiodine ablation allows post treatment surveillance with serial thyroglobulin measurements and identification/treatment of metastases with radioiodine.

* Hurthle cell tumours (Oxyphil) may behave more aggressively. Malignant Hurthle cell tumours are less likely to concentrate radioiodine and there is a case for treating them by total thyroidectomy.

Tumour Staging

We advise that pathological TNM staging is recorded for all patients with follicular thyroid cancer to facilitate comparative studies of treatment and outcome (local/regional/systemic relapse and death) – see page 6

Post-operative Management

As for papillary thyroid cancer, all patients diagnosed with follicular thyroid cancer should be reviewed by the Cancer Centre designated specialist multidisciplinary team.

MEDULLARY THYROID CANCER (MTC)

This is a rare tumour which can occur in four clinical settings:

* Sporadic
* Familial
  (a) as part of multiple endocrine neoplasia type IIa (b) as part of multiple endocrine neoplasia type IIb (c) as familial medullary thyroid cancer without other endocrine disease (FMTC)

Although appearing microscopically identical, the behaviour of the MTC varies according to the setting. The tumour is most indolent in FMTC and most aggressive in MEN IIb. The spectrum from indolent to aggressive is: FMTC * MEN IIa * Sporadic * MEN IIb. The familial forms are inherited autosomal dominant conditions – there is a 50/50 chance of any child of an affected parent inheriting the condition.

Surgery for medullar thyroid cancer should be performed by an endocrine surgeon in a unit with access to appropriate support in cytology, biochemistry, clinical and molecular genetics, endocrinology and oncology.

Diagnosis

The disease may present as a solitary thyroid nodule or as lymph node enlargement. Cytology is characteristic in MTC and should prompt a specific diagnosis. A positive family history of thyroid tumours will suggest a diagnosis of familial MTC.

Pre-operative Investigations in all cases

* The surgeon has the responsibility to distinguish between familial and sporadic disease and to exclude phaeochromocytoma in all cases.

* Whether the disease appears to be sporadic or familial, pre-operative or intra-operative staging should include:

* Basal calcitonin levels.

* Ultra-sonography of the neck to determine if there are multiple thyroid nodules or lymphadenopathy, or MRI/CT scans to assess the potential burden of cervical and/or mediastinal disease.

Investigations in Familial Cases

In the presence of a family history suggestive of MEN 2 or FMTC family genetic screening is required. It must be remembered that absence of family history does not preclude this patient being the index case for a new kindred.

Absence of biochemical expression (elevated calcitonin) in a relative of an index case after the age of 40 coupled with absence of a germ line mutation and the presence of a somatic mutation in the tumour suggests sporadic disease.

Molecular Genetics

Ninety-seven percent of individuals with MEN IIa have germ line mutations in the RET gene and 99% of those with MEN IIb have a RET germ line mutation in codon 918. Up to 80% of those with FMTC have these mutations. Within proven kindreds these molecular tests should, wherever and whenever possible, be used to diagnose disease in family members at risk (ante- and post-cedent). Children with these mutations should be offered prophylactic surgery. Provocative biochemical testing is not required in RET positive individuals but baseline calcitonin concentrations should be taken, though they may be normal in the presence of MTC.

The unit must be able to manage the logistics, ethics, clinical genetics and treatment of patients, including children, with inherited disease.

Preparation for Surgery

Preoperative laryngoscopy is indicated in the presence of voice change, clinically suspected or proven malignant disease.

Patients must be informed of the risks of thyroid surgery and the increased risk of more radical surgery – hypoparathyroidism and recurrent laryngeal nerve injury – see Section 6.1.

Surgical Treatment

The prime objective in surgery is to remove all malignancy in the thyroid, draining lymph nodes and adjacent structures. This usually requires total thyroidectomy and central compartment lymph-adenectomy. Patients with sporadic tumours may have a single focus of disease in one lobe but, because of its aggressive nature and multifocality, total thyroidectomy is still recommended.

Total thyroidectomy should include clearance of the central and paratracheal nodes from the level of the thyroid cartilage to the upper mediastinum and include a cervical thymectomy. The lateral nodes, if enlarged or suspicious around the jugular veins on each side, can be sampled and subjected to frozen section histology. If involvement is present, a modified neck dissection of the affected node groups should be performed at the time of the first operation sparing the jugular vein, the sternomastoid muscle and the accessory nerve.

Thyroidectomy should minimise the risk of avoidable injury to the recurrent laryngeal nerves and attempt to conserve functioning parathyroid tissue. If preoperative investigations have demonstrated associated hyperparathyroidism, enlarged glands should be removed at the time of thyroid exploration.

Prophylactic thyroidectomy is indicated in kindred members without clinically apparent disease but who are carriers of the germ line RET mutation. There is no current evidence on which to base a firm recommendation for the age at which prophylactic surgery should be performed but, on current knowledge, it should be between four and seven years of age. Emerging evidence suggests that certain RET mutations are associated with early and aggressive phenotypes of the disease requiring early operation (e.g. Men IIb) but others may have a less aggressive form and surgery can be postponed.

Postoperative management

Adequate replacement with thyroxine is required. Suppressive doses of thyroxine are not necessary and Thyroglobulin is not used as a marker in patients with this type of thyroid cancer. There is no role for postoperative iodine scanning in MTC. Calcitonin is used as a marker of recurrent or persistent disease. All patients diagnosed with medullary thyroid cancer should be reviewed by the Cancer Centre designated specialist multidisciplinary team.

Life long surveillance is required in a specialist endocrine clinic where biochemical and radiological expertise is available to detect and localise clinically apparent/occult disease. In patients with MEN II an annual test to exclude hyperparathyroidism and phaeochromocytoma should be performed.

Tumour staging

We advise that pathological TNM staging is recorded for all patients with medullary thyroid cancer to facilitate comparative studies of treatment and outcome (local/regional/systemic relapse and death) – see page 6.

Pathology

If MTC is diagnosed only after a hemithyroidectomy then completion thyroidectomy and central node clearance as described above should be undertaken.

Surgeons should seek advice from colleagues in clinical genetics about every case of diagnosed MTC [vide supra].

Familial disease is implied when there is multifocal, bilateral malignancy with or without a background of C-cell hyperplasia and in these cases germ line RET mutation analysis is mandatory.

Recurrent or Persistent Disease

When persisting or recurrent disease is suggested by rising levels of tumour markers, an attempt should be made to locate the disease by appropriate imaging which may involve Ultrasound, CT/MRI and scanning with octreotide, pentavalent DMSA or MIBG. The nature of palpable recurrences should be confirmed with FNAB. Accessible focal recurrences in the neck may be treated by excision but there is little evidence that radical re-operative surgery is beneficial. Patients with confirmed recurrent disease should be managed by a specialist multidisciplinary team so that all forms of palliative treatment can be considered. There may be a limited role for external beam radiotherapy or chemotherapy. There is no role for radioiodine therapy for patients with these tumours.

THYROID LYMPHOMA

Thyroid Lymphoma may present as a dominant nodule, a nodular goitre, or an 'inflammatory mass'. Surgery has little value in treatment.

Diagnosis

Thyroid function is usually normal but abnormal titres of thyroid auto-antibodies may be detected.

Fine Needle Aspiration cytology may be used to distinguish lymphoma from lymphocytic thyroiditis but usually the diagnosis is difficult, especially in distinguishing lymphoma from anaplastic carcinoma.

Core biopsy is necessary to allow immunohistochemical sub-typing of the lymphoma which will have implications for further treatment. It is recommended that this is undertaken in theatre or with image-guidance (USI-core). Open biopsy is rarely needed.

Treatment

All patients diagnosed with thyroid lymphoma should be reviewed by the Cancer Centre designated specialist multidisciplinary team.

When a diagnosis of lymphoma has been established by FNA or core biopsy, oral/parenteral steroids are of value to treat acute symptoms of airway compromise rapidly.

When the diagnosis has been made the patient must be referred promptly to an oncologist or haematologist for staging and definitive treatment with radiotherapy and/or chemotherapy.

2.3 ANAPLASTIC THYROID CANCER

This disease has a very poor prognosis and 85% of patients die within a year of diagnosis. Cure is exceptionally rare and the primary aim of treatment is local control.

Diagnosis

Core or open biopsy is the most appropriate investigation as FNAB will often be unhelpful. The particular object of the biopsy is to distinguish anaplastic carcinoma from medullary carcinoma and from lymphoma. Immuno-cytochemistry may be needed to achieve a diagnosis.

CT or MRI give excellent information on local extension of tumour.

Treatment

All patients diagnosed with anaplastic thyroid cancer should be reviewed by the Cancer Centre designated specialist multidisciplinary team.

There is no consistent evidence that radiotherapy or chemotherapy is useful, although radiotherapy may aid local control. Hyper-fractionated radiotherapy with doxyrubicin as a sensitiser may be more effective. Palliative resection of the tumour should be considered but is not normally feasible or useful unless the tumour is less than 5 cm in diameter. Tracheostomy should be avoided if possible, tracheal stents should be considered in patients with airway obstruction.

2.4 THYROTOXICOSIS

Surgery for thyrotoxicosis can be technically demanding and should be performed by endocrine surgeons with appropriate training and organisational support. Patients should be treated by a team which includes an endocrinologist and an anaesthetist with appropriate training and experience of thyroid anaesthesia.

A decision to recommend surgical management of thyrotoxicosis should be made jointly by a surgeon and an endocrinologist or physician experienced in the management of thyroid disease. The treatment options of anti-thyroid drugs, radioactive iodine ablation (RAI) and surgery must be discussed with patients, and informed consent taking account of patient preference must be obtained – see Section 6.2.

Diagnosis

The diagnosis is based on the clinical features of hyperthyroidism with additional specific features in Graves' disease. Serum levels of free T4 and TSH must be measured in all patients. The free T3 level is mandatory only if there is a suppressed TSH and normal or minimally elevated T4 level. Measurement of serum anti-body levels is recommended. When the possibility of toxic uninodular or multinodular goitre or toxic adenoma exists, an isotope scan to localise area(s) of hyperactivity is indicated. When the biochemical diagnosis is borderline, and in children, neonates and pregnant women, more detailed assessment by an endocrinologist is recommended.

Indications for Surgery

There are no fixed indications for surgery as the preferred treatment of thyrotoxicosis. Normally surgery is considered only after failed medical treatment in the young (particularly adolescents) and in women who wish to, or may become, pregnant within two years of radioiodine treatment. Surgery also has a role in the management of the large toxic goitre.

Preparation for Surgery

Patients undergoing surgery for thyrotoxicosis should be euthyroid or effectively 'beta blocked' to eliminate the risk of a thyrotoxic crisis. The following regimens can be combined if required.

* A titrated dose of antithyroid drugs to normalise serum levels of T3, T4 and TSH.

* A block and replace regimen of a large dose of anti-thyroid drug and replacement thyroxine.

* The effects of sympathetic over-activity may be blocked with an appropriate beta blocking drug.

* Oral iodine for ten days before surgery in Graves' disease has been shown to reduce blood flow in the thyroid and may be considered.

Preoperative discussion with the patient should outline the objectives of the operation taking into account family planning, social circumstances, compliance with medication, ophthalmopathy and patient preference. The likely effect of surgery and the requirement for postoperative thyroxine replacement and calcium/Vitamin D supplements must be explained.

Surgery

The surgery must be performed with a view to achieving the preoperative objective and minimising surgical morbidity (avoidable injury to the recurrent and superior laryngeal nerves and conservation of functioning parathyroid tissue). Total/sub-total thyroidectomy is recommended. Routine DVT chemoprophylaxis is not required, but mechanical DVT prevention should be used.

Postoperative Management

Facilities to deal with acute post-operative airway and metabolic problems must be available. The serum calcium should be checked after bilateral thyroid lobe resections. When pre-operative preparation includes beta blockade this must be continued for 7 days post-operatively.

Temporary early postoperative hypocalcaemia is common after surgery (up to 30% patients) for thyrotoxicosis. This must be identified but requires treatment only if severe. Serum calcium should be measured within the first 24 hours after surgery and regularly thereafter until normal levels are achieved by spontaneous recovery or institution of calcium and/or vitamin D therapy.

Following complete ablation of thyroid function, thyroxine replacement must be instituted before thyroid failure develops. The follow-up of patients after surgery to conserve function should continue until a stable euthyroid state is achieved. Such patients may develop late hypothyroidism, the likelihood and timing of this is not predictable. Long term monitoring of thyroid function is recommended in these patients.

Ophthalmopathy

Patients should be warned that surgery does not have a predictable effect on eye signs or prevent future development or progression. Total thyroidectomy may, in some patients, favourably influence the course of

ophthalmopathy. Progressive eye signs may require referral to a specialist surgeon for treatment.

Surgical Approach to the Thyroid Gland

The standard approach for the thyroid is via an open operation through a "collar" incision. Minimally invasive techniques to remove the thyroid gland are being developed in several centres globally, and when a surgeon has had specific training in this technique this may be appropriate. Patients should discuss options for the surgical approach with their surgeon and come to a joint decision on the final procedure taking into account available expertise and the underlying thyroid disease.

SURGICAL TREATMENT OF THE PARATHYROID GLANDS

3.1 PRIMARY HYPERPARATHYROIDISM

Although some patients are genuinely 'asymptomatic' at presentation, a full history will usually uncover symptoms that may be due to Primary Hyperparathyroidism (HPT). The surgeon has a responsibility to ensure that alternative causes of hypercalcaemia have been excluded.

A family history must be taken to identify familial hyperparathyroidism, multiple endocrine neoplasia types I and II or familial hypercalcaemic hypocalciuria (FHH).

Indications for Surgery

A liberal approach to surgery for patients with HPT is recommended as there are no clear means to predict which patients will deteriorate if left unoperated upon. There is equivalent benefit from surgery in patients with symptomatic and with 'asymptomatic' mild hypercalcaemia. Patients aged 70 or less have a higher mortality with untreated disease. The increased risk of death decreases after successful parathyroid surgery. Successful operation improves reported functional health status and quality of life. Cardiac changes have been shown to be reversible after treatment of asymptomatic disease. Symptoms of depression and anxiety may be reversed. Loss of bone mineral content is partially reversible.

Investigation

The biochemical diagnosis of HPT requires elevated levels of corrected serum calcium and an intact parathyroid hormone level that is raised or inappropriate for the calcium reading. Total urinary calcium or fractional urinary calcium excretion may be helpful in borderline cases. Normocalcaemia does not exclude hyperparathyroidism. Hypocalciuria (< 2 mmol/day) should prompt the diagnosis of FHH.

For an experienced surgeon, when full cervical exploration is performed, preoperative localisation of parathyroid glands does not improve the likelihood of cure at first time neck surgery. In this setting objective evidence of a benefit from routine scanning in terms of efficacy, safety, cost effectiveness and treatment has not been established. If preoperative localisation is considered necessary the investigations of choice are 99Tc Sestamibi scanning, and high-resolution ultrasonography, and this will be required if a "focused" approach and limited neck dissection is to be performed.

Surgery

The patient should receive an information sheet and consent form specific to surgery for HPT and should have the opportunity to discuss these with the operating surgeon prior to the operation – see Section 6.5.

Surgery will result in normocalcaemia in excess of 95% of patients with minimal morbidity. Unilateral neck exploration, whether or not aided by preoperative imaging studies and minimally invasive parathyroid surgery (video assisted or probe-guided unilateral approach) are feasible , and these techniques can produce equivalent results to bilateral neck dissection. Approximately 50% of patients will not be suitable for a focused approach because of discordant scans or coincidental thyroid disease.

In single gland disease the abnormal parathyroid gland should be removed. If multiple gland enlargement is found only enlarged glands should be removed. If 4 large glands are identified a remnant of one should remain in situ and should be marked in case subsequent re-exploration is necessary.

The success of parathyroidectomy may be confirmed in the majority of cases by the use of intra-operative PTH assay techniques. If excision has been effective, the PTH should fall to less than half the pre-operative value within fifteen minutes of excision, and should be in the normal range. False positive results can occur, but are rare.

Biopsy of normal glands is associated with increased incidence of postoperative hypocalcaemia. When abnormal parathyroid tissue cannot be identified, normal parathyroid glands should not be removed. Routine DVT chemoprophylaxis is not recommended but mechanical prevention is advised.

Patients with familial HPT should be treated in specialised centres because different strategies are required for MEN I, II and Familial HPT. In familial syndromes the surgeon must decide whether to perform a subtotal parathyroidectomy (leaving a remant in the neck) and cervical thymectomy, or a total parathyroidectomy with thymectomy and autotransplantation of parathyroid tissue.

Pathology

Intraoperative frozen section examination can be used to confirm the removal of parathyroid tissue but cannot be relied upon to distinguish between hyperplasia and adenoma. The use of paraffin-embedded sections of the parathyroid gland may similarly prove difficult to distinguish adenoma form hyperplasia.

Postoperative Management

Serum calcium should be checked within the first 24 hours after surgery and subsequently if abnormal.

Patients treated successfully for solitary adenoma should have a serum calcium checked at one year and may be

discharged. Long-term follow up is recommended for patients with multiple gland disease.

Recurrent or Persistent HPT

Reoperative parathyroid surgery requires a multi-disciplinary approach and should be managed by a specialist team in a centre with appropriate facilities. The management will normally include reconfirmation of the diagnosis including a review of the original histopathology material. Extensive attempts to localise the abnormal gland/s should be made with the object of achieving concordance in two or more different test modalities. The clinical indications for operation should be reappraised.

3.2 RENAL HYPERPARATHYROIDISM (SECONDARY OR TERTIARY HYPERPARATHYROIDISM)

Renal Hyperparathyroidism (RHPT) is one facet of a multisystem disease and patients should therefore only be accepted for surgery at the recommendation of a consultant Nephrologist or consultant Endocrinolgist. Patients should be treated within a multispecialty environment where continuing nephrological care and dialysis is available during the admission for surgery.

Indications for Surgery

The presence of RHPT is not of itself an indication for surgery. Surgery is normally only indicated in patients refractory to medical treatment who have very high levels of serum calcium or PTH or complications of RHPT, in particular bone disease, bone pain, soft tissue calcification, pruritus, unexplained myopathy or progressive vascular calcification. Aluminium toxicity must be excluded as a cause for symptoms.

The decision to refer for surgery will require a longitudinal review of symptoms, serum calcium, PTH and measures of bone metabolism, taken in the context of the patient's general condition and the effect of dialysis and medications on the biochemical results.

Routine imaging of the parathyroid glands is not indicated prior to a first operation.

Preoperative Preparation

Normocalcaemic patients should be pretreated with Vitamin D. During the admission for surgery the patient should be under the joint care of the surgical and nephrology teams. The anaesthetist should be experienced in the care of patients in renal failure. There should be explicit liaison concerning the timing of surgery in relation to concurrent nephrological care.

The patient should receive an information sheet and consent form specific to surgery for RHPT and should have the opportunity to discuss these with the operating surgeon prior to the operation – see Section 6.4 (page 22). Routine DVT chemoprophylaxis is not recommended.

Surgery

The operation should be conducted by a surgeon experienced in parathyroid surgery. The operation should attempt to identify all parathyroid tissue. This will normally involve the identification of at least four parathyroid glands and a search for ectopic parathyroid tissue in the neck. Routine cervical thymectomy is recommended.

Operative strategies include:

* Total parathyroidectomy with or without autografting of parathyroid tissue.

* Subtotal parathyroidectomy.

Each of these procedures can achieve the required biochemical and symptomatic benefits and all may be associated with recurrence. There is currently no consistent evidence to indicate that one strategy is preferable but total parathyroidectomy without reimplantation may be less satisfactory in patients expected to receive a renal transplant. The cryopreservation of parathyroid tissue for possible later implantation is an option in units with specific experience of this technique.

Postoperative Management

Postoperative hypocalcaemia may be more profound and symptomatic than after surgery for PHPT and adjustments may be needed to dialysis regime in response to this. The majority of patients will require vitamin D and Calcium supplementation.

Recurrent or Persistent Disease

Detailed attempts at imaging are imperative before any reoperation to establish whether the cause of excessive PTH is located in the neck, from an ectopic parathyroid or from hyperfunction of any autografted tissue. Reoperation **MUST** be carried out by an experienced endocrine surgeon.

Surgical approach

Open neck exploration of the neck is a well recognised approach with constantly good results in the hands of experienced endocrine surgeons.

A "focused" approach following imaging of the parathyroids my permit a smaller surgical scar and can be open or using minimally invasive techniques (MI). MI surgery should only be performed by those surgeons explicitly trained in the procedure, (and will probably be applicable in no more than 50% of patients).

4. SURGICAL TREATMENT OF DISEASES OF THE ADRENAL GLAND

Introduction

Adrenal pathology requiring surgery is rare. Disorders of adrenal function are often complex and the involvement of an experienced endocrine team supported by sophisticated

clinical biochemistry and radiology is essential to secure a reliable diagnosis. Successful surgical management is dependent on expert preoperative therapeutic preparation and skilled anaesthetic care, especially for patients with phaeochromocytoma. In practice the whole range of required expertise is only likely to be found in a Regional centre to which most patients should be referred.

The commonest diseases of the adrenal glands that require surgical management are:

* Hypercortisolism – ACTH independent – due to adrenocortical adenoma (Cushing's syndrome).
* Primary hyperaldosteronism due to adrenocortical adenoma (Conn's Syndrome).
* Phaechromocytoma.
* Incidentally discovered adrenal tumours, ('incidentaloma').
* Adrenocortical carcinoma.

The diagnosis of adrenal hyperplasia secondary to Cushing's disease or ectopic ACTH syndrome is not straightforward and such patients may require multiple investigations under the endocrine medical team. If pituitary surgery and bronchopulmonary surgery fail, bilateral adrenalectomy may be indicated.

There are other rare indications for surgical management of adrenal disease such as metastatic disease and tumours producing sex hormones but these are not addressed in these guidelines.

Investigations:

Cushing's Syndrome secondary to adrenocortical adenoma

These patients have the clinical stigmata of Cushing's syndrome and the following biochemical abnormalities: high levels of urinary free cortisol, loss of diurnal rhythm of plasma cortisol, an abnormal dexamethasone suppression test. The level of ACTH in the blood is suppressed. An adrenal mass is identified on a CT or MRI scan, 5 mm cuts are taken after enhancement with intravenous contrast. Typically the contralateral adrenal is suppressed, reflected in a smaller size on imaging.

Primary hyperaldosteronism – adrenocortical adenoma – Conn's Syndrome.

Investigation will classically reveal hypokalaemia, inappropriately raised urinary potassium, a high plasma aldosterone and a suppressed renin level. The aldosterone level does not suppress when the intake of sodium chloride is increased and it is unaffected by posture. Normal individuals typically increase their aldosterone levels markedly (3-4 times) when recumbency is followed by several hours of activity. Those with Conn's tumour show no change from this pattern whereas patients with idiopathic hyperaldosteronism – a bilateral nodular hyperplastic condition show a modest increase (30%). Serial 3 mm CT (or MRI) scans will identify the majority of Conn's tumours arising in the cortex of one adrenal. Attenuation is not improved by intravenous contrast. Occasionally adrenal scintiscanning or selective venous sampling is required to lateralise the disease, often with ACTH stimulation.

Phaeochromocytoma

Although the majority of patients have the classical triad of headaches, palpitations and hyperhidrosis with sporadic bouts of hypertension, fifteen percent of phaeochromo - cytomas are clinically silent and only declare themselves as an abdominal mass on routine scanning or in response to anaesthesia or surgery for an unrelated condition. In some there will be family history of phaeochromocytoma or Multiple Endocrine Neoplasia 2 or Von Hippel-Lindau disease. The absence of a family history does not preclude the patient being the index case of a new mutation.

In patients suspected of the diagnosis of phaeochromo-cytoma, 24 hour total urinary metanephrines and catecholamines are measured. Where the combined urinary catecholamines are measured and are raised, a phaeochromocytoma is confirmed in more than 95% of patients. Localising an extra adrenal parganglioma is best achieved with whole body scanning using MIBG labelled with 123I or 131I and MRI. MRI is better than CT at detecting extra adrenal paraganglioma and distinguishes phaeochromocytomas from the surrounding structures without the need for intravenous contrast.

Malignant change in a phaeochromocytoma is only certain where overt local invasion and distant metastases are observed. Neither of these may be evident at the time of investigation and surgery. Malignancy cannot be excluded on histology, long-term follow up is therefore advised.

Incidentaloma

This term includes all tumours which are a chance finding on abdominal scanning by whatever modality when adrenal disease was unsuspected. Approximately 1-3% of abdominal CT scans reveal such adrenal tumours. Half of them are metastases from known primaries and a third are cortical adenomas. The remainder comprise stromal tumours such as myelolipoma, haemangioma and lymphangioma.

A full clinical history may reveal previously unrecognised symptoms of adrenal over-activity. Even if there are no symptoms to indicate hyperfunction, a basic biochemical screen should be undertaken to exclude a functioning adrenal tumour.

Assessment of serum cortisol and potassium and urinary metanephrines and catecholamines is recommended. If a functioning lesion is discovered, surgical removal will usually be appropriate but incidentalomas less than 3 cm in diameter can safely be observed with repeat CT scanning to monitor any increase in size. Surgical removal is indicated if growth is rapid or if the diameter exceeds 3 cm. Needle biopsy is only indicated to confirm metastatic disease in a patient with a history of primary malignancy elsewhere but no other evidence of metastases.

Adrenocortical carcinoma

These tumours are typically bulky, greater that 1.5 cm in diameter and invade locally with early spread to the liver, regional lymph nodes and the lungs. Half the tumours are physiologically active with hypercortisolism and/or excessive androgen production. Weight loss and anaemia signal malignancy and an MRI scan provides the most complete assessment of operability. Measurement of dihydroepiandrosterone sulphate levels may assist in the diagnosis.

Preoperative Preparation

Adrenal surgery is a major procedure even when undertaken laparoscopically. Cardiac and respiratory function should be optimised. Anaemia and any electrolyte imbalance should be corrected and any intercurrent infection treated. Patients undergoing open or laparoscopic surgery should receive thromboprophylaxis.

Hypercortisolism:

Cushing's disease and Cushing's syndrome

Prophylactic antibiotics and thromboprophylaxis are particularly required for this group. Anticipating the dramatic fall of circulating cortisol after bilateral adrenalectomy and unilateral adrenalectomy where the contralateral adrenal is likely to be suppressed, 100 mg of parenteral hydrocortisone hemisuccinate is given in the anaesthetic room and again on completion of the operation.

Conn's Disease

Spironolactone 200-400 mg daily can be given for four weeks to stabilise hypertension and reverse hypokalaemia.

Phaeochromocytoma

Blockade of alpha-adrenergic receptors, (with doxazosin or phenoxybenzamine) is essential prior to surgery. The dosage and duration of treatment is highly individualised according to the severity of the disease and patients will sometimes require admission to stabilise hypertension and expand the intravascular volume. This is reflected in a fall in the haematocrit and an increase in the body weight. Patients invariably experience some postural hypotension with a dry mouth and a degree of sedation. Where tachycardia or arrhythmias develop a beta-blocker such as atenolol may be given but only when alpha blockade is complete.

The anaesthetist involved in the surgery of phaeochromocytoma **MUST** be familiar with the complexities of the anaesthesia for such patients. The anaesthetist should be closely involved in the preoperative preparation of the patient. The technical details of appropriate anaesthesia for these patients is outside the scope of these guidelines.

Surgery

Solitary tumours should normally be treated by total removal of the affected adrenal gland. Where the underlying pathology is the adrenal hyperplasia of Cushing's disease with a continuing source of over secretion of ACTH, all adrenal tissue must be removed. Subtotal excision of an adrenal gland or reimplantation of adrenal tissue is not of proven value and should only be undertaken as part of an ethically approved, scientific study.

Benign tumours in which there is no suspicion of malignancy can normally be removed laparoscopically by a transperitoneal or retroperitoneal route. The surgical approach to large tumours (>6 cm) will depend upon the experience of the surgeon, the patient's habitus, and the size of the tumour. There is no longer any requirement that phaeochromocytomas should be removed by the transperitoneal route to allow palpation of the contralateral adrenal bed. The superiority of one technique of adrenalectomy over the other has not yet been determined, but the laparoscopic approach is rapidly gaining popularity.

Postoperative Management

Patients undergoing bilateral adrenalectomy require lifelong adrenal replacement medication. In the immediate postoperative phase this will be by bolus iv administration of Hydrocortisone hemisuccinate repeated six hourly until the patient can be switched to oral replacement. This should be by a locally agreed protocol but as a guideline 20 mg of hydrocortisone is taken in the morning and 10 mg of hydrocortisone in the evening. The need for fludrocortisone is determined by the serum potassium level, doses range from 50-200 μg per day. It is vital that patients appreciate the importance of maintaining their replacement therapy and the need to increase the steroid dose if infection or stress arises.

All patients should be given a 'steroid card' indicating their steroid doses, hospital contact details and advice. Patients undergoing unilateral adrenalectomy for a cortisol secreting adenoma will also require cortisone postoperatively as the remaining adrenal will have been suppressed. The suppressed adrenal may require up to 18 months to recover and cortisone should not be discontinued until that recovery has been documented by a Synacthen test.

Occasionally aldosterone secreting tumours also secrete cortisol and where this is detected suppression of the contra lateral adrenal should be anticipated by giving cortisone replacement pre and post adrenalectomy as for patients with Cushing's syndrome.

5. SURGICAL TREATMENT OF THE ENDOCRINE PANCREAS

Pancreatic endocrine tumours (insulinoma and gastrinoma) are rare and require complex diagnostic and preoperative assessment. Surgical decision making and operative technique require specific experience of these conditions which should therefore be treated by a specialist endocrine team with the requisite experience. This will normally require referral to a regional centre.

5.1 INSULINOMA

Diagnosis

A full clinical assessment must be made in an endocrine clinic. Particular care must be taken to identify familial disease and exclude other endocrinopathies. Regional genetics units should be consulted in familial syndromes.

Biochemical

* The demonstration of a low fasting blood glucose and a high insulin.

* A 72 hour fast to provoke and unmask hypoglycaemia. A blood sugar below 2 mmo1/1 and an inappropriately raised insulin greater than 16 mU/1 is usually required.

* A high level of pro-insulin suggests malignancy.

* A high titre of insulin antibodies and low C peptide suggests a factitious hypoglycaemia due to exogenous insulin administration.

* A gut hormone profile, serum calcium, PTH and prolactin should be measured to identify other endocrine adenopathies.

Localisation

Precise localisation of an insulinoma is not essential before laparotomy is performed. A variety of localisation procedures are available which include endoscopic ultrasound, CT, MRI and octreotide scanning. Approximately 30% of insulinomas will be identified by Octreoscan. Endoscopic ultrasound is currently the most effective localisation procedure. Venous sampling and selective arterial calcium infusion stimulation testing is conducted in some centres.

Surgery

Once the diagnosis has been established almost all patients are submitted to surgery and pancreatic exploration even in the absence of preoperative localisation of the tumour. Only those patients unfit for surgery are treated with diazoxide or octreotide.

Even though only 10% of insulinomas are malignant, liver secondaries are excluded at laparotomy by palpation and intraoperative ultrasound. The pancreas is fully mobilised in order to permit careful palpation from head to tail. Intraoperative ultrasound should be used to localise the tumour and demonstrate its relationship to important vascular structures and the pancreatic duct. Most insulinomas are treated by enucleation but lesions in the tail of the pancreas are best treated by distal pancreatectomy. When the tumour is closely related to the pancreatic duct a distal pancreatectomy is a safer procedure. Lesions in the head of the pancreas should be enucleated and a Whipple's procedure is rarely necessary.

Frozen section histology is used to confirm the neuro-endocrine nature of the lesion. A spontaneous rise in blood glucose will demonstrate success but failure to note such a rise should not be taken to indicate failure. If no tumour is found at operation, blind distal pancreatectomy is NOT recommended.

Surgical Strategy for MEN 1

In this familial disorder the pancreatic lesions are multiple and require treatment by enucleation of those situated within the head of the organ and by a generous distal pancreatectomy as far as the portal vein.

Postoperative Management

The patient is best managed for the first few hours in a high dependency unit with regular monitoring of blood glucose.

Unresectable primary disease or hepatic metastases should be managed medically with octreotide, hepatic embolisation or resection.

5.2 GASTRINOMA
(ZOLLINGER-ELLISON SYNDROME)

Diagnosis

Full clinical assessment must be made in an endocrine clinic. Particular care must be taken to identify familial disease and exclude other endocrinopathies.

Biochemical

* High serum Gastrin when atrophic gastritis has been excluded by biopsy (Atrophic gastritis is the commonest cause of hypergastrinaemia).

* Secretin stimulation test (2U/kg body weight) is required to confirm the diagnosis of gastrinoma and distinguish it from antral G-cell hyperplasia.

* Measurement of gut hormone profile, calcium, PTH and prolactin to exclude MEN 1.

Localisation

Recommended tests include ultrasound, CT, MRI, octreotide scintigraphy and endoscopic ultrasound. Selective arterial calcium stimulation with venous sampling for gastrin (Imamura test) is a helpful method for regionalisation but not true localisation of gastrinoma.

Surgery

Patients with confirmed Zollinger-Ellison syndrome may have sporadic disease or MEN 1 syndrome. Both groups should be treated by active surgical intervention if hepatic metastatic disease has been excluded. Elective total gastrectomy or vagotomy is NOT indicated.

All patients should be treated with proton pump inhibitors up to the time of surgery.

Sporadic Gastrinoma

The primary tumours are frequently located outside of the pancreas, within the duodenal or even jejunal mucosa. The pancreas and duodenum should be fully mobilised, carefully palpated and also examined by intra operative ultrasound. In sporadic disease 30-40% of primary tumours will be within the duodenum and frequently accompanied by lymph node metastasis.

Identification of the duodenal wall lesion can be aided by endoscopic trans illumination as well as careful palpation, the latter being performed after a generous duodenotomy. These lesions are usually too small to be identified by ultrasound.

Once the primary lesion has been identified either within the gut or pancreas, it is excised and all adjacent lymph nodes meticulously cleared.

If a primary tumour has been located in sporadic Zollinger-Ellison syndrome, no other search for additional tumours is required. Frozen section histology will confirm the neuro-endocrine nature of the lesion.

MEN 1 Syndrome

HPT should be excluded and, if present, treated first. A complete pancreatic exploration for gastrinoma similar to that performed for sporadic disease is required and excision of any lesion within the wall of the gut, enucleation of lesions within the head of the pancreas, and a completion distal pancreatectomy performed.

Multiple neuro-endocrine tumours will inevitably be present and more than 60% will be within the wall of the duodenum. Duodenotomy is therefore mandatory. A Whipple's resection of the head of pancreas is rarely required. All draining nodes are resected and subjected to histology.

Postoperative Management

Initial recovery should be in a high dependency unit. The patient should remain on proton pump inhibitors as there is likely to be significant gastric hypertrophy and continuing hypersecretion of acid. Long term follow up and regular review in out patient clinic is required.

After three to four months the proton pump inhibitor should be stopped and the patient re-evaluated by measurement of fasting and secretin stimulated gastrin levels. In persistent or recurrent disease the patient should remain on proton pump inhibitors.

Unresectable primary disease or hepatic metastases should be managed medically with octreotide and proton pump inhibitors. Hepatic embolisation or resection is occasionally indicated. Chemotherapy is not recommended.

Operation in the presence of biochemical abnormality but no identified lesion on imaging is increasingly advised in confirmed MEN I. In this setting distal pancreatectomy and a search for lesions in the duodenum and head of the pancreas with enucleation as necessary is advised.

6. PATIENT INFORMATION SHEETS

*** 6.1 THYROIDECTOMY**
FOR BENIGN NODULAR DISEASE OF THE THYROID

*** 6.2 THYROTOXICOSIS**
(OVER ACTIVE THYROID)

*** 6.3 ADVICE TO PATIENTS LEAVING HOSPITAL AFTER**
ADMINISTRATION OF 131-IODINE

*** 6.4 RENAL HYPERPARATHYROIDISM (RHPT)**

*** 6.5 PRIMARY HYPERPARATHYROIDISM (HPT)**

INFORMATION SHEET FOR PATIENTS

6.1 THYROIDECTOMY FOR BENIGN NODULAR DISEASE OF THE THYROID

Introduction

This sheet is intended to tell you about the thyroid gland and give you information about the operation of thyroidectomy. Your surgeon will explain to you the reason for recommending surgery as treatment for your thyroid gland. The surgeon will also discuss whether in your case it is planned to remove all of the thyroid or only part of it.

The Thyroid Gland

The thyroid gland produces a chemical substance (a hormone) called thyroxine. This hormone circulates around the body in the blood and controls the speed at which the body's chemical processes work. The normal thyroid has considerable spare capacity for making thyroxine, and so normally removal of as much as half of the gland can be undertaken without any need to give thyroxine replacement in the form of daily tablets after the operation. If however the whole thyroid has been removed you will need to take thyroxine for the rest of your life.

Very close to the thyroid glands are four tiny glands called parathyroid glands, each not much bigger than a grain of rice. These produce a hormone, which controls the level of calcium in your body. The parathyroid glands are normally left in place when the thyroid gland is operated upon but their function may be affected by the operation on the thyroid; there is more information about this later in this document.

Surgery

The operation requires a general anaesthetic and a stay in hospital, which is normally between 2 and 4 days. Access to the thyroid obviously requires that the surgeon make an incision in the neck. This is made a couple of finger breadths above the top of the breastbone. It is made in a skin crease or following the "grain" of the skin. This collar incision is symmetrical even if the thyroid abnormality is only on one side. Most thyroidectomy incisions heal to produce a very satisfactory scar. At the end of the operation the surgeon may consider it appropriate to leave a small "drain" tube in the neck. This will normally be removed on the first or second day after surgery. In some thyroid operations it is necessary to remove some of the lymph glands from the neck. The absence of these glands does not normally produce any problems; if your surgeon expects to remove lymph glands it will have been discussed with you.

Possible complications

Most thyroid operations are straightforward and associated with few problems. However all operations carry risks which include postoperative infections (e.g. in the wound or chest), bleeding in the wound and miscellaneous problems due to the anaesthesia, but these are very rare. Bleeding in the wound can be a serious problem if it occurs but the chance of a significant bleed needing you to return to the operating theatre within a day or two after your operation to clear out the blood is small (in the region of 1`in 50).

Scar:

The scar may become relatively thick and red for a few months after the operation before fading to a thin white line. Very rarely some patients develop a thick exaggerated scar but this is uncommon.

Voice Change:

It is impossible to operate on the neck without producing some change in the voice; fortunately this is not normally detectable. A specific problem related to thyroid surgery is injury to one or both of the Recurrent Laryngeal Nerves. These nerves pass close to the thyroid gland and control movement of the vocal cords. Injury to these nerves causes hoarseness and weakness of the voice. The nerve may not work properly after thyroid surgery due to bruising of the nerve but if this should occur, it recovers over a few weeks or months. Rarely, the nerve may be permanently injured and the nerve function will not recover. The External Laryngeal Nerve may also be injured and this results in a weakness in the voice although the sound of the voice is unchanged. Difficulty may be found in reaching the high notes when singing, the voice may tire more easily and the power of the shout reduced. Careful surgery reduces the risk of permanent accidental injury to a very low level but cannot absolutely eliminate it. Injury to both recurrent laryngeal nerves is extremely rare but is a serious problem and may require a tracheostomy (tube placed through the neck into the windpipe).

Low blood calcium levels:

Patients undergoing surgery to the thyroid gland are at risk of developing a low calcium level if the four tiny parathyroid glands which control the level of calcium in the blood stop working after the operation. It is normally possible to identify and preserve some if not all of these glands and so avoid a long-term problem. Unfortunately even when the glands have been found and kept they may not function. If this happens then you will require to take extra calcium and/or vitamin D on a permanent basis. The risk of you needing long term medication because of a low calcium level is small (about 1 in 50). It is quite common to require calcium and/or vitamin D tablets for a few weeks or months after the operation.

Thyroid function:

If it has been decided to remove all the thyroid gland then you will require lifelong replacement of thyroxine. Fortunately this is a straightforward once a day regimen with little requirement for adjusting dosage. There is a prescription charge exemption for patients requiring thyroxine tablets so you will not have to pay for these (or any other tablets as the law currently stands). If most, but

not all, of the thyroid gland is removed then in the early weeks after the operation the remaining thyroid may not produce enough thyroxine and you may require replacement tablets temporarily until the retained thyroid produces enough hormone itself. This will be monitored.

Swallowing difficulty:
Usually swallowing is improved following thyroid surgery, especially for large goitres or those which have extended down into the chest, but occasionally some mild difficulty may develop or be persistent. Similarly, if you are experiencing any difficulty with your breathing before the operation, then this may also be eased.

We wish to emphasise that these potential side effects and complications are unusual, but we believe it is essential to tell you about these rather than have you develop a complication without having been forewarned. If you are unclear about the topics in this sheet or if you are unclear about any other details of your operation please ask one of the surgical team.

I confirm that I have read the above and have discussed any queries with the surgical team.

Name .....
Signature .....
Date .....

Note: If you search the Internet for information on this subject you should remember that some sites will describe calcium levels using different units of measurement. Additionally many sites are in effect advertising for patients and may propose untried or non standard procedures and treatments, so beware and discuss what you read with your doctors.

INFORMATION SHEET FOR PATIENTS

6.2 THYROTOXICOSIS (OVER ACTIVE THYROID)

Investigations have shown you to have an overactive thyroid gland. Three treatment options are available for this:-

* Antithyroid Drugs

* Radioiodine

* Surgery.

Most patients will need antithyroid drugs initially to control the thyroid overactivity. These drugs may be continued for around 18 months to allow the thyroid to recover spontaneously but in many cases such recovery will not occur and a more permanent treatment will be needed using radio-iodine or surgery. Often the illness will recur even after a full length course of antithyroid drugs and in these circumstances treatment with radioiodine or surgery will normally be advised.

Antithyroid drugs:
These are relatively slow acting and several months after withdrawal of treatment a significant number of patients will experience recurrence of the problem. There may be side effects associated with these drugs which may require them to be stopped.

Radioactive Iodine:
This is probably the simplest option for treating most overactive thyroid glands. It requires swallowing a capsule containing radioactive iodine which is taken up by the thyroid gland. The radioactivity destroys the thyroid cells that are causing the overactivity. Most patients are successfully treated with only one course of treatment but some require more than one. The major side effects of this treatment is that you have to keep a distance away from other people, particularly pregnant women and young children for several days until the level of radioactivity drops. This usually takes 2 to 3 weeks. (See advice sheet below).
Radioactive iodine therapy is not recommended for young women who are or wish to become pregnant within the next few months. You must not take radioactive iodine if you are currently pregnant or breast feeding. There is no evidence of long-term harm from taking radioactive iodine although most patients who take it will develop an under active thyroid sooner or later and require to take thyroid tablets.

Surgery:
Surgery is a suitable option for many patients with an overactive thyroid gland when antithyroid drugs do not work and a decision must be made between an operation and radioactive iodine. The advantages of an operation are that it removes an enlarged gland (goitre) and eases any symptoms caused by pressure of the gland on the other neck structures. It rapidly cures the overactivity and can restore normal function to patients for many years. However a substantial number of patients will have an under active thyroid after the operation and other patients will become under active with each passing year. For this reason it is sometimes recommended that all or virtually all the thyroid is removed and replacement thyroid hormone is started just after the operation. If some of the thyroid gland is left behind, then this can occasionally become overactive again in the future. Removing the whole of the gland has the great advantage of solving the problem once and for all with no risk of recurrence of the overactivity and no requirement for careful follow-up provided the thyroid tablets are taken once a day for the rest of your life.

Preparation for surgery:
When a decision has been made to proceed with surgery it is important that the effects of thyroid over activity are controlled before hand. This requires you to take the prescribed medication up to the time of the operation. If you have been prescribed a beta blocker (e.g. propranolol or nadolol) you may need to continue to take these for a few days after the operation. Your doctors will advise you if this is necessary.

Possible complications:
Most thyroid operations are straightforward and associated with few problems. However all operations carry risks which include postoperative infections (e.g. in the wound or chest), bleeding in the wound and miscellaneous problems due to the anaesthesia, but these are very rare. Bleeding in the wound can be a serious problem if it occurs but the chance of a significant bleed needing you to return to the operating theatre within a day or two after your operation to clear out the blood is small (in the region of 1`in 50).

Scar:
The scar may become relatively thick and red for a few months after the operation before fading to a thin white line. Very rarely some patients develop a thick exaggerated scar but this is uncommon.

Voice Change:
It is impossible to operate on the neck without producing some change in the voice; fortunately this is not normally detectable. A specific problem related to thyroid surgery is injury to one or both of the recurrent laryngeal nerves. These nerves pass close to the thyroid gland and control movement of the vocal cords. Injury to these nerves causes hoarseness and weakness of the voice. It is uncommon for the nerve not to work properly after thyroid surgery due to bruising of the nerve but if this should occur, it recovers over a few weeks or months. The external laryngeal nerve may also be injured and this results in a weakness in the voice although the sound of the voice is unchanged. Difficulty may be found in reaching the high notes when

singing, the voice may tire more easily and the power of the shout reduced. Careful surgery reduces the risk of permanent accidental injury to a very low level but cannot absolutely eliminate it. Injury to both recurrent laryngeal nerves is extremely rare but is a serious problem and may require a tracheostomy (tube placed through the neck into the windpipe).

Low blood calcium levels:
Patients undergoing surgery to the thyroid gland are at risk of developing a low calcium level if the four tiny parathyroid glands which control the level of calcium in the blood stop working after the operation. It is normally possible to identify and preserve some if not all of these glands and so avoid a long-term problem. Unfortunately even when the glands have been found and kept they may not function. If this happens then you will require to take extra calcium and/or vitamin D on a permanent basis. The risk of you needing long term medication because of a low calcium level is small (about 1 in 50). It is quite common to require calcium and/or vitamin D tablets for a few weeks or months after the operation.

Thyroid function:
If it has been decided to remove all the thyroid gland then you will require lifelong replacement of thyroxine. Fortunately this is a straightforward once a day regimen with little requirement for adjusting dosage. There is a prescription charge exemption for patients requiring thyroxine tablets so you will not have to pay for these (or any other tablets as the law currently stands). If most, but not all, of the thyroid gland is removed then in the early weeks after the operation the remaining thyroid may not produce enough thyroxine and you may require replacement tablets temporarily until the retained thyroid produces enough hormone itself. This will be monitored.

Swallowing difficulty:
Usually swallowing is improved following thyroid surgery, especially for large goitres or those which have extended down into the chest, but occasionally some mild difficulty may develop or be persistent. Similarly, if you are experiencing any difficulty with your breathing before the operation, then this may also be eased.

Pregnancy:
Once the thyroid hormone levels are normal after surgery, there is no reason not to become pregnant if desired. If you become pregnant during treatment with antithyroid medications such as carbimazole or propylthiouracil, it is not a major problem, but it is important to notify your hospital doctor as soon as possible.

We wish to emphasise that these potential side effects and complications are unusual, but we believe it is essential to tell you about these rather than have you develop a complication without having been forewarned. If you are unclear about the topics in this sheet or if you are unclear about any other details of your operation please ask one of the surgical team.

I confirm that I have read the above and have discussed any queries with the surgical team.

Name .....
Signature .....
Date .....

PATIENT INFORMATION SHEET

6.3 ADVICE TO PATIENTS LEAVING HOSPITAL AFTER ADMINISTRATION OF 131-IODINE

These notes are to help you understand some of the restrictions that you will need to follow after receiving a radioactive treatment for an over active thyroid.

The radioactive treatment is prescribed for you and only you will benefit from the treatment. The following instructions are to be adhered to in order to limit the amount of radiation to other people. Remember they have not been prescribed the radiation and will receive no benefit from it.

Make sure you understand the restrictions. There will be opportunities while at the hospital to ask any questions you may have.

The Card:
You will be given a card when the treatment is administered. The card lists the restrictions and the number of days for which the restrictions will apply.

With regards to children and pregnant women the card says:-

(a) Refrain from all close contact
This means – cuddling is not permitted. You should generally keep a minimum distance of 1 metre (3 feet) away from a child (except for occasional very brief periods). This degree of restriction should last for either 9 or 15 days, depending on the activity given.

(b) Refrain from extended periods of close contact
This means – limited contact is permitted, but you should not cuddle a child for more than 15 minutes per day and should not sleep with a child or pregnant woman in the same bed. This degree of restriction should last for either 21 – 25 days depending on the activity given. Normal contact can be resumed after this time.

Remember – occasional brief periods of contact with children or pregnant women will not be harmful, but you should restrict your contact by observing the two restrictions above.

In cases where you have sole responsibility for the care of a child, it should normally be recommended that alternative arrangements be made for the childs care for up to 25 days. This should be discussed with the doctor at your appointment at the hospital, before the treatment.

Travel – On leaving the hospital after the treatment there are no restrictions on the use of seated private or public transport provided the journey is less than 3 hours.

Spills – Please contact the hospital Nuclear Medicine Department if you should vomit or be incontinent of urine with 24 hours of the treatment.

Returning to work – Return to work on the day following treatment. This advice may be different for people who work close to others. If you work, please discuss your work with hospital staff.

Partners – Providing your partner is aware of the radiation and is not pregnant, you may sleep in the same bed. However there is a restriction if you have received the larger activity treatment. Then you should have separate beds for 4 days.

PATIENT INFORMATION SHEET

6.4 RENAL HYPERPARATHYROIDISM (RHPT)

The parathyroid glands are small glands usually situated behind the thyroid gland at the base of neck and which produce parathyroid hormone (PTH) which controls the level of calcium in the blood. The control of blood calcium levels is important for the proper functioning of the brain and nervous tissue and the maintenance of healthy bones.
There are normally four parathyroid glands which are usually situated close to the thyroid gland. Their function is completely unrelated to that of the thyroid.

In some patients with kidney disease a complex sequence of events can cause the parathyroid glands to become overactive and produce too much hormone. This leads to an excess of calcium in the blood, most of which is drawn from the bones which are thereby damaged. Some of this excess calcium may be deposited in other structures such as blood vessel walls where it may cause damage. This sequence of events is known as renal hyperparathyroidism (RHPT). RHPT may persist even when the original damage to the kidney as been corrected by dialysis or by transplantation.

The treatment of RHPT is by medication in the first instance, but this is not always effective and sometimes it becomes necessary surgically to remove all or most of the parathyroid glands. The operation is known as parathyroidectomy.

PARATHYROIDECTOMY

The technical details of the operation will be explained to you by your surgeon. At the operation the surgeon will try to locate all your parathyroid glands and will remove all or most of them. If all are removed part of one may be implanted into a forearm muscle. This procedure is undertaken so that if the overactivity of the parathyroid tissue recurs a further operation maybe necessary and it may be easier to remove tissue from the arm than from the neck.

The Operation

Operations on the parathyroid glands are very safe but sometimes there are complications and this section summarises these for you. If having read it there is anything that worries you and about which you would like further information please ask the surgical team.

Scar:
The scar may become relatively thick and red for a few months after the operation before fading to a thin white line. Very rarely some patients develop a thick exaggerated scar but this is uncommon.

Voice Change:
It is virtually impossible to operate on the neck without producing some change in the voice; fortunately this is not normally detectable. A specific problem related to parathyroid surgery is injury to one or both of the recurrent laryngeal nerves. These nerves pass close to the thyroid gland and control movement of the vocal cords. Injury to these nerves causes hoarseness and weakness of the voice. The nerve may not work properly after parathyroid surgery due to bruising of the nerve but if this should occur, it recovers over a few weeks or months. Rarely, the nerve may be permanently injured and the function will not recover. The external laryngeal nerve may also be injured and this results in a weakness in the voice although the sound of the voice is unchanged. Difficulty may be found in reaching the high notes when singing, the voice may tire more easily and the power of the shout reduced. Careful surgery reduces the risk of permanent accidental injury to a very low level but cannot absolutely eliminate it. Injury to both recurrent laryngeal nerves is extremely rare but is a serious problem and may require a tracheostomy (tube placed through the neck into the windpipe).

Other hazards:
Most parathyroid operations are straightforward and associated with few problems. However all operations carry risks which include postoperative infections (e.g. in the wound or chest), bleeding in the wound and miscellaneous problems due to the anaesthesia, but these are very rare. Bleeding in the wound can be a serious problem if it occurs but the chance of a significant bleed needing you to return to the operating theatre within a day or two after your operation to clear out the blood is small (in the region of 1`in 50).

Results of surgery

Operations to correct RHPT are intended to remove or reduce symptoms which you are having which may be attributable to the RHPT. You should discuss with your surgeon or renal physician the reasons why you have been referred for surgery and their expectations of improvement. Parathyroidectomy may be effective at controlling the RPHT and reducing future damage but may not necessarily improve your current symptoms or restore damaged bones, nor can it be expected to alter your need for renal replacement therapies such as dialysis. In most people the stimulus to parathyroid overactivity remains even after successful surgery, but there is always a risk that parathyroid tissue which has been left in your body may enlarge and lead to recurrence of your symptoms. About one patient in ten undergoing parathyroidectomy for RPHT will require a further operation on the parathyroid glands in the future. In other patients parathyroidectomy may remove all functioning parathyroid tissue and you may require continuing medical treatment to sustain a normal blood calcium level. It is not possible to predict in advance what the long term result of parthyroidectomy will be on your blood calcium levels, but in the majority of patients the operation will be beneficial.

Note: If you search the Internet for information on this subject you should remember that some sites will describe calcium levels using different units of measurement. Additionally many sites are in effect advertising for patients and may propose untried or non standard procedures and treatments, so beware and discuss what you read with your doctors.

I confirm that I have read the above and have discussed any queries with the surgical team.

Name .....
Signature .....
Date .....

PATIENT INFORMATION SHEET

6.5 PRIMARY HYPERPARATHYROIDISM (HPT)

The parathyroid glands are small glands usually situated behind the thyroid gland at the base of the neck and which produce parathyroid hormone (PTH) which controls the level of calcium in the blood. There are normally four parathyroid glands; although they are situated close to the thyroid gland their function is completely unrelated to that of the thyroid. The control of blood calcium levels is important for the proper functioning of the brain and nervous tissue and the maintenance of healthy bones. Too much PTH leads to too high a level of calcium in the blood and this can have various damaging effects, especially on the skeleton and kidneys. One of the surgical team will have discussed with you why it is necessary to remove one or more of your parathyroid glands to control the overproduction of PTH. Usually there is only one abnormal gland but there may be two or more affected glands. Parathyroid cancer is very rare and the abnormality in the parathyroid glands is nearly always benign.

The Operation

During your operation the surgeon will attempt to find and remove the abnormal parathyroid gland or glands and identify, inspect and possibly take samples from the other parathyroid glands. Abnormal glands may not be found at the time of operation in about one patient in twenty. One reason is that the gland may be so small or hidden that it cannot be found or seen. This happens very rarely. Another reason may be that the gland lies not in the neck but in the chest. If that is the case you may need another operation at another time to remove it.

Operations on the parathyroid glands are very safe but sometimes there are complications and this section summarises these for you.

Scar:

The scar may become relatively thick and red for a few months after the operation before fading to a thin white line. Very rarely some patients develop a thick exaggerated scar but this is uncommon.

Voice Change:

It is virtually impossible to operate on the neck without producing some change in the voice; fortunately this is not normally detectable. A specific problem related to parathyroid surgery is injury to one or both of the recurrent laryngeal nerves. These nerves pass close to the thyroid gland and control movement of the vocal cords. Injury to these nerves causes hoarseness and weakness of the voice. The nerve may not work properly after parathyroid surgery due to bruising of the nerve but if this should occur, it recovers over a few weeks or months. Rarely, the nerve may be permanently injured and the function will not recover. The external laryngeal nerve may also be injured and this results in a weakness in the voice although the sound of the voice is unchanged. Difficulty may be found in reaching the high notes when singing, the voice may tire more easily and the power of the shout reduced. Careful surgery reduces the risk of permanent accidental injury to a very low level but cannot absolutely eliminate it. Injury to both recurrent laryngeal nerves is extremely rare but is a serious problem and may require a tracheostomy (tube placed through the neck into the windpipe).

Other hazards:

Most parathyroid operations are straightforward and associated with few problems. However all operations carry risks which include postoperative infections (e.g. in the wound or chest), bleeding in the wound and miscellaneous problems due to the anaesthesia, but these are very rare. Bleeding in the wound can be a serious problem if it occurs but the chance of a significant bleed needing you to return to the operating theatre within a day or two after your operation to clear out the blood is small (in the region of 1`in 50).

The Results of Surgery

From the situation of having too high a level of calcium before the operation, the calcium often falls to quite a low level shortly after the operation. This is because the other glands have become lazy or under active. The other glands will recover their normal function quite quickly but you may require calcium and vitamin D tablets temporarily after the operation to boost the level of calcium in the body. Rarely, the other glands may not recover and you will need to take calcium and/or vitamin D tablets.

Operations to correct HPT are intended to remove or reduce symptoms which you are having which may be attributable to the HPT. You should discuss with your surgeon or medical doctor the reasons why you have been referred for surgery and their expectations of improvement. Parathyroidectomy may be effective at controlling the HPT and reducing future damage but may not necessarily improve your current symptoms.

I confirm that I have read the above and have discussed any queries with the surgical team.

Name .....
Signed .....
Date .....

Note: If you search the Internet for information on this subject you should remember that some sites will describe calcium levels using different units of measurement. Additionally many sites are in effect advertising for patients and may propose untried or non standard procedures and treatments, so beware and discuss what you read with your doctors.

APPENDIX 1

TRAINING REQUIREMENTS

AND

CURRICULUM

FOR ENDOCRINE SURGERY

REQUIREMENTS FOR TRAINING IN ENDOCRINE SURGERY

LENGTH OF TRAINING

One year in a BAES recognised HST Training Unit. Because of the rarity of some endocrine tumours it may be advisable that trainees spend more than one year in such a unit if this is practical or possible. Flexible rotations between regions may be required.

LOCATION OF TRAINING

Training should be undertaken in Units consisting of one or more surgeons approved as endocrine surgical trainers by the BAES and the SAC in General Surgery.
Approved endocrine surgical training Units will normally consist of:

* One or more surgeons with a declared interest in endocrine surgery.
* An annual operative throughput in excess of 50 cases
* On site cytology and histopathology services
* At least one consultant endocrinologist on site holding one or more dedicated endocrinology clinics per week.
* A Department of Nuclear Medicine on site.
* On site MR and CT scanning facilities.

It is accepted that candidates may need to have access to other units to ensure some experience of specialist techniques such as laparoscopic adrenal surgery.

THE SYLLABUS

The BAES believes that in addition to proper knowledge and experience of the principles and practice of general surgery candidates offering a special interest in endocrine surgery:

* Must have a firm grounding in the basic and clinical science aspects of the organs and diseases with which they should be familiar. A syllabus for this appears below.

* Must have an adequate operative experience in endocrine surgery. The BAES do not believe it is necessary to stipulate exact numbers of cases that must appear in the trainees log book expecting an appropriate practical experience to flow from attachment to approved endocrine surgical trainers/units as defined above. It is however expected that the number of index cases specified below (page 28) would be achieved.

* Should have an appreciation of relevant current research based on attendance at least one national or international endocrine surgical meeting per year.

BASIC SCIENCE CURRICULUM

GENERAL

An understanding of:

* The development of the endocrine glands and a detailed knowledge of their anatomy including common variations in position.
* Endocrine physiology as outlined below.
* The essentials of current techniques of radioimmunoassay of hormones and of radiographic and isotopic techniques for localising abnormal endocrine tissue.
* A basic knowledge of the genetics of inherited endocrine disorders.

SPECIFIC GLANDS

Thyroid

The role of Iodine in the normal function of the thyroid including pathways of iodine metabolism. The physiology of TSH and Thyrotrophin Releasing Factor. The functions of T3 and T4. Role of thyroglobulin in thyroid physiology. Thyroid hormone release. The principles underlying the functioning of the pituitary thyroid axis. Tests of thyroid function. Thyroid antibodies and their significance. An understanding of the physiological impact of Graves Disease on normal bodily functions. Mechanisms of ophthalmic manifestations of Graves Disease.. Calcitonin and its significance within the body.

Parathyroid glands

An understanding of the metabolism of calcium. The structure of parathyroid hormone. The control of calcium metabolism. The activity of PTH on kidney, gut and bone. D Vitamins and their function. The measurement of PTH.

The pituitary

Structure. Cells of origin, the basic metabolism and function of anterior pituitary hormones without feedback loops (growth hormone and prolactin) and those with feedback loops (FSH, LH, TSH and ACTH). Hypothalamic pituitary pathways and related releasing substances. Corticotrophin releasing factor and its relationship to ACTH. Physiology of ACTH including diurnal variation. ACTH changes in response to stress, illness and trauma.

Adrenal cortex

The biosynthesis of glucocorticoids. Physiology of glucocorticoids. Metabolism of cortisol and a knowledge of those metabolites which are measured in clinical practice. The physiology of adrenal androgens and the effects of pathological overproduction. The mineral-ocortcoids. The physiology of aldosterone. An understanding of the renin angiotensin mechanisms. A knowledge of tests of adrenal cortical function.

Adrenal Medulla

Metabolic pathways of Adrenaline and Noradrenaline production. The assessment of adrenal medullar activity. An understanding of the pathophysiology of excess catecholamines.

The gut as an endocrine organ

An appreciation of the physiology of gastrin, insulin, glucagon, pancreatic polypeptide, VIP, secretin and somatostatin.

CLINICAL CURRICULUM

An understanding of the principles of endocrine investigation, biochemical, radiological, isotopic, cytological and histological. Including strategies for minimising intervention and cost. Limitations of investigation. An understanding of the concepts of sensitivity, specificity etc.

FOR SPECIFIC CONDITIONS:

Knowledge of pathophysiology and pathology.
Outline of medical investigations.
Details of imaging.
Methods of biopsy.
Interpretation of histological and cytological specimens
Treatment options. Surgical vs. other.
Preoperative preparation
Anaesthetic and pharmacological peculiarities.
Operative techniques and strategies.
Postoperative care including substitution therapies.
Additional and adjuvant treatment.
Prognosis and strategies for follow up.
Counselling and screening in familial diseases.

SPECIFIC TOPICS

Pituitary.Cushing's including different options for managing microadenoma

Thyroid. Thyrotoxicosis including:
Aetiology
Medical and radioisotope therapy
Management of eye disease
Surgical strategies
Organisation of follow up
Monitoring of replacement therapy
Thyrotoxicosis in Pregnancy
Thyrotoxicosis in Childhood

Solitary nodule
Multinodular Goitre
Retrosternal goitre
Thyroid malignancy of ALL types
Medullary carcinoma and MEN type 2
Ectopic thyroid
Thyroiditis

Technique of thyroidectomy

Techniques and use of block dissections of neck.
Complications of thyroidectomy.
Management of RLN palsy.
Control of suppressive thyroxine therapy.
Role of Radiation therapy in thyroid malignancy.

Parathyroid

Primary, secondary and tertiary HPT.
Investigation of hypercalcaemia.
Management of acute hypercalcaemia.
Strategies in parathyroidectomy.
Technique of parathyroidectomy.
Management of HPT in renal patients.
Management of familial HPT.
Management of HPT in MEN patients.
Management of recurrent and persistent HPT.
Parathyroid autotransplantation.
Parathyroid carcinoma.
Management of hypocalcaemia.

Other Neck Pathology

Familiarity with parotid and submandibular salivary gland disease, Thyroglossal cysts, Branchial Cysts and the causes and management of cervical lymphadenopathy.

Adrenal

Cushing's Disease and Syndrome:
Investigation.
Indications for adrenalectomy.
Nelson's syndrome.
Conn's syndrome.
Incidentalomas.
Phaeochromocytoma.
Pharmocological and anaesthetic management of phaeochromocytoma.
MEN type 2
Other adrenal endocrine tumours
Adrenal malignancy
Strategies and routes for adrenalectomy
Replacement therapy

Pancreas and gut.

Insulinomas
Gastrinomas and ZE Syndrome
MEN 1
Carcinoid syndrome
Other endocrine tumours of gut.

INDEX CASES

	Performed	Assisted
Thyroid Lobectomy	20	30
Parathyroid	10	20

TO BE A GENERAL SURGEON WITH AN INTEREST IN ENDOCRINE SURGERY

The trainee must be competent to perform:

Thyroid Lobectomy/isthmectomy.

Sub-total thyroidectomy (including Graves Disease).

Total Thyroidectomy.

Thyroglossal cyst excision.

TO BE A SUB-SPECIALIST IN ENDOCRINE SURGERY

The trainee would normally be expected to be competent to perform:

Thyroid Lobectomy/isthmectomy.

Sub-total thyroidectomy (including Graves Disease)

Total Thyroidectomy.

Thyroglossal cyst excision.

Thyroidectomy for advanced malignancy.

Re-operative thyroid surgery.

Modified block dissection of neck for thyroid malignancy.

Parathyroidectomy.

Adrenalectomy.

Surgery for tumours of the endocrine pancreas.

Surgery for gastrointestinal endocrine disease.

This operative experience will be assessed from the SAC approved Log Book.