Diseases in the neck include a wide variety of pathological conditions, some of whose symptoms and signs are confined to the neck, e.g. thyroglossal cyst and carotid body tumours, to diseases that originate in the neck but present with the symptoms of dysfunction in organs outside the neck, e.g. the systemic symptoms caused by a toxic goitre, and to diseases that originate outside the neck but present with neck symptoms, e.g. cancers that spread to and enlarge the lymph glands of the neck.

Patients with neck problems may be referred to different specialists – general surgeons, endocrine surgeons, ENT surgeons and physicians.

Because the diagnosis can often be made from the symptoms and signs, the family doctor often initiates the initial diagnostic workup – blood tests and simple imaging – leaving the specialist to order the more complex haematological and imaging investigations required to help choose the best management.

Patients with neck diseases commonly present with the following problems:

- localized or diffuse swelling
- pain or discomfort
- deformity
- voice change
- stridor
- dysphagia
- clinical syndromes associated with hormone dysfunction.

A complete history and examination will often indicate the source of the symptoms and signs, commonly the lymph nodes and the thyroid gland. Neck symptoms can, however, arise from any of the structures in neck: from the musculoskeletal system (cervical spine spondylosis, osteophytes, cervical rib), from the vascular and lymphatic structures (carotid body tumour, subclavian aneurysm, cystic hygroma), from the pharynx (pharyngeal pouch), from the nervous system (neuralgia, neurogenic tumours), from the skin and soft tissues (lipoma, sebaceous cyst) and from congenital lesions (thyroglossal tract, branchial clefts).

Conditions which are not specific to the neck, i.e. those affecting the spine, nerves, skin and soft tissue, are not discussed in this chapter.

To help clinicians describe the exact anatomical location of any abnormality, the neck is subdivided into several compartments. For example, the anterior border of the sternocleidomastoid muscle is used to divide the neck into anterior and posterior triangles, a distinction particularly relevant to the definition of the site of lymph node disease and lymph node dissection.

If the exact location of the disease and its nature is ambiguous, two simple investigations, an ultrasound scan of the neck and fine needle aspiration or core biopsy, can usually speedily reveal the source of the patient’s problems, confirm the clinical diagnosis and expedite management. Biopsy should never be used before excluding lesions such as a carotid body tumour, an aneurysm or a pharyngeal pouch because a needle inserted into these swellings can be fatal.

Diseases of the larynx (laryngocele), hypopharynx (globus, pharyngeal pouch) and the cervical oesophagus (tumours, dysmotility) are discussed in other chapters.

### CERVICAL LYMPHADENOPATHY

Lymphadenopathy presents in the neck more often than elsewhere because the cervical lymph nodes are close to the skin and receive lymph from the structures of the upper airway, the mouth and pharynx, the limbs and all the organs in the chest abdomen.

The common causes of cervical lymph node enlargement are listed in Table 13.1.
The neck and the salivary glands

Investigation
Clinical diagnostic indicators
A detailed history and physical examination of a patient with cervical lymphadenopathy often reveals not only the underlying diagnosis but also the optimum management pathway.

**A history of blood transfusions or intravenous drug abuse** should alert the clinician to a possible viral cause.

A history of **tuberculosis** may point to tubercular lymphadenitis.

**Travel** to certain tropical countries or specific occupations can suggest specific chronic infections such as trypanosomiasis, leishmaniasis and tularaemia.

The presence of **other associated conditions** such as upper respiratory tract infection, pharyngitis, conjunctivitis, periodontal disease, insect bites and dermatitis are obvious pointers to the source of a reactive lymphadenopathy.

**The physical features of the lymph nodes** such as their size and location can point to the underlying diagnosis. Multiple small lymph nodes tend to be reactive, while large, matted nodes that may occasionally be fluctuant are likely to be caused by an acute bacterial infection or tuberculosis with cold abscess formation.

Diseases such as lymphoma and certain systemic autoimmune conditions can present with a generalized as well as cervical lymphadenopathy which are large and rubbery.

Hard enlarging cervical lymph nodes often contain carcinoma originating in the organs the nodes drain. Identification of the specific groups of lymph nodes enlarged should indicate to the clinician which drainage areas to examine and investigate.

Lymph nodes greater than 1 cm diameter, nodes that are increasing in size and nodes that persist for more than a month should be considered significant and investigated.

The investigations of a patient with significant lymphadenopathy can be used just to screen for the various conditions listed in Table 13.1 or designed, with the clinical features already established, to be diagnostic.

**Blood tests**
A full blood count and erythrocyte sedimentation rate (ESR) is almost always helpful but rarely identifies the cause of the lymphadenopathy unless a marked leucocytosis indicates leukaemia.

**Imaging**
An ultrasound scan of the neck often helps reveal the origin of the lymphadenopathy and establish its extent. It will also identify or exclude associated lesions in the neck such as thyroid lumps and facilitate an image-guided biopsy or needle aspiration.

An **ear, nose and throat examination, an upper gastrointestinal (GI) endoscopy and CT scans** of the chest and abdomen may be needed if metastatic lymphadenopathy is suspected to identify the primary tumour before considering an excision biopsy.

**Tissue biopsy**
Fine needle aspiration (FNA) is often the test that reveals the diagnosis, but if it does not and if a

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**Table 13.1**
**Causes of cervical lymphadenopathy**

<table>
<thead>
<tr>
<th>Reactive or inflammatory</th>
</tr>
</thead>
<tbody>
<tr>
<td>Viral illness – Epstein–Barr virus, cytomegalovirus, human immunodeficiency virus, hepatitis B virus</td>
</tr>
<tr>
<td>Acute suppuration in the head and neck</td>
</tr>
<tr>
<td>Granulomatous diseases such as tuberculosis, syphilis and sarcoidosis</td>
</tr>
<tr>
<td>Other infections – toxoplasmosis, cat scratch disease, lymphogranuloma venereum, chancroid, typhoid, brucellosis, plague, tularaemia, measles, rubella, lyme disease</td>
</tr>
<tr>
<td>Systemic lupus erythematosis, rheumatoid arthritis, Kawasaki disease, Still’s disease, dermatomyositis</td>
</tr>
</tbody>
</table>

| Malignant disease                                                                            |
| Primary – lymphoma, leukaemia                                                               |
| Secondary – metastases from primaries in the head and neck, breast, lungs, abdomen and testes |

| Drug induced                                                                                 |
| By allopurinol, carbamazepine, hydralazine, phenytoin and certain antibiotics               |

| Miscellaneous                                                                                 |
| Amyloidosis                                                                                  |
detailed pathological examination of the lymph node is essential for accurate classification, as in suspected lymphoma, an **excision biopsy** may be required.

Fine needle aspiration has a good chance (85 per cent) of revealing caseation and epithelioid cells if tuberculosis is suspected, but acid-fast bacilli will only be seen in 50 per cent. If pus is aspirated it must be sent for culture and tested for antibiotic sensitivity.

It is important to exclude a metastatic neck malignancy before performing an open biopsy, to avoid compromising a subsequent block dissection.

**Management**

The management of cervical lymphadenopathy depends on the underlying cause. If the cause is reactive or inflammatory, treatment of the underlying or associated illness is all that is required.

Combined drug therapy for tuberculosis may be started on the basis of a positive FNA with further supervision by a surgeon and physician.

If an abscess develops it should be drained by aspiration or incision while medical therapy continues.

Occasionally, after successful medical treatment, a large residual fibrotic mass of glands may need excision.

The treatment of other serious underlying illnesses, such as malignancy, requires discussion with a specialist team to review and choose the best oncological and surgical options.

**THYROID SWELLINGS**

**Investigation**

**Clinical diagnostic indicators**

A neck swelling is considered to arise from the thyroid gland if it is in the region of the thyroid gland (front of the neck) and **moves on swallowing**. Rarely a solitary colloid cyst arising from one of the lobes may lie in a more lateral position.

A centrally placed swelling just below or above the hyoid bone that moves with protrusion of the tongue is suggestive of a **thyroglossal cyst**.

The examination of a patient with a thyroid lump should be aimed towards obtaining answers to the following questions.

- Is the lump solitary, multi-nodular or diffuse?
- Is the patient clinically euthyroid, hypothyroid or hyperthyroid?
- If a thyroid cancer is suspected, is there associated lymphadenopathy or evidence of distant disease in the lungs, bones etc?

Although the clinical features may not accurately distinguish between specific diseases of the thyroid, they provide valuable clues about the possible diagnosis. For example, a diffuse goitre in a patient with obvious hyperthyroidism suggests Graves’ disease, whereas a long-standing nodular goitre in a patient living in an endemic area suggests a benign multinodular goitre.

Table 13.2 lists the various possible causes of goitre and relates them to the patient’s thyroid functional status and their presentation in the neck.

The investigations used to identify thyroid function, in addition to clinical examination, are mostly blood tests. Imaging is used for establishing the anatomical abnormality and guiding tissue biopsy.

**Blood tests**

**Thyroid function tests** evaluate thyroid function and determine whether the cause of any thyroid dysfunction originates within the thyroid or the pituitary gland.

The tests of function include measurements of the levels of **TSH** (thyroid-stimulating hormone), **free T4** (thyroxine) and occasionally **free T3** (tri-iodothyronine). Normal T3 and T4 levels with abnormal TSH levels indicate subclinical dysfunction. High and low TSH levels indicate subclinical hypothyroidism and hyperthyroidism respectively.

**Antithyroid antibodies** may be present in patients with suspected autoimmune thyroid disorders.

**Serum calcium** levels should be checked to confirm normal parathyroid function.

**Calcitonin** levels should be performed in patients with a family history of **medullary thyroid carcinoma** and are increasingly being recommended in all patients with thyroid nodules for the early detection and appropriate treatment of sporadic medullary thyroid cancer.

**Imaging**

An **ultrasound scan** of the neck is performed as routine by many surgeons on patients with thyroid
### Table 13.2
Relationship between clinical features, function and pathology

<table>
<thead>
<tr>
<th>Clinical features</th>
<th>Hypothyroid</th>
<th>Euthyroid</th>
<th>Hyperthyroid</th>
</tr>
</thead>
<tbody>
<tr>
<td>Diffuse</td>
<td>Thyroiditis</td>
<td>Iodine deficiency</td>
<td>Primary hyperthyroidism</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Enzyme defects</td>
<td>(Graves’ disease)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Goitrogens</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Thyroiditis</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Amyloid</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Pregnancy, puberty</td>
<td></td>
</tr>
<tr>
<td>Multinodular enlargement</td>
<td>Multinodular goitre with gross degeneration</td>
<td>Multinodular goitre</td>
<td>Secondary hyperthyroidism</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Anaplastic carcinoma</td>
<td>(Plummer’s syndrome)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Medullary carcinoma</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Lymphoma</td>
<td></td>
</tr>
<tr>
<td>Solitary nodule</td>
<td>Coincidental nodule with myxoedema</td>
<td>Cyst</td>
<td>Autonomous toxic nodule</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Dominant nodule</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Adenoma</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Follicular or papillary carcinoma</td>
<td></td>
</tr>
<tr>
<td>No goitre</td>
<td>Thyroiditis</td>
<td>Normal gland</td>
<td>Primary hyperthyroidism</td>
</tr>
<tr>
<td></td>
<td>Primary myxoedema</td>
<td></td>
<td>Thyroxine overdose</td>
</tr>
<tr>
<td></td>
<td>Post-thyroidectomy</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Post-radioiodine</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Lumps (Fig 13.1). It has several uses: it can confirm the presence of thyroid enlargement, differentiate between solitary and multiple nodules, differentiate between solid and cystic lumps, identify retrosternal extension and any associated cervical lymphadenopathy, and be used to target FNA or biopsy.

Recent advances in technology also allow the ultrasonographer to distinguish between benign and malignant lumps.

**CT or MRI scans** of the neck should be performed in patients with large thyroid masses with suspected retrosternal extension (Fig 13.2), tracheal deviation and compression or locally advanced malignancy (Fig 13.3).

**Thyroid scintigraphy** is occasionally undertaken to look for ectopic thyroid tissue and in those with subclinical hyperthyroidism to identify functioning (hot) nodules.

**Laryngoscopic** vocal cord examination is performed routinely in most centres before thyroid
Euthyroid benign nodules

Surgery to establish a pre-operative baseline of recurrent laryngeal nerve function against which any postoperative vocal disturbance may be compared and as a medico-legal precaution.

Tissue biopsy

Fine needle aspiration (either free hand or ultrasound guided) should be performed on all solitary or dominant nodules to determine their morphology (Fig 13.4).

Ultrasound-directed FNA provides a more accurately defined source and representative specimen.

FNA cannot differentiate between a follicular adenoma and a follicular carcinoma as the latter is distinguished from a benign adenoma by the presence of vascular and/or capsular invasion, something which can only be identified on histological examination.

Fine needle aspirations can and often are repeated if the aspirate is inadequate/insufficient or if a benign result does not corroborate with clinical suspicion. FNA can be therapeutic for thyroid cysts.

An ultrasound-guided core biopsy or an open incisional biopsy is occasionally performed on tumours of uncertain origin and anaplastic cancers.

Patients with medullary thyroid cancer should undergo investigations to exclude hyperparathyroidism and phaeochromocytoma in case they have the MEN II (multiple endocrine neoplasia) syndrome and be offered genetic screening for RET mutations irrespective of family history.

Management

The results of thyroid function tests, ultrasound imaging and FNA cytology help determine the management options of a thyroid nodule as set out in Fig 13.5.

Euthyroid Benign Nodules

Surgical excision (partial thyroidectomy) is indicated for benign nodules, single or multiple, that are increasing in size, causing pressure effects or are causing cosmetic concern.
EUTHYROID MALIGNANT NODULES

Surgical excision is recommended for all thyroid lumps that are malignant on cytological examination, whether definite, suspicious or indeterminate, except in cases of lymphoma, secondary deposits or anaplastic cancer.

For a well-differentiated thyroid cancer of less than 1 cm diameter, a lobectomy may be sufficient treatment, provided there is no family history of thyroid cancer or a history of neck irradiation.

Large (>1 cm diameter) malignant tumours are treated with a total thyroidectomy.

Masses that are suspicious or indeterminate on preoperative cytology such as follicular neoplasms may be offered a hemi-thyroidectomy, but if the lesion is found to be malignant and of more than 1 cm diameter a second operation should be performed to complete the total thyroidectomy, often accompanied by a central lymph node dissection.

Lateral lymph nodal excision is recommended in patients with palpable lymph node disease.

Radio-iodine ablation and TSH suppressive doses of thyroxine are recommended as adjuvant treatments for differentiated thyroid cancer of >1 cm diameter, while thyroxine replacement is sufficient in medullary thyroid cancer.

External radiotherapy and chemotherapy may be required in advanced disease and in those with anaplastic cancer. All such treatment options should be considered at a multidisciplinary team meeting.

DIFFUSE OR NO ENLARGEMENT WITH DISORDERED THYROID FUNCTION

The management of patients with thyroid dysfunction (hypo- or hyperthyroidism) with or without enlargement of the gland or benign lesions such as nodules, without pressure effects or cosmetic concerns, is usually referred to an endocrinologist for long-term medical management.

Thyroxine replacement is given for hypothyroidism.

Anti-thyroid drugs, such as carbimazole or propylthiouracil, and/or radio-iodine are given for hyperthyroidism. The risk of hypothyroidism after radio-iodine may be as high as 30 per cent 3 years after treatment and then increases steadily thereafter.

Local problems are dealt with once the patient has been rendered euthyroid.

Total thyroidectomy (with careful preservation of the parathyroid glands) may be offered to patients with hyperthyroidism caused by recurrent or poorly controlled Graves’ disease who are young, female or have a large toxic goitre, as an alternative to long-term anti-thyroid drugs or radio-iodine.

For many years the standard operation for hyperthyroidism was subtotal thyroidectomy, an operation that preserves just 2–10 g of thyroid tissue; but because the results of pharmacological and radioiodine treatment are now so good, and because this operation has a 5 per cent recurrence of hyperthyroidism at 5 years and a slowly increasing rate of hypothyroidism, it is rarely practised.

Before the operation all patients with hyperthyroidism should receive a course of anti-thyroid drugs to establish a euthyroid state – a prerequisite for a successful surgical intervention.
All patients need thyroxine after total thyroidec-
tomy, as do some after subtotal thyroidectomy.

**BENIGN SOLITARY TOXIC NODULE**

Patients with a toxic autonomous nodule should be offered thyroid lobectomy after being rendered clinically euthyroid with anti-thyroid drugs.

**SOLITARY THYROID CYSTS**

Thyroid cysts are usually managed by aspiration. **Excision** is reserved for those cysts which are large, symptomatic or recurrent.

**THYROGLOSSAL CYST**

The thyroglossal tract is the remnant of the diverticu-
lum that develops in the floor of the primitive embry-
onic pharynx at the base of the tongue (foramen caecum) which, after extending caudally, develops into the isthmus and pyramidal lobe of the thyroid gland. The tract normally disappears but occasionally part of it can persist to become a cyst.

**Investigation**

**Clinical diagnostic indicators**

A thyroglossal cyst usually presents early in life, but may appear in adult life, as a **midline swelling**, most often **in front of or below the hyoid cartilage**. However, it can develop anywhere from the base of the tongue to the manubrium sterni.

In addition to moving upwards during swallow-
ing, as other thyroid swellings, the thyroglossal cyst also **moves upwards with protrusion of the tongue**.

**Differential diagnoses** include a mid-line lymph node, an enlarged pyramidal lobe of the thyroid gland, a dermoid cyst and a sebaceous cyst. Very occasionally, the cyst wall contains the patient’s only functioning thyroid tissue (an ectopic thyroid) and so the demonstration of a normal thyroid gland by ultrasound imaging is an important precaution.

**Imaging**

**An ultrasound scan** will confirm the diagnosis.

**Table 13.3**

**Causes of hypercalcaemia**

Malignancies – primary solid tumours, metastases and haematological malignancies

Hyperparathyroidism

Other endocrine diseases – hyperthyroidism, Addisonian crises

Drugs – thiazide diuretics, lithium, vitamin A and vitamin D toxicity, aluminium toxicity

Miscellaneous – immobilization, laboratory error, milk alkali syndrome, granulomatous diseases, e.g. sarcoidosis

An **isotope scan** should be performed before excising the cyst if there is doubt about the presence of normal thyroid tissue in the orthotopic location.

**Management**

**Surgical excision with the tract above it** including the central portion of the hyoid bone (Sistrunk’s procedure) is indicated for infection, cosmetic rea-
sons, pressure effects or malignant change (rarely diagnosed before definitive histology is available).

Recurrence is possible if the tract is not com-
pletely excised.

**THE PARATHYROID GLANDS**

The four parathyroid glands (a superior and an inferior on either side) normally lie close to the thy-
roid gland. The clinical manifestations of parathy-
roid diseases are invariably the result of abnormal levels of serum calcium caused by pathologically high or low levels of parathyroid hormone (PTH).

**Hypercalcaemia** is often a chance finding detected during the screening investigations for gastrointes-
tinal, cardiovascular, renal and neuro-psychiatric disorders and other symptom patterns such as myalgia, lethargy, visual changes and pruritus.

The causes of hypercalcaemia and the disorders of the parathyroid glands that cause it are listed in Tables 13.3 and 13.4.

Patients with **hypoparathyroidism** are managed by endocrinologists with calcium and vitamin D supplementation.
The neck and the salivary glands

Table 13.4
Disorders of the parathyroid glands

Hyperparathyroidism
Primary hyperparathyroidism
Single adenoma (89 per cent)
Double adenoma (4 per cent)
Hyperplasia (6 per cent)
Parathyroid cancer (1 per cent)
Secondary hyperparathyroidism
Associated with renal failure
Associated with vitamin D deficiency
Tertiary hyperparathyroidism
Autonomous function after long-standing secondary hyperparathyroidism
Familial hypocalciuric hypercalcaemia
Idiopathic hypercalcaemia
Lithium

Hypoparathyroidism
Anatomical damage – post surgery, radiotherapy, autoimmune
Reduced function – hypomagnesaeemia, PTH and calcium sensing receptor gene defects
Parathyroid agenesis

Table 13.5
Biochemical characteristics in hyperparathyroidism

<table>
<thead>
<tr>
<th>Test</th>
<th>Primary HPT</th>
<th>Secondary HPT</th>
<th>Tertiary HPT</th>
</tr>
</thead>
<tbody>
<tr>
<td>Serum PTH</td>
<td>High</td>
<td>High</td>
<td>High</td>
</tr>
<tr>
<td>Serum calcium</td>
<td>High</td>
<td>Low/normal</td>
<td>High</td>
</tr>
<tr>
<td>Serum phosphate</td>
<td>Low</td>
<td>High/normal</td>
<td>High</td>
</tr>
<tr>
<td>Vitamin D</td>
<td>Normal/low</td>
<td>Low</td>
<td>Low/normal</td>
</tr>
</tbody>
</table>

HPT, hyperparathyroidism.

The presence and type of hyperparathyroidism and, when appropriate, the necessary imaging tests to localize the parathyroid glands must be performed before considering surgical treatment.

Blood and urine tests
The serum calcium, phosphate, PTH and vitamin D levels and 24-hour urinary calcium excretion should be measured.

Table 13.5 shows the various biochemical results found in the different types of hyperparathyroidism.

In patients with suspected primary hyperparathyroidism, determination of the 24-hour urinary excretion of calcium is essential to confirm high levels and thus rule out the autosomal dominant benign disorder called familial hypocalciuric hypercalcaemia (FHH). In this condition, PTH levels may be elevated but urinary calcium levels are typically low because of an inability of the kidneys to secrete calcium. These patients do not benefit from a parathyroidectomy.

Imaging
Imaging for localization of the abnormal parathyroid gland/s is currently undertaken in patients with primary hyperparathyroidism (and by some surgeons in secondary/tertiary hyperparathyroidism) to facilitate the performance of a unilateral or focused parathyroidectomy.

The investigation and management of patients with an autoimmune hypoparathyroidism are beyond the scope of this book.

The following paragraphs focus on the investigation and management of hyperparathyroidism.

The results of thyroid function tests, ultrasound imaging and FNA cytology that help determine the management options of a thyroid nodule are set out in Fig. 13.5).
The tests used to identify an abnormal gland include a combination of a **preoperative ultrasound scan** and a **99Tc sestamibi radionuclide subtraction scan (MIBI)** (Fig 13.6).

**Intraoperative localization techniques** such as an intraoperative ultrasound, radio-guided surgery and methylene blue injection may improve the success of the primary operation, and facilitate a focused approach and thus avoid extensive dissection and the risk of damaging the normal parathyroid glands in patients with a single parathyroid adenoma (Fig 13.7).

**An intraoperative PTH assay** may be used to confirm successful excision of the overactive parathyroid tissue.

**CT or MRI scanning** may be required for patients with recurrent disease prior to re-operation, with or without **selective venous sampling for PTH levels**.

---

**Management of primary hyperparathyroidism caused by an adenoma**

Parathyroidectomy is the gold standard in the treatment of symptomatic primary hyperparathyroidism.

The indications for parathyroidectomy in symptomless hyperparathyroidism include patients at high risk of developing complications, i.e. those with high calcium levels (>2.85 mmol/L), high urinary calcium (excretion of >10 mmol/day), reduced creatinine clearance (<70 per cent of normal), significant osteoporosis (bone density T score less than −2.5), young patients (<50 years of age) and patients in whom surveillance is not possible.

Lithium therapy can increase both calcium and PTH levels and should be excluded as a cause of hyperparathyroidism before surgery.

Thiazide diuretics can exacerbate hypercalcaemia in primary hyperparathyroidism. Patients on these medications should be taken off these drugs if possible, and the levels of calcium and PTH rechecked.

As the majority of cases of primary hyperthyroidism are caused by a solitary adenoma, preoperative localization with or without intraoperative localization facilitates unilateral exploration. If the preoperative imaging is inconclusive, as is often the case with glandular hyperplasia, bilateral neck exploration is needed.

The success of a focused parathyroidectomy is thought to be equivalent to a formal bilateral neck exploration in terms of resolution of hypercalcaemia.

---

**Management of primary hyperparathyroidism caused by general hyperplasia**

Parathyroid hyperplasia can be familial and can occur as part of the MEN syndromes (MEN I and MEN IIa). Other endocrine abnormalities (especially a co-existing phaeochromocytoma) should be excluded by biochemical testing in all patients with suspected hyperplasia (young patients with a family history) prior to parathyroidectomy.

**Excision of three glands and one-half of the fourth gland**, leaving the remaining half gland carefully marked, or else autotransplanted, is the most common surgical treatment for patients with primary general parathyroid hyperplasia.
Management of secondary and tertiary hyperparathyroidism

This condition is usually associated with renal failure and patients on haemodialysis. It occurs in renal transplant patients and in those with long-standing end-stage renal disease, where one or more of the glands assume autonomous function following chronic stimulation by hypocalcaemia.

Medical management consists of calcium supplements, vitamin D analogues and phosphate binding agents in an attempt to regulate the levels of calcium, phosphate and vitamin D.

New organic agents (calcimimetics) are now being used to increase the sensitivity of calcium receptors to extracellular calcium and suppress PTH secretion.

Surgery is occasionally required; but as only a small proportion of these patients have single gland disease, and even though this can be localized preoperatively by imaging and excised, the traditional treatment in these patients is a full neck exploration with either subtotal excision of the parathyroid glands (three and a half glands) or total parathyroidectomy with autotransplantation of some parathyroid tissue into either the sternocleidomastoid muscle or the forearm muscles.

Many surgeons perform a thymectomy in addition to excision of the parathyroid glands as these patients can have accessory parathyroid tissue within their thymus.

Complications of parathyroidectomy

Specific complications of parathyroid surgery include persistent or recurrent hypercalcaemia needing further treatment, hypoparathyroidism and the uncommon possibility of recurrent laryngeal nerve damage. Short-term hypocalcaemia is common and requires intravenous calcium.

Hypoparathyroidism in patients with renal transplants can contribute to worsening of graft function.

Prognosis Failure to control hyperparathyroidism is usually the result of a failed surgical exploration. Recurrence after two explorations is over 10 per cent. Hypoparathyroidism is treated with vitamin D and calcium supplements.

CAROTID BODY TUMOUR

A number of different tumours of neuro-ectodermal tissue origin can develop in the neck, including neuroma, neurofibroma, neurilemmoma, carotid body tumour and glomus tumour. The last one develops from paraganglionic cells in close anatomical association with the carotid artery bifurcation and occasionally from cells adjacent to the jugular bulb, the middle ear cavity and the ganglion nodosum of the vagus nerve.

Carotid body tumours may be associated with other paragangliomata in the neck, chest and abdomen and with phaeochromocytomata. This is especially true in 10 per cent of familial cases where around 30 per cent are also bilateral.

These tumours are very vascular and approximately 10 per cent are malignant.

Investigation

Clinical diagnostic indicators

The presence of a relatively long-standing painless firm mass below the angle of the mandible should raise the suspicion of a carotid body tumour. The swelling is often associated with a transmitted pulsation and a bruit and occasionally with pressure effects on adjacent vital structures. It is easily compressible and refills on release. It can be moved from side to side but not vertically.

Imaging

A CT scan, MRI or duplex scan can provide accurate localization and define the tumour’s relationships to adjacent structures while excluding bilateral disease (Fig 13.8).

Screening

The family members of patients with a family history should be screened in view of the conditions autosomal dominant mode of inheritance.

Tissue biopsy

Percutaneous aspiration or biopsy can be hazardous and should not be attempted.

Management

Surgical resection should be carried out through the same incision as a carotid endarterectomy. The
tumour, which is very vascular, needs to be dissected off the artery. It is rarely necessary to resect the artery unless the tumour is malignant.

Damage to the cranial nerves in the neck is a potential complication. Stroke is rare.

Radiotherapy may be considered in patients with inoperable malignant tumours, incomplete resections, recurrent disease or metastases but is rarely effective.

**BRANCHIAL CYST**

Branchial cysts may be present at birth or appear later in life (commonly in the third decade). They are thought by some to be unrelated to branchial abnormalities and arise in the lymphatic tissue of the neck.

**Investigation**

**Clinical diagnostic indicators**

The patient is usually the first to notice the external skin opening.

Both a sinus and fistula may be associated with an intermittent mucoid discharge. Both may become infected to form an abscess with or without purulent discharge.

**Management**

**Surgical excision** is recommended both for treatment and for confirmation of the diagnosis.

If the cyst is infected it should be aspirated under the cover of a course of antibiotics and excision delayed until the infection has resolved.
The neck and the salivary glands

**CYSTIC HYGROMA**

A cystic hygroma is a collection of dilated lymphatic sacs that develop and fill during prenatal life from sequestered (unconnected) embryonic lymph vessels. They usually present before the second year of birth and can sometimes be diagnosed prenatally by antenatal ultrasonography, but they may appear in adult life.

**Investigation**

**Clinical diagnostic indicators**

Cystic hygromata present as a **brilliantly translucent, fluctuant mass**. Although commonly found in the posterior triangle of the neck, they can often extend across the boundaries of the neck compartments. They occasionally cause compression of the oesophagus, trachea or the great vessels. In a neonate a complication such as haemorrhage into the cyst or acute infection can be fatal.

The **differential diagnoses** include other causes of cystic neck masses such as branchial cysts, thyroglossal cysts, thymic cysts, cervical bronchogenic cysts, dermoid and epidermoid cysts.

**Imaging**

**Ultrasound scanning** will confirm the cystic nature of the mass and determine its extent and relationship to other neck structures.

A **CT scan or MRI** provides superior resolution and is very helpful in planning the excision of an extensive hygroma especially in young children or neonates (Fig 13.9).

**Management**

**Surgical excision** is the appropriate treatment of symptomatic or large hygromas at any age. Small cystic lesions in children can be left until the child is 3–4 years old.

![Figure 13.9](image)

**FIGURE 13.9** Nuclear magnetic resonance scans of a large lobulated cystic hygroma in the neck. (A) Axial STIR scan showing the cyst surrounding the major neck vessels and extending down below the clavicle. (B) Coronal STIR scan showing the cyst extending medially deep to the mandible and displacing the pharynx to the right. Reproduced by kind permission of the Department of Paediatric Surgery, Queen’s Medical Centre, Nottingham.
Although a complete excision is usually possible, technical difficulties caused by the adherence of the cyst to nearby nerves and vessels and the weak friable cyst wall are not uncommon, making the chance of an incomplete removal and subsequent recurrence not insignificant.

**Obliterative sclerosis** is an alternative to surgery. Intralesional injections of compounds such as Ethibloc or OK 432 have been used with variable results.

### CERVICAL RIB AND THORACIC OUTLET SYNDROME

One per cent of the population have an accessory rib at the level of the seventh cervical vertebra which can be the cause of the thoracic outlet syndrome, i.e. upper limb symptoms caused by compression of nerves and/or blood vessels at the thoracic outlet.

Compression of the brachial plexus and subclavian vessels occurs in the triangle bounded by the scalenus anterior and scalenus medius muscles and a cervical rib or the first rib, but can also be caused by accessory muscle bands and tendons, fibrous bands and hyperextension neck injury.

**Investigation and management**

See Chapter 11.

### PHARYNGEAL POUCH

See Chapter 18.

### THE SALIVARY GLANDS

Salivary glands are traditionally classified as being major (the parotid, submandibular and sublingual glands) and minor (those widely distributed below the mucosa of the lips, palate, pharynx and larynx). The following only discusses problems in the parotid and submandibular glands because they are the commonest sites of salivary gland pathology. Many of the problems described can be encountered in the sublingual and minor salivary glands.

Table 13.6 gives a comprehensive list of salivary gland pathology and the distribution of the different diseases in the different glands.

Infective, inflammatory and neoplastic conditions occur more frequently in the parotid gland; the submandibular gland is more often the site of obstructive calculus disease. This is attributed to the increased mucin content in the secretions of the submandibular gland and the ascending course of its duct.

**Investigation**

**Clinical diagnostic indicators**

The commonest presenting symptoms in patients with salivary gland pathology are pain and swelling.

Patients with Sjögren’s syndrome may present with dry mouth and eyes.

Examination of these patients should not be limited to the symptomatic gland but include an examination of all the major salivary glands, the seventh cranial nerves and the neck.

**Blood tests**

Blood tests demonstrating the presence of anti-nuclear factor (ANF), rheumatoid factor (RF) and raised gamma globulin levels can be useful in providing corroborative evidence of Sjögren’s syndrome.

Serum angiotensin-converting enzyme levels (SACE) may be useful in suspected sarcoidosis.

**Imaging**

A chest X-ray may reveal sarcoidosis.

An X-ray of the floor of the mouth and cheek may reveal stones in the salivary ducts and in the glands (Fig 13.10).

An ultrasound scan is often employed to confirm that the swelling is in a salivary gland and exclude other swellings such as dental and branchial cysts and abnormalities of the masseter muscle and the mandible.

Although further investigations are rarely required, CT scanning and MRI are now replacing plain X-rays, sialography and radionuclide scanning.

If a neoplastic lesion is suspected, MRI can provide more information than CT about the involvement of the deep lobe, the extent of invasion and any associated soft tissue abnormalities.

**Cytology**

Fine needle aspiration is of variable value in benign conditions. It is used routinely by some and rarely by others but can be helpful in ruling out conditions such as lymphadenopathy, lipoma, neuroma and non-salivary gland cysts, and, when the nature of the pathological change is in doubt, malignant change.
The neck and the salivary glands

Table 13.6
Conditions of the salivary glands

<table>
<thead>
<tr>
<th>Infections</th>
<th>Carcinoma in a pleomorphic adenoma</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acute bacterial infection – e.g. suppurative parotitis</td>
<td>Acinic cell carcinoma</td>
</tr>
<tr>
<td>Viral – mumps (commonest) and other viruses including coxsackie, influenza and herpes</td>
<td>Squamous cell carcinoma</td>
</tr>
<tr>
<td>Other – tuberculosis, syphilis, cat-scratch disease, toxoplasmosis, actinomycosis</td>
<td>Miscellaneous rare tumours (benign and malignant)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Obstructive salivary disease</th>
<th>Haemangioma</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sialectasis (recurrent parotitis)</td>
<td>Lymphangioma</td>
</tr>
<tr>
<td>Calculus disease</td>
<td>Lipoma</td>
</tr>
<tr>
<td>Inflammatory and autoimmune diseases</td>
<td>Sarcoma</td>
</tr>
<tr>
<td>Sarcoidosis</td>
<td>Lymphoma</td>
</tr>
<tr>
<td>Sjögren’s disease</td>
<td>Secondary deposits</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Tumours</th>
<th>Sialomegaly associated with chronic disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>Benign</td>
<td>Endocrine diseases – Myxoedema, Cushing’s disease and diabetes mellitus</td>
</tr>
<tr>
<td>Pleomorphic adenoma</td>
<td>Cirrhosis</td>
</tr>
<tr>
<td>Warthin’s tumour</td>
<td>Alcoholism</td>
</tr>
<tr>
<td>Oncocytoma</td>
<td>Gout</td>
</tr>
<tr>
<td>Malignant</td>
<td>Bulimia</td>
</tr>
<tr>
<td>Mucoepidermoid carcinoma</td>
<td>HIV</td>
</tr>
<tr>
<td>Adenoid cystic carcinoma</td>
<td>Drugs – thiouracil, phenylbutazone, isoprenaline, dextropropoxyphene, oral contraceptive pill</td>
</tr>
<tr>
<td>Adenocarcinoma</td>
<td></td>
</tr>
</tbody>
</table>

Figure 13.11 shows how the combination of clinical features and investigations can lead to a diagnosis.

Management

Infection: bacterial and viral

Institute adequate analgesia and hydration, and give antibiotics for bacterial infections. Investigate other sites for chronic infections such as tuberculosis and granulomatous diseases such as sarcoidosis and institute specific treatment as appropriate.

Sjögren’s disease

Lubrication of the oral cavity and eye and pilocarpine to stimulate residual function are required.

Sialectasis

Institute analgesia and hydration during acute episodes.
The salivary glands

Sialography can help in confirming the diagnosis, and may also help in relieving symptoms.

Duct dilatation may improve flow, while very occasionally radiotherapy and division of parasympathetic nerve supply may help.

Total parotidectomy is the last resort and is fraught with complications.

Salivary gland calculi
The treatment is stone removal from the duct via the oral cavity or elective excision of the gland for intraglandular calculi or recurrent disease. These can also now be treated by lithotripsy (as for urinary calculi).

Pleomorphic adenoma
Pleomorphic adenoma is the commonest salivary gland tumour and occurs predominantly in the superficial lobe of the parotid gland. Local excision of the involved lobe (superficial parotidectomy) or total parotidectomy are the treatments of choice (Fig 13.12). Malignant transformation is very rare, but local recurrence is common.

Warthin’s tumour
Warthin’s tumour (papillary cystadenoma lymphomatosum) often occurs in older people and can be multiple or bilateral in up to 10 per cent. It occurs in the parotid gland usually in the superficial lobe. Superficial parotidectomy (Fig 13.12) is the treatment of choice. Local recurrences after excision are rare.

Oncocytoma
Oncocytoma (oxyphil cell adenoma) often occurs in minor salivary glands. It is often seen in older people as a slow-growing soft tumour and rarely turns malignant. Complete excision usually requiring no more than a superficial parotidectomy (Fig 13.12) is the treatment of choice.

Mucoepidermoid carcinoma and other malignant conditions
Malignant salivary gland tumours are rare. The commonest type is mucoepidermoid tumour; the other types are listed in Table 13.6.

Fortunately, the vast majority are amenable to surgical excision but the proximity of the major...
The neck and the salivary glands

The neck and the salivary glands to important nerves and vessels significantly influences the extent of radical resection. Nevertheless, surgery is often the only curative option for overtly malignant tumours. There is no alternative effective treatment although radiotherapy may be a useful adjunct.

Preoperative imaging with MRI is useful to stage the local extent of the disease and is helpful in preoperative planning.

Radical excision of the tumour, including sacrificing the facial nerve if necessary, is the only treatment likely to succeed.

Lymph node dissection is indicated only if the nodes are involved.

Postoperative radiotherapy is often used after surgery for mucoepidermoid and adenoid cystic carcinoma. This serves to reduce local recurrence especially after incomplete excision.

The facial nerve, if excised, can be bridged by a nerve graft. Alternatively, the corner of the mouth can be pulled up with a facial sling. This can be combined with a lateral tarsorrhaphy to protect the eye from exposure damage.

Complications of parotid gland surgery

Damage to the facial nerve, salivary fistula and paraesthesia of the ear lobe (from damage to the greater auricular nerve) is possible.

Frey’s syndrome is discomfort, sweating and redness of the skin overlying the parotid gland – related to the innervation of the skin by the parasympathetic secretory nerve fibres that normally supply the parotid gland.

Complications of submandibular gland surgery

Damage to the mandibular branch of the facial nerve, the hypoglossal nerve and the lingual nerve is possible.

TRACHEOSTOMY

A tracheostomy is an artificial opening between the cervical trachea and the external air. It may be made to assist inadequate natural ventilation and/or provide mechanical ventilation, to protect the lungs from regurgitated gastric contents, secretions and blood and to provide access for bronchial toilet. These and other indications are listed in Table 13.7.

Investigation

Clinical Diagnostic Indicators

A patient’s inability to inhale is usually obvious from their gasping struggles to breath and in extreme circumstances the rapid development of acute cyanosis, collapse and death.

Consequently it is always better to anticipate the need for and to perform a tracheostomy before the situation becomes an acute life threatening emergency.

Management

Surgical technique

Except in extreme circumstances perform the procedure in an operating room together with an anaesthetist.
Tracheostomy

Place the patient supine with a firm pillow between the shoulder blades to make the neck extend.

If time allows, infiltrate the skin incision midway between the larynx and the suprasternal notch with a local anaesthetic but if the patient is becoming unconscious make the incision without any anaesthesia.

Identify the position of the trachea by palpating the larynx between two fingers and moving them downwards. Then hold the trachea still. Fixing the trachea between the fingers prevents the incision slipping lateral to the trachea (Fig 13.13).

In a dire emergency, a vertical incision is made through the skin and all the underlying structures right down to and through the second, third tracheal rings. This may involve cutting through the thyroid isthmus which will bleed profusely, but the bleeding will diminish rapidly once the trachea is open and the cervical venous congestion and cyanosis relieved. Attention may be paid to any continuing haemorrhage later.

If a tracheostomy tube is unavailable, any hollow tube, such as the plastic tube of a pen, will suffice. If a hollow tube is unavailable a rigid structure must be pushed between the divided tracheal rings to hold them apart and allow the ingress of air until a proper tracheostomy tube is obtained.

In less urgent circumstances, a transverse incision is made midway between the cricoid cartilage and the suprasternal notch. This leaves a better scar and should be at the level of the 2nd and 3rd tracheal rings. Divide and ligate or retract any tissues such as the thyroid isthmus before opening the trachea.

In children, make a vertical incision in the trachea. In adults remove a square of trachea between the second and fourth rings large enough to admit the tracheostomy tube.

In both situations, insert a silastic tracheostomy tube with an inflatable balloon (Fig 13.14) and connect it to the ventilation mechanism of an anaesthetic machine.

It is better to make the incision in the trachea in the form of an inverted U, the top lying just above the second ring and the sides cutting through the second and third rings, to form a flap of trachea.

Table 13.7
Indications for tracheostomy

<table>
<thead>
<tr>
<th>Indications for tracheostomy</th>
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<tbody>
<tr>
<td>To bypass acute upper airway obstruction by:</td>
</tr>
<tr>
<td>Foreign body</td>
</tr>
<tr>
<td>Infection (Diphtheria, Acute epiglossitis, Acute laryngitis)</td>
</tr>
<tr>
<td>Oedema</td>
</tr>
<tr>
<td>Haemorrhage</td>
</tr>
<tr>
<td>Tumours</td>
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<tr>
<td>Bilateral vocal cord paralysis</td>
</tr>
<tr>
<td>To assist natural respiration or facilitate artificial ventilation, as in:</td>
</tr>
<tr>
<td>Unconsciousness associated with head injuries</td>
</tr>
<tr>
<td>Neurological conditions causing coma, e.g. bulbar palsy</td>
</tr>
<tr>
<td>Chest injuries (such as flail chest)</td>
</tr>
<tr>
<td>Fulminating bronchopneumonia</td>
</tr>
<tr>
<td>Severe emphysema</td>
</tr>
<tr>
<td>By reducing the dead space</td>
</tr>
<tr>
<td>To protect the airway</td>
</tr>
<tr>
<td>from e.g. blood, gastic reflux, secretions</td>
</tr>
<tr>
<td>For bronchial toilet</td>
</tr>
<tr>
<td>e.g. in bronchiectasis and encrustations</td>
</tr>
<tr>
<td>Following laryngeal excision</td>
</tr>
</tbody>
</table>

Figure 13.13 (A–B) Surgical technique of a tracheostomy
The neck and the salivary glands

Most tracheostomies in intensive care are now performed using a percutaneous closed insertion under local anaesthetic. A gide wire is inserted into the trachea, the trachea is dilated and an endotrachial tube is inserted.

After care

Keep a suction device, a sterile catheter, tracheal dilator and a spare tube beside the bed at all times.

Ensure that the inhaled air is warm and humidified to stop the encrustation of bronchial secretions. Commercial humidifiers are available.

Clean the incision and suck out the bronchial tree regularly.

Try not to replace a silastic tube for 48 hours, i.e. until the track has started to organise and become fixed, but deflate the balloon at regular intervals to prevent the development of pressure necrosis in the tracheal endothelium.

A silver tube with a talking piece can be inserted when positive pressure ventilation is no longer required. The inner speaking tube of a silver tracheostomy tube must be removed and cleaned at regular intervals – at least every 24 hours.

Complications

- Accidental dislodgement or obstruction.
- Haemorrhage, from the wound or the trachea.
- Surgical emphysema in the neck and mediastinum.
- Pneumothorax.
- Rarely a fistula may develop between the trachea and the oesophagus.
- The trachea may become permanently stenosed at an angle of 5–10 degrees.

OTHER METHODS OF EMERGENCY VENTILATION

There are a number of other methods of assisting ventilation when the airway is obstructed. These include:

- cricothyroidotomy
- fibroptic endotracheal intubation
- the laryngeal mask
- transtracheal ventilation.