EFFECTIVE HEAD AND NECK CANCER MANAGEMENT

Third Consensus Document • 2002
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The British Association of Otorhinolaryngologists – Head and Neck Surgeons remains committed to raising standards and providing the best possible care for patients with head and neck cancer and, to this end, the first Consensus Document was published by the BAO-HNS in 1998.

I am pleased to introduce this third Consensus Document to you. It updates the 2000 issue, but retains a multidisciplinary authorship. This expanded edition has a greater emphasis on the evidence base for management decisions and has been able to draw on the teams of reviewers who contributed to the NOTO annual EBM conferences, co-ordinated by Andrew Robson. Also new to this edition is the introduction of a team of Section Editors, who have streamlined the production process.

The Association did consider deferring publication until the latest UK Guidance was published, but this document will not appear until 2003 at the earliest. Developments in Head and Neck cancer progress rapidly, and we feel that an updated version of this widely used handbook is now timely.

IAN S. MACKAY FRCS
President
Acknowledgements & Contributors

This document has been produced by the British Association of Otorhinolaryngologists - Head and Neck Surgeons. The Association gratefully acknowledge the efforts of the many contributors who include Otolaryngologists, maxillofacial surgeons, radiologists, clinical oncologists, pathologists, dieticians, speech pathologists, palliative care practitioners, and not forgetting our patient representatives, many officiated to the National Association of Laryngectomee Clubs.

We also gratefully acknowledge permission via Mr Pat Bradley, Section Editor, from the AJCC and Springer Verlag to include the latest version of the TNM Staging System for Head and Neck Cancer.

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Cancer of the head and neck does not refer to a single histological diagnosis or organ of origin. Rather, it refers to malignant tumours arising from the upper aerodigestive tract (UADT), salivary glands, thyroid and parathyroid glands, Paranasal sinuses, and the skin of the head and neck. Because of this heterogeneity, a wide spectrum of aetiologies, disease presentations, and clinical characteristics must be considered when caring for these patients. However the term “head and neck cancer” usually refers to squamous cell carcinomas arising from the mucosal surfaces of the upper aerodigestive tract. These tumours make up more than 95% of the cases of head and neck cancer. Head and neck cancers comprise 3.0% of all new cancers in the UK per year. In the UK the major head and neck sites are the larynx, oral cavity, hypopharynx and oropharynx, which together account for 90% of head and neck squamous cell carcinoma.

Head and neck cancer in the past was predominantly a disease of men and of urban areas. The main identifiable causes of head and neck squamous cell carcinoma are smoking and excessive alcohol consumption. Other factors such as poor dentition and dietary factors may also increase the risk of oral cancer. There is more recently identified a subgroup of patients without the traditional risk factors [1,2]. Recent clinical and molecular investigations suggest that there are distinctive clinical entities, particularly affecting young patients with cancers of the oral tongue and oropharynx. It is therefore possible to prevent the development of cancer by reducing the use of tobacco and alcohol intake. The NHS Cancer Plan [3] and the DOH “Smoking Kills” [4] are investing millions of pounds annually on a rolling programme to reduce cancer overall. There is no mention of head and neck cancer by tumour site in either of these documents! A recent study [5] on delays in diagnosis in head and neck cancers recommends that special educational initiatives should be directed to those patients most at risk and their general practitioners. There continues to be reported evidence that general practitioners, medical and dental, are still in need of further education on the aetiology and health education of patients who may be at risk of developing head and neck cancer [6-8]. The absence of definite early warning signs for most head and neck cancers suggests the need to develop essential screening criteria [9]. It is essential to define the population that is at high risk for head and neck cancer and to subject it to aggressive screening protocols [10].

Making the diagnosis of head and neck cancer is based on clinical symptoms and signs, confirmation of the presence of cancer by histopathology [11]. The current staging of the disease, now universally involves each patient undergoing radiological imaging with computer tomography [CT] and/or magnetic resonance imaging [MRI] to assess the true extent of the disease. Positron emission tomography [PET] may help by evaluation of tumour biological aggression, and, thus, improved staging PET detection of residual or recurrent tumour activity following definitive treatment, by surgery or radiotherapy, may aid with treatment planning [12]. Developments in histopathology have improved the ability to predict which areas of pre-malignancy [leukoplakia] in the oral cavity and the larynx are likely to progress to malignancy and require aggressive therapy and be given long-term follow up clinic observation [13-14]. A recent study [15] of UK waiting times during the management of head and neck tumours showed that there was an inordinate delay in time to radiology, 4.1 – 5.5 weeks, and primary radiotherapy - 10.3 weeks and to surgery – 5.5 weeks. It is recognised that the provision of a head and neck oncology service, currently not widely recognised or resourced in the UK, will require appropriate funding and staffing which is sustainable and recurrent [16].

The untreated head and neck cancer patient is seldom encountered nowadays, but a report from Brazil on 808 patients diagnosed and followed up between 1953 and 1990, showed that approximately 50% of untreated patients died within 4 months, but the remaining patients survived up to 4 or more years following diagnosis [17].
Treatment for head and neck squamous cell carcinomas in the UK is currently by surgery or radiotherapy or a combination of both. In early disease the treatment is tending to move from radiotherapy to conservation surgery [18], while in advanced disease the treatment in the main is surgery with adjuvant radiotherapy. Combined chemoradiation has steadily grown in popularity.

The evidence base for head and neck cancers is low with relatively few randomised controlled trials of the two main treatments. In a review of treatments in three large areas of England, the patterns of surgery and radiotherapy treatments for head and neck cancers were studied and their effects on survival [19]. There was evidence of regional variations in the treatments given and four patients in ten did not receive currently recommended treatments. Better survival was associated with surgery for the mouth cancers, radiotherapy for laryngeal cancers and with a combined treatment regime for pharyngeal cancers independent of tumour and demographic factors. The use of altered fraction schedules of radiotherapy and the addition of chemotherapeutic agents to standard treatments in advanced disease needs to be further explored by the inclusion of patients in randomised controlled trials. A greater research culture in UK head and neck cancer management is required before the practice develops in a haphazard, unregulated fashion [20-22]. The results of aggressive primary treatment including surgery, radiotherapy, or both show that more than 60% of these patients develop loco regional recurrence or distant metastases. Survival rates for recurrent disease are very poor, despite further surgery, radiation therapy, or chemotherapy [23]. The other difficulty with the management of head and neck squamous cell carcinoma patients is the presence of synchronous primary tumours which are present in 1 – 15% and are most commonly associated with an index tumour of the larynx. There is also the continued risk of developing a metachronous second primary malignancy of 4 – 7% per year of survival [24].

Ultimately it is the quantity and quality of the patient’s survival that is the aim of treatment interventions. Where once there was little demonstrable improvement in survival of patients over several decades, recent evidence has been provided which shows improved outcomes [25]. It is of utmost importance to audit and review activity [26] and to evaluate the quality of life [QoL] [27] of all patients who are definitively treated for head and neck cancer.

It is now recognised since the reorganisation of cancer services that care provision is best provided by a structured and weekly run multidisciplinary clinic [MDC] [28]. These clinics require resourcing - adequate space, staff and equipment, to provided optimum convenience and minimal disruption to the patient and their carers where they can be informed of their diagnosis and plan of treatment once the diagnosis of head and neck cancer has been confirmed. It is also recognised that the staff require resources in communication skills training in oncology [29]. Currently, there is a need to define who and what personnel are essential to attend the MDC, and the optimum scheduling of the care discussion and the presence of the patient [30].

To summarise: Head and neck cancer is an uncommon cancer, seldom recognised in the early stages by the patients and their “carers”, and thus requires widespread public health education of its existence to aid with earlier stage disease detection. To diagnose the cancer requires pathologists and radiologists to aid with staging and planning of treatment. The treatment of patients requires a co-ordination of clinicians - surgeons, radiotherapists and oncologists to minimise delays and interruptions to planned treatments. These patients subsequently require rehabilitation of their breathing, eating, swallowing and voice, as well as clinical follow up to detect recurrent, persistent or new disease. The current 5 year survival outlook for a patient diagnosed with a head and neck cancer is more than 60%.
References:

In the 1998 edition of *Effective Head and Neck Cancer Management*, it was recognised that the single most major deficiency in the United Kingdom head and neck cancer management had been the absence of accurate systematic prospective data collection. A prototype minimum dataset was proposed for the UK, to assist in:-

- accreditation of cancer service providers
- prospective audit
- more accurate prognosis
- long term population based observational studies

The minimum dataset seeks to record the minimum amount of data to allow an individual patient and groups of patients to be identified, their management quantified and analysis made of simple defined outcomes.

The essential outcomes being :-

- survival
- complications of treatment
- quality of life measures eg University of Washington QOL

Utilising the minimum of data improves speed of entry, compliance, and accuracy. However every clinician is free to collect additional data to meet local needs and research needs. Within the dataset standard coding practice should be followed, to allow cross comparison of data.

**Development of unified datasets for professions managing head and neck cancer:**

Clinicians from all specialities managing head and neck cancer, representatives from the cancer registries, Office for National Statistics, Department of Health and coding advisors met in March 1998 under the auspices of BAHNO (British Association of Head and Neck Oncologists). From this emerged a multi-agency working group, who built upon the BAO-HNS initiatives outlined above, to define a multidisciplinary dataset to be utilised by all professions managing head and neck cancer. This incorporated the 1998 Report from The Royal College of Pathologists in which a minimum dataset for histopathology reporting in head and neck cancer was described. This group’s report was published in June 1999 – **BAHNO NATIONAL MINIMUM AND ADVISORY HEAD AND NECK CANCER DATA SETS**, which utilised wherever possible international classifications of coding to allow cross-comparison of data. The BAHNO minimum and advisory data sets were formally adopted by BAHNO’s Council and its membership in 1999, and a recent survey has shown a steady rise in their usage. The full text document is available on the BAO-HNS website - www.orl-baohns.org.

It was thus appropriate that BAHNO in May 2000, formed one of the four established clinical areas (lung, colon, breast and head and neck) who contributed to the evolution of a **National Cancer Dataset** with the aim to provide a generic core common to all cancers, with a site specific extension designed to meet the needs of professionals managing head and neck cancer.

The work was commissioned by the National Cancer Director in conjunction with the National Health Service Information Authority (NHSIA) and was delivered by a working group to which BAHNO’s representative contributed and linked to the BAHNO Data Set Working Group.
The **National Cancer Dataset** delivers both the information for performance management required by the NHS, as well as matching the requirements set out above, to achieve co-ordinated data collection, across cancer networks and nationally, meet the requirements for Cancer Registration, and assist clinical teams to better follow the patients journey.

The latest iteration **Version 1.1ISB** can be found on the NHSIA website at [www.nhsia.nhs.uk/cancer](http://www.nhsia.nhs.uk/cancer) and has now successfully received draft NHS Information Standards Board Approval, which is the first step to becoming mandated for all NHS providers to use and report on.

It is accompanied on the website above by a **head and neck cancer data manual** jointly developed with BAHNO defining all items and codes, for head and neck cancer. The Cancer Dataset is intended to be used for all patients identified as having cancer; and all stages of cancer should be included i.e. primary, secondary/recurrences, second primaries. Within this data manual, site specific items for head and neck cancer which fall outside the generic core National Cancer Dataset are defined. These are of two types, those fields common to most cancers’ staging but with their head and neck subsite variants described, and those items felt to be specifically relevant only to head and neck cancer. The latter have been derived from the original BAHNO data set and have been re-evaluated by the dataset working group to ensure their purpose remains and their continuation is justified. Item codes have been modified if required as well as aligning smoking history with the lung group, but including other tobacco usage. A new section has been created on **nutrition** confirming the aims to reflect a multidisciplinary approach, whilst a **section on surgical voice restoration** is in preparation.

An addendum to the manual will be issued to meet any specific requirements from thyroid cancer, which are being examined by a multi-speciality group, with reporting expected in **early 2003**. Companion developments in lymphoma and skin cancer will need to be accommodated.

**KEY SECTIONS IN THE NATIONAL CORE CANCER DATASET for HEAD AND NECK CANCER** (see [www.nhsia.nhs.uk/cancer](http://www.nhsia.nhs.uk/cancer) for latest details)

1. Demographics
2. Referrals
3. Imaging
4. Diagnosis
5. Cancer Care Plan
6. Staging
7. Surgery an other procedures
8. Pathology details –both diagnostic and resective matching to RCPPath reporting
9. Chemotherapy and other drugs
10. Radiotherapy
11. Brachytherapy
12. Palliative care (in development)
13. Clinical trials
14. Clinical status assessment
15. Death details

**HEAD AND NECK SPECIFIC ITEMS SECTION**

- History non head and neck cancer
- Smoking and alcohol history
- Quality of life
- Family history of cancer
- Nutritional support
It is recognised that the development and implementation of this dataset is a significant undertaking for all those involved in the management and delivery of cancer services. It is also recognised that until appropriate IT systems are in place, collection of some data items will be more difficult than others. However, the National Cancer Dataset is a significant step in the drive to improve cancer services and it is only through its collection and use that further development can take place.

The timetable for implementation of the National Cancer dataset is now becoming clearer, with a three phase introduction:

**PHASE 1** – focuses on waiting times information to key referral, diagnosis and treatment points. Information on head and neck cancer will be required to be reported by September 2003.

**PHASE 2** – is based on subset of the full dataset to provide National Comparative Clinical Audit, with waiting times as a component. Introduction will occur between 2003 and 2005 support from the National Clinical support program. Initial piloting of DAHNO (DAta for Head and Neck Oncology) will commence in spring 2003, and then be progressively made available across England. Regular updates will be provided at the BAHNO website. (www.bahno.org) Separate arrangements will apply for Scotland, Wales and Northern Ireland with the exact timing for head and neck cancer yet to be confirmed.

**PHASE 3** – is the implementation of all remaining items.

By 2004 all cancer data will be transmitted to registries electronically.

All colleagues managing head and neck cancer are requested to consider moving to use the National Cancer Dataset for Head and Neck as soon as is practicable. This will support both the move towards universal high quality data collection for every head and neck cancer patient throughout their cancer journey and to achieve the aims of true prospective audit on a routine basis.

A migratory map is available for those wishing to transfer previous NMDS/NADS data to the latest format, as well as paper proformas for those not able to enter electronically at the point of contact.(see NHSIA website)

**DEFINING RESPONSIBILITIES IN HEAD AND NECK CANCER DATA COLLECTION**

To achieve the overall aim of comprehensive head and neck cancer data collection, collation and analysis, wide scale support and ultimately financial provision will be needed. The agenda requires a recognition of the responsibilities outlined below:-

that BAO-HNS will :-
- Continue to liaise with BAHNO in future development of the head and neck specific elements of the National Core Cancer Dataset
- Recommend all its members to follow the National Core Cancer Dataset
- Encourage development of common standards for data transfer to support National Comparative audit
- Liaise with National Clinical Audit support program (NCASP) to develop statistical analysis and regional/central collation

that EACH TRUST/ CANCER NETWORK will:-
- develop a cancer information strategy by Autumn 2002
- Seek to provide hardware/software/provision for data collection input at the point of patient contact
- assist the unit in its responsibilities by providing technical support in computing
- seek solutions to support data transfer where the patients path moves across unit and centre

that EACH HEAD and NECK UNIT will :-
- accept responsibility for organising data collection along the patient pathway/collation / validation / presentation and assist in interpretation for central/regional reporting
that EACH HEAD AND NECK SURGEON/ONCOLOGIST will :-
- facilitate data collection by leading other professionals by example
- ensure data is correct and true
- hold regular local/regional meetings to review surgeon/unit specific data

USEFUL ADDRESSES AND SOURCES OF INFORMATION:

NHS Information Authority website – for details of National core cancer dataset for head and neck, cancer data manual and up to date details on implementation http://www.nhsia.nhs.uk/cancer
BAHNO National Minimum and Advisory Head and Neck Cancer Data Sets Version 1.0 June 1999 Available at www.orl-baohns.org
(Nonepithelial tumours such as those of lymphoid tissue, soft tissue, bone and cartilage are not included.)

C10.1 Anterior (lingual) surface of epiglottis
C32.0 Glottis
C32.1 Supraglottis (laryngeal surface)
C32.2 Subglottis
C32.3 Laryngeal cartilage
C32.8 Overlapping lesion
C32.9 Larynx, NOS

ANATOMY

Primary Site. The following anatomic definition of the larynx allows classification of carcinomas arising in the encompassed mucous membranes but excludes cancers arising on the lateral or posterior pharyngeal wall, pyriform fossa, postcricoid area, or base of tongue. The anterior limit of the larynx is composed of the anterior or lingual surface of the suprahypoid epiglottis, thyrohyoid membrane, the anterior commissure, and the anterior wall of the subglottic region, which is composed of the thyroid cartilage, the cricothyroid membrane, and the anterior arch of the cricoid cartilage. The posterior and lateral limits include the laryngeal aspect of the aryepiglottic folds, the arytenoid region, the interarytenoid space, and the posterior surface of the subglottic space, represented by the mucous membrane covering the surface of the cricoid cartilage. The superolateral limits are composed of the tip and the lateral borders of the epiglottis. The inferior limits are made up of the plane passing through the inferior edge of the cricoid cartilage. For purposes of this clinical stage classification, the larynx is divided into three regions: supraglottis, glottis, and subglottis. The supraglottis is composed of the epiglottis (both its lingual and laryngeal aspects), aryepiglottic folds (laryngeal aspect), arytenoids, and ventricular bands (false cords). The epiglottis is divided for staging purposes into suprahypoid and infrahypoid portions by a plane at the level of the hyoid bone. The inferior boundary of the supraglottis is a horizontal plane passing through the lateral margin of the ventricle at its junction with the superior surface of the vocal cord. The glottis is composed of the true vocal cords, including the anterior and posterior commissures, superior and inferior surfaces. It occupies a horizontal plane 1 cm in thickness, extending inferiorly from the lateral margin of the ventricle. The subglottis is the region extending from the lower boundary of the glottis to the lower margin of the cricoid cartilage.

The division of the larynx is summarized in the following table:

<table>
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<th>Site</th>
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<tr>
<td>Supraglottis</td>
<td>Suprahypoid epiglottis</td>
</tr>
<tr>
<td></td>
<td>Infrahypoid epiglottis</td>
</tr>
<tr>
<td></td>
<td>Aryepiglottic folds (laryngeal aspect)</td>
</tr>
<tr>
<td></td>
<td>Arytenoids</td>
</tr>
<tr>
<td></td>
<td>Ventricular bands (false cords)</td>
</tr>
<tr>
<td>Glottis</td>
<td>True vocal cords including anterior and posterior commissures</td>
</tr>
<tr>
<td>Subglottis</td>
<td>Subglottis</td>
</tr>
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Regional Lymph Nodes. The incidence and distribution of cervical nodal metastases from cancer of the larynx varies with the site of origin and the “T” classification of the primary tumour. The true vocal cords are nearly devoid of lymphatics and tumours of that site alone rarely spread to regional nodes. On the contrary, the supraglottis has a rich and bilaterally interconnected lymphatic network and primary supraglottic cancers are commonly accompanied by regional larynx node spread. Glottic tumours may spread directly to adjacent soft tissues and prelaryngeal, pretracheal, paralaryngeal and paratracheal nodes as well as upper, mid and lower jugular nodes. Supraglottic tumours commonly spread to upper and midjugular nodes, considerably less commonly to submental or submandibular nodes, but occasionally to retropharyngeal nodes.
The rare subglottic primary tumours spread first to adjacent soft tissues and prelaryngeal, pretracheal, paralaryngeal and paratracheal nodes, then to mid and lower jugular nodes. Contralateral lymphatic spread is common. It is recognized that most masses over 3 cm in diameter are not single nodes but are confluent nodes or tumour in soft tissues of the neck. There are three categories of clinically positive nodes: N1, N2, and N3. Midline nodes are considered ipsilateral nodes.

**Metastatic Sites:** Distal spread is common only for patients who have bulky adenopathy. When distant metastases occur, spread to the lungs is most common; skeletal or hepatic metastases occur less often. Mediastinal lymph node metastases are considered distal metastases.

**RULES FOR CLASSIFICATION**

**Clinical Staging.** The assessment of the larynx is accomplished primarily by inspection, using indirect mirror and direct endoscopic examination. The tumour must be confirmed histologically and any other data obtained by biopsies may be included. Cross-sectional imaging in laryngeal carcinoma is recommended when the primary tumour extent is in question based upon clinical examination. Radiologic nodal staging should be done simultaneously to supplement clinical examination. Complete endoscopy under general anesthesia is usually performed after completion of other diagnostic studies to accurately assess, document and biopsy the tumour.

**Pathological Staging.** Pathologic staging requires the use of all information obtained in the clinical staging in addition to histologic study of the surgically resected specimen. The surgeon’s evaluation of gross unresected residual tumour must also be included. Specimens that are resected after radiation or chemotherapy need to be identified and considered in context. The pathologic description of any lymphadenectomy specimen should describe size, number, position of the involved node(s), and the presence or absence of extracapsular extension.

**DEFINITION OF TNM**

**Primary Tumour (T)**

| TX | Primary tumour cannot be assessed |
| T0 | No evidence of primary tumour |
| Tis | Carcinoma in situ |

**Supraglottis**

| T1 | Tumour limited to one subsite of supraglottis with normal vocal cord mobility |
| T2 | Tumour invades mucosa of more than one adjacent subsite of supraglottis or glottis or region outside the supraglottis (e.g. mucosa of base of tongue, vallecula, medial wall of pyriform sinus) without fixation of the larynx. |
| T3 | Tumour limited to larynx with vocal cord fixation and/or invades any of the following postcricoid area, pre-epiglottic tissues, paraglottic space, and/or minor thyroid cartilage erosion (e.g. inner cortex). |
| T4A | Tumour invades through the thyroid cartilage and or invades tissues beyond the larynx (e.g., trachea, soft tissues of neck including deep extrinsic muscle of the tongue, strap muscles, thyroid, or esophagus). |
| T4B | Tumour invades prevertebral space or encases carotid artery or invades mediastinal structures |

**Glottis**

| T1 | Tumour limited to the vocal cord(s) (may involve anterior or posterior commissure with normal mobility |
| T1a | Tumour limited to one vocal cord |
| T1b | Tumour involves both vocal cords |
| T2 | Tumour extends to supraglottis and/or subglottis, and/or with impaired vocal cord mobility |
| T3 | Tumour limited to the larynx with vocal cord fixation, and/or invades paraglottic space, and or minor thyroid cartilage erosion (e.g. inner cortex). |
| T4a | Tumour invades through the thyroid cartilage and/or invades tissues beyond the larynx (e.g., trachea, soft tissues of neck including deep extrinsic muscle of the tongue, strap muscles, thyroid or esophagus). |
| T4b | Tumour invades prevertebral space or encases carotid artery or invades mediastinal structures |
**Subglottis**

T1  Tumour limited to the subglottis
T2  Tumour extends to vocal cord(s) with normal or impaired mobility
T3  Tumour limited to larynx with vocal cord fixation
T4a Tumour invades cricoid or thyroid cartilage and/or invades tissues beyond the larynx (e.g., trachea, soft tissues of neck including deep extrinsic muscles of the tongue, strap muscles, thyroid or esophagus)
T4b Tumour invades prevertebral space or encases carotid artery or invades mediastinal structures

**STAGE GROUPING**

<table>
<thead>
<tr>
<th>Stage</th>
<th>Ti</th>
<th>N</th>
<th>M</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stage 0</td>
<td>Tis</td>
<td>N0</td>
<td>M0</td>
</tr>
<tr>
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<td>M0</td>
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<td>T3</td>
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<td>M0</td>
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<td>M0</td>
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<tr>
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<tr>
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<tr>
<td>Stage IVA</td>
<td>T4a</td>
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<td>N3</td>
</tr>
<tr>
<td>Stage IVC</td>
<td>Any</td>
<td>Any</td>
<td>N</td>
</tr>
</tbody>
</table>

**HISTOPATHOLOGIC TYPE**

The predominant cancer is squamous cell carcinoma. The staging guidelines are applicable to all forms of carcinoma. Nonepithelial tumours such as those of lymphoid tissue, soft tissue, bone and cartilage (i.e., lymphoma, melanoma, and sarcoma) are not included. Histologic confirmation of diagnosis is required.

Histopathologic grading of squamous carcinoma is recommended: the grade is subjective and uses a descriptive as well as numerical form, i.e., well differentiated, moderately differentiated, and poorly differentiated, depending upon the degree of closeness to or deviation from squamous epithelium in mucosal sites. Also recommended where feasible is a quantitative evaluation of depth of invasion of the primary tumour and the presence or absence of vascular invasion and perineural invasion. Although the grade of tumour does not enter into the staging of the tumour, it should be recorded. The pathologic description of any lymphadenectomy specimen should describe the size, number, position of the involved node(s) and the presence or absence of extracapsular extension.

**HISTOPATHOLOGIC GRADE (G)**

<table>
<thead>
<tr>
<th>Grade</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>GX</td>
<td>Grade cannot be assessed</td>
</tr>
<tr>
<td>G1</td>
<td>Well differentiated</td>
</tr>
<tr>
<td>G2</td>
<td>Moderately differentiated</td>
</tr>
<tr>
<td>G3</td>
<td>Poorly differentiated</td>
</tr>
</tbody>
</table>
TNM Classification: AJCC 2002-3
Head and Neck Sites

The T classifications indicating the extent of the head and neck primary tumour differ in specific details. The N classification for cervical lymph node metastasis is uniform for all mucosal sites except nasopharynx. The N classifications for thyroid and nasopharynx are unique to those sites and are based upon tumour behaviour and prognosis. The clinical staging systems presented in this section are all clinical staging, based on the best possible estimate of the extent of the disease before first treatment. Imaging techniques (computed tomography (CT), magnetic resonance imaging (MRI), ultrasonography) may be applied, and in more advanced tumour stages, have added to the accuracy of primary (T) and nodal (N) staging, especially in the nasopharyngeal, paranasal sinuses and regional lymph nodal areas. Appropriate imaging studies should be obtained whenever the clinical findings are uncertain. Similarly, endoscopic evaluation of the primary tumour, when appropriate, is desirable for detailed assessment of the primary tumour for accurate “T” staging. Fine needle aspiration biopsy (FNAB) may confirm the presence of tumour and its histopathologic nature, but cannot rule out the presence of tumour.

Any diagnostic information which contributes to the overall accuracy of the pretreatment assessment should be considered in clinical staging and treatment planning. When surgical treatment is carried out, cancer of the head and neck can be staged (pathologic stage (pTNM) using all information available from clinical assessment as well as from the pathologic study of the resected specimen. The pathologic stage does not replace the clinical stage, which should be reported as well.

In reviewing the staging systems, several changes in the T classifications as well as stage groupings are made to reflect current practices of treatment, clinical relevance and contemporary data. Uniform “T” staging for oral cavity, oropharynx, salivary and thyroid cancers greatly simplifies the system and will improve compliance by clinicians. T4 tumours are subdivided into advanced resectable (T4a) and advanced unresectable (T4b) categories. Regrouping of Stage IV disease for all sites into advanced resectable (Stage IVA), advanced unresectable (Stage IVB), and distant metastatic (Stage IVC) also simplifies advanced disease staging.

A major revision of the nasopharynx classification has been stimulated by clinical experience from several Asian sources.

Regional Lymph Nodes. The status of the regional lymph nodes in head and neck cancer is of such prognostic importance that the cervical nodes must be assessed for each patient and tumour. The lymph nodes may be subdivided into specific anatomic subsites and grouped into seven levels for ease of description.

<table>
<thead>
<tr>
<th>Level</th>
<th>Subsite</th>
</tr>
</thead>
<tbody>
<tr>
<td>Level I:</td>
<td>Submental</td>
</tr>
<tr>
<td></td>
<td>Submandibular</td>
</tr>
<tr>
<td>Level II:</td>
<td>Upper jugular</td>
</tr>
<tr>
<td>Level III:</td>
<td>Midjugular</td>
</tr>
<tr>
<td>Level IV:</td>
<td>Lower jugular</td>
</tr>
<tr>
<td>Level V:</td>
<td>Posterior triangle (Spinal accessory and transverse cervical)</td>
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<tr>
<td></td>
<td>(Upper, mid and lower, corresponding to the levels that define upper,</td>
</tr>
<tr>
<td></td>
<td>mid, and lower jugular nodes)</td>
</tr>
<tr>
<td>Level VI:</td>
<td>Prelaryngeal (Delphian)</td>
</tr>
<tr>
<td></td>
<td>Pretracheal</td>
</tr>
<tr>
<td></td>
<td>Paratracheal</td>
</tr>
<tr>
<td>Level VII:</td>
<td>Upper mediastinal</td>
</tr>
</tbody>
</table>

Other groups: Retropharyngeal
Parapharyngeal
Buccinator (facial)
Preauricular
FIG. 1/2 Schematic diagram indicating the location of the lymph node levels in the neck as described in the text.

The location of the lymph node levels conforms to the following clinical descriptions which also correlate with surgical landmarks at the time of surgical neck exploration.

Level I: Contains the submental and submandibular triangles bounded by the anterior and posterior bellies of the digastric muscle, the hyoid bone inferiorly and the body of the mandible superiorly.

Level II: Contains the upper jugular lymph nodes and extends from the level of the skull base superiorly to the hyoid bone inferiorly.

Level III: Contains the middle jugular lymph nodes from the hyoid bone superiorly to the level of the lower border of the cricoid cartilage inferiorly.

Level IV: Contains the lower jugular lymph nodes from the level of the cricoid cartilage superiorly to the clavicle inferiorly.

Level V: Contains the lymph nodes in the posterior triangle bounded by the anterior border of the trapezius muscle posteriorly, the posterior border of the sternocleidomastoid muscle anteriorly, and the clavicle inferiorly. For descriptive purposes, Level V may be further subdivided into upper, middle, or lower levels corresponding to the superior and inferior planes that define Levels II, III, and IV.

Level VI: Contains the lymph nodes of the anterior central compartment from the hyoid bone superiorly to the suprasternal notch inferiorly. On each side, the lateral boundary is formed by the medial border of the carotid sheath.

Level VII: Contains the lymph nodes inferior to the suprasternal notch in the superior mediastinum.

The pattern of the lymphatic drainage varies for different anatomic sites. However, the location of the lymph node metastases has prognostic significance in patients with squamous cell carcinoma of the head and neck. Survival is significantly worse when metastases involve lymph nodes beyond the first echelon of lymphatic drainage and, particularly, lymph nodes in the lower regions of the neck, i.e. Level IV and Level V (supraclavicular region). Consequently, it is recommended that each “N” staging category be recorded to show, in addition to the established parameters, whether the nodes involved are located in the upper (U) or lower (L) regions of the neck, depending upon their location above or below the level of the cricoid cartilage.

The natural history and response to treatment of cervical nodal metastases from nasopharynx primary sites is different, in terms of its impact on prognosis, and, therefore, justifies a different “N” classification scheme. Regional node metastases from well differentiated thyroid cancer do not significantly impact upon the ultimate prognosis and, therefore, also justify a unique staging system for thyroid cancers.

Histopathologic examination is necessary to exclude the presence of tumour in lymph nodes. No imaging study (as yet) can identify microscopic tumour foci in regional nodes or distinguish between small reactive nodes and small malignant nodes.

When enlarged lymph nodes are detected, the actual size of the nodal mass(es) should be measured. It is recognized that most masses over 3 cm in diameter are not single nodes but are confluent nodes or tumour in soft tissues of the neck. Imaging studies showing amorphous spiculated margins of involved nodes or involvement of internodal fat resulting in loss of normal oval to round nodal shape strongly suggest extracapsular (extranodal) tumour spread. Pathologic examination is necessary for documentation of tumour extent in terms of the location or level of the lymph node(s) involved, the number of nodes containing metastases and the presence or absence of extracapsular spread of tumour.
For pN, a selective neck dissection will ordinarily include 6 or more lymph nodes and a radical or modified radical neck dissection will ordinarily include 10 or more lymph nodes. Negative pathologic examination of a lesser number of nodes still mandates a pN0 designation.

Metastatic Sites. The most common sites of distant spread are in the lungs and bones; hepatic or brain metastases occur less often. Mediastinal lymph node metastases are considered distant metastases.

Regional Lymph Nodes (N)

NX Regional lymph nodes cannot be assessed
N0 No regional lymph node metastasis
N1 U L Metastasis in a single ipsilateral lymph node, 3 cm or less in greatest dimension
N2 U L Metastasis in a single ipsilateral lymph node, more than 3 cm but not more than 6 cm in greatest dimension; or in multiple ipsilateral lymph nodes, none more than 6 cm in greatest dimension; or in bilateral or contralateral lymph nodes, none more than 6 cm in greatest dimension
N2a Metastasis in single ipsilateral lymph node more than 3 cm but not more than 6 cm in greatest dimension
N2b Metastasis in multiple ipsilateral lymph nodes, none more than 6 cm in greatest dimension
N2c Metastasis in bilateral or contralateral lymph nodes, none more than 6 cm in greatest dimension
N3 U L Metastasis in a lymph node more than 6 cm in greatest dimension

U = Upper neck: Metastasis located above the level of the cricoid cartilage
L = Lower neck: Metastasis located below the level of the cricoid cartilage

Distant Metastasis (M)

MX Distant metastasis cannot be assessed
M0 No distant metastasis
M1 Distant metastasis

References:
TNM Classification: AJCC 2002 –3
Lip and Oral Cavity

(Nonepithelial tumours such as those of lymphoid tissue, soft tissue, bone, and cartilage are not included.)

Anatomical Sites and Subsites

Lip
External upper lip (vermilion border) (C00.0)
External lower lip (vermilion border) (C00.1)
Commissures (C00.6)
Oral cavity
Buccal mucosa
  - mucosa of the upper and lower lips (C00.3, 4)
  - cheek mucosa (C06.0)
  - retromolar areas (C06.2)
  - bucco-alveolar sulci, upper and lower (vestibule of mouth) (C06.1)
Upper alveolus and gingiva (upper gum) (C03.0)
Lower alveolus and gingiva (lower gum) (C03.1)
Hard palate (C05.0)
Tongue
  - dorsal surface and lateral borders anterior to vallate papillae (anterior two-thirds) (C02.0, 1)
  - inferior (ventral) surface (C02.2)
Floor of mouth (C04)

ANATOMY

Primary Site. The oral cavity extends from the skin-vermilion junction of the lips to the junction of the hard and soft palate above and to the line of circumvallate papillae below and is divided into the following specific areas:

Mucosal Lip. The lip begins at the junction of the vermilion border with the skin and includes only the vermilion surface or that portion of the lip that comes into contact with the opposing lip. It is well defined into an upper and lower lip joined at the commissures of the mouth.

Buccal Mucosa. This includes all the membrane lining of the inner surface of the cheeks and lips from the line of contact of the opposing lips to the line of attachment of mucosa of the alveolar ridge (upper and lower) and pterygo-mandibular raphe.

Lower Alveolar Ridge. This refers to the mucosa overlying the alveolar process of the mandible which extends from the line of attachment of mucosa in the buccal gutter to the line of free mucosa of the floor of the mouth. Posteriorly it extends to the ascending ramus of the mandible.

Upper Alveolar Ridge. This refers to the mucosa overlying the alveolar process of the maxilla which extends from the line of attachment of mucosa in the upper gingival buccal gutter to the junction of the hard palate. Its posterior margin is the upper end of the pterygopalatine arch.

Retromolar Gingiva (Retromolar Trigone). This is the attached mucosa overlying the ascending ramus of the mandible from the level of the posterior surface of the last molar tooth to the apex superiorly, adjacent to the tuberosity of the maxilla.

Floor of the Mouth. This is a semilunar space over the mylohyoid and hyoglossus muscles, extending from the inner surface of the lower alveolar ridge to the undersurface of the tongue. Its posterior boundary is the base of the anterior pillar of the tonsil. It is divided into two sides by the frenulum of the tongue and contains the ostia of the submaxillary and sublingual salivary glands.
**Hard Palate.** This is the semilunar area between the upper alveolar ridge and the mucous membrane covering the palatine process of the maxillary palatine bones. It extends from the inner surface of the superior alveolar ridge to the posterior edge of the palatine bone.

**Anterior Two-Thirds of the Tongue (Oral Tongue).** This is a freely mobile portion of the tongue that extends anteriorly from the line of circumvallate papillae to the undersurface of the tongue at the junction of the floor of the mouth. It is composed of four areas: the tip, the lateral borders, the dorsum, and the undersurface (nonvillus ventral surface of the tongue). The undersurface of the tongue is considered as a separate category by the World Health Organization (WHO).

**Regional Lymph Nodes.** Mucosal cancer of the oral cavity may spread to regional lymph node(s). Tumours of each anatomic site have their own predictable patterns of regional spread. The risk of regional metastasis generally relates to the T category and probably more importantly to the depth of infiltration of the primary tumours. Cancer of the lip carries a low metastatic risk and initially involves adjacent submental and submandibular nodes, then jugular nodes. Cancers of the hard palate and alveolar ridge likewise have a low metastatic potential and involve buccinator, submandibular, jugular and occasionally retropharyngeal nodes. Other oral cancers will primarily spread to submandibular and jugular nodes, uncommonly posterior triangle/supraclavicular nodes. Cancer of the anterior oral tongue may spread directly to lower jugular nodes. The closer to the midline the primary, the greater the risk of bilateral cervical nodal spread. Any previous treatment to the neck, surgical and/or radiation, may alter normal lymphatic drainage patterns resulting in unusual distribution of regional spread of disease to the cervical lymph nodes. In general, cervical lymph node involvement from oral cavity primary sites is predictable and orderly, spreading from the primary to upper, then middle, and subsequently lower cervical nodes. However, disease in the anterior oral cavity may also spread directly to the midcervical lymph nodes. The risk of distant metastasis is more dependent upon the "N" than the "T" status of the head and neck cancer.

**Metastatic Sites.** The lungs are the commonest site of distant metastases; skeletal or hepatic metastases occur less often. Mediastinal lymph node metastases are considered distant metastases.

**RULES FOR CLASSIFICATION**

**Clinical Staging.** The assessment of the primary tumour is based upon inspection and palpation of the oral cavity and neck. Additional studies may include CT or MRI. When imaging is utilized one study will generally suffice to evaluate primary and nodal tumour extent. Clinical assessment of extent of mucosal involvement is more accurate than is radiographic assessment. The radiographic estimate of deep tissue extent and of regional lymph node involvement is usually more accurate than clinical assessment. MRI is generally more revealing of extent of soft tissue, perivascular and perineural spread, skull base involvement and intracranial tumour extension. On the other hand, high resolution CT with contrast will often provide similar information if carefully done and will provide better images of bone and larynx detail and is minimally affected by motion. CT or MR imaging may be more useful in evaluation of advanced tumours for assessment of bone invasion (mandible or maxilla) and deep tissue invasion (deep extrinsic tongue muscles, midline tongue, soft tissues of neck). Clinical examination supplemented with dental films or panoramic X-rays may be helpful in determining cortical bone involvement. If CT or MR imaging is undertaken for primary tumour evaluation, radiologic assessment of nodal involvement should also be done simultaneously. For lesions of an advanced extent appropriate screening for distant metastases should be considered. Ultrasonography may be helpful in assessment of major vascular invasion as an adjunctive test. The tumour must be confirmed histologically. All clinical, imaging, and pathologic data available prior to first definitive treatment may be used for clinical staging.

**Pathologic Staging.** Complete resection of the primary site and/or regional nodal dissections followed by pathologic examination of the resected specimen(s) allow the use of this designation for pT and/or pN, respectively. Specimens that are resected after radiation or chemotherapy need to be identified and considered in context. pT is derived from the actual measurement of the unfixed tumour in the surgical specimen. It should be noted, however, that up to 30% shrinkage of soft tissues may occur in the resected specimen. Pathological staging represents additional and important information and should be included as such in staging, but does not supplant clinical staging as the primary staging scheme.
DEFINITION OF TNM

Primary Tumour (T)

TX Primary tumour cannot be assessed
T0 No evidence of primary tumour
Tis Carcinoma in situ
T1 Tumour 2 cm or less in greatest dimension
T2 Tumour more than 2 cm but not more than 4 cm in greatest dimension
T3 Tumour more than 4 cm in greatest dimension
T4 (lip) Tumour invades adjacent structures (e.g., through cortical bone, inferior alveolar nerve, floor of mouth, skin of face, i.e. chin or nose)
T4a (oral cavity) Tumour invades adjacent structures (e.g., through cortical bone, into deep [extrinsic] muscle of tongue (genioglossus, hyoglossus, palatoglossus, and styloglossus), maxillary sinus, skin of face. Superficial erosion alone of bone/tooth socket by gingival primary is not sufficient to classify as T4)
T4b Tumour invades masticator space, pterygoid plates, skull base and internal carotid artery

NOTE: Superficial erosion alone of bone/tooth socket by gingival primary is not sufficient to classify a tumour as T4.

STAGE GROUPING – Identical to Larynx

HISTOPATHOLOGIC TYPE

The predominant cancer is squamous cell carcinoma. The staging guidelines are applicable to all forms of carcinoma. Nonepithelial tumours such as those of lymphoid tissue, soft tissue, bone and cartilage (i.e., lymphoma, melanoma, and sarcoma) are not included. Histologic confirmation of diagnosis is required. Histopathologic grading of squamous carcinoma is recommended; the grade is subjective and uses a descriptive as well as numerical form, i.e., well, moderately well, and poorly differentiated, depending upon the degree of closeness to or deviation from squamous epithelium in mucosal sites. Also recommended is a quantitative evaluation of depth of invasion of the primary tumour and the presence or absence of vascular invasion and perineural invasion.

Characteristics of Tumours

Endophytic. The measurement using an ocular micrometer is taken perpendicular from the surface of the invasive squamous cell carcinoma (A) to the deepest area of involvement (B) and recorded in millimeters. The measurement should not be done on tangential sections or in lesions without a clearly recognizable surface component.

Exophytic. The measurement which is better characterized as tumour thickness rather than depth of invasion is taken from the surface (A) to the deepest area (B).

Ulcerated. The measurement is taken from the ulcer base (A) to the deepest area (B) as well as from the surface of the most lateral extent of the invasive carcinoma (C) to the deepest area (D).

Depth of tumour invasion (mm) should be recorded. Depth is NOT used for T staging. Although the grade of the tumour does not enter into staging of the tumour, it should be recorded. The pathologic description of any lymphadenectomy specimen should describe the size, number, and position of involved lymph node(s), and the presence or absence of extracapsular extension.

HISTOPATHOLOGIC GRADE (G)

GX Grade cannot be assessed
G1 Well differentiated
G2 Moderately differentiated
G3 Poorly differentiated
PROGNOSTIC FACTORS

In addition to the importance of the TNM factors outlined previously, the overall health of these patients clearly influences outcome. Comorbidity can be classified by more general measures, such as the Karnofsky Performance status (KPS), or more specific measures, such as the Kaplan-Feinstein Index. The KPS provides a uniform, objective assessment of an individual’s functional status. The scale, in 10 point increments from 0 (dead) to 100 (normal, no complaints, no evidence of disease), was devised in 1948 by David A. Karnofsky. The KPS is a reliable, independent predictor of survival outcome for patients with solid tumors and, therefore, is a required baseline assessment in clinical protocols in head and neck and other cancers.

The AJCC strongly recommends recording of KPS along with standard staging information.

Karnofsky Scale: Criteria of Performance Status (PS)

<table>
<thead>
<tr>
<th>Score</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>100</td>
<td>Normal; no complaints; no evidence of disease.</td>
</tr>
<tr>
<td>90</td>
<td>Able to carry on normal activity; minor signs or symptoms of disease</td>
</tr>
<tr>
<td>80</td>
<td>Able to carry on normal activity with effort; some signs or symptoms of disease</td>
</tr>
<tr>
<td>70</td>
<td>Cares for self; unable to carry on normal activity or do active work.</td>
</tr>
<tr>
<td>60</td>
<td>Requires occasional assistance but is able to care for most of own needs.</td>
</tr>
<tr>
<td>50</td>
<td>Requires considerable assistance and frequent medical care.</td>
</tr>
</tbody>
</table>

Disabled; requires special care and assistance.

Diagnosis and treatment of depression may also aid in symptom control and improved quality of life. Continued exposure to carcinogens, such as alcohol and tobacco smoke, likely also affects patients' outcome adversely.
TNM Classification: AJCC 2002–3
Major Salivary Glands
(Parotid, Submandibular and Sublingual)

C07.9 Parotid gland
C08.0 Submandibular gland
C08.1 Sublingual gland
C08.8 Overlapping lesion of major salivary glands
C08.9 Major salivary gland, NOS

This staging system is based on an extensive retrospective review of the world literature regarding malignant tumors of the major salivary glands.

Numerous factors affect patient survival, including the histologic diagnosis, cellular differentiation of the tumor (grade), site, size, degree of fixation, or local extension, facial nerve involvement and the status of regional lymph nodes as well as distant metastases. The classification here proposed involves the four dominant clinical variables: tumor size, local extension of the tumor, nodal metastasis, and distant metastasis. The T4 category has been subdivided (T4a, T4b). T4a indicates advanced lesions which are resectable with grossly clear margins; T4b reflects extension to areas which preclude resection with clear margins. Histologic grade, patient age and tumor site are important additional factors that should be recorded for future analysis and potential inclusion in the staging system.

ANATOMY

Primary Site The major salivary glands include the parotid, submandibular and sublingual glands.

Tumors arising in minor salivary glands (mucous-secreting glands in the lining membrane of the upper aerodigestive tract) are properly staged according to the anatomic site of origin (e.g., oral cavity, sinuses, etc). Primary tumors of the parotid comprise the largest proportion of salivary gland tumors. Sublingual primary cancers are rare and may be difficult to distinguish with certainty from minor salivary gland primary tumors of the anterior floor of the mouth.

Regional Lymph Nodes Regional lymphatic spread from salivary gland cancer is less common than from head and neck mucosal squamous cancers and varies according to the histology and size of the primary tumor. Most nodal metastases will be clinically apparent on initial evaluation. Low-grade tumors rarely metastasize to regional nodes, while the risk of regional spread is substantially higher from the high-grade cancers. Regional dissemination tends to be orderly, progressing from intraglandular to adjacent (periparotid, submandibular nodes) then to upper and midjugular, and occasionally retropharyngeal nodes. Bilateral lymphatic spread is rare.

Metastatic Sites Distant spread is most frequently to the lungs.

RULES FOR CLASSIFICATION

Clinical Staging The assessment of primary salivary gland tumors includes a pertinent history (pain, trismus, etc), inspection, palpation and evaluation of the cranial nerves. Radiologic studies may add valuable information for staging. The soft tissues of the neck from the skull base to the hyoid bone must be studied with the lower neck included whenever lymph node metastases are suspected. Images of the intratemporal facial nerve are critical to the identification of perineural tumor in this area. Cancers of the submandibular and sublingual salivary glands merit cross-sectional imaging. Computed tomography (CT) or MRI may be useful in assessing extent of deep extraglandular tumor, bone invasion, deep tissue extent (extrinsic tongue muscle and/or soft tissues of the neck).

Pathologic staging. The surgical pathology report and all other available data should be used to assign a pathologic classification to those patients who have resection of the cancer.
DEFINITION OF TNM

Primary Tumor (T)

TX Primary tumor cannot be assessed
T0 No evidence of primary tumor
T1 Tumor 2 cm or less in greatest dimension without extraparenchymal extension*
T2 Tumor more than 2 cm but not more than 4 cm in greatest dimension without extraparenchymal extension*
T3 Tumor more than 4 cm and/or tumor having extraparenchymal extension*
T4a Tumor invades skin, mandible, ear canal, and/or facial nerve
T4b Tumor invades skull base, pterygoid plates, and/or carotid artery

Note: Extraparenchymal extension is clinical or macroscopic evidence of invasion of soft tissues or facial Nerve.

STAGE GROUPING - Identical to Larynx

HISTOPATHOLOGIC TYPE

The suggested histopathologic typing is that proposed by the World Health Organization.

Acinic cell carcinoma
Mucoepidermoid carcinoma
Adenoid cystic carcinoma
Polymorphous low grade adenocarcinoma
Epithelial-myoepithelial carcinoma
Basal cell adenocarcinoma
Sebaceous carcinoma
Papillary cystadenocarcinoma
Mucinous adenocarcinoma
Oncocytic carcinoma
Salivary duct carcinoma
Adenocarcinoma
Myoepithelial carcinoma
Carcinoma ex pleomorphic adenoma
Squamous cell carcinoma
Small cell carcinoma
Other carcinomas
HISTOLOGIC GRADE (G)

Histologic grading is applicable only to some types of salivary cancer:

Mucoepidermoid carcinoma, adenocarcinoma not otherwise specified, or when either of these is the carcinomatous element of carcinoma ex pleomorphic adenoma.

In most instances, the histologic type defines the grade (i.e. salivary duct carcinoma is high grade; basal cell adenocarcinoma is low grade).

TNM Classification: AJCC 2002 -3
Paranasal Sinuses

(Nonepithelial tumours such as those of lymphoid tissue, soft tissue, bone and cartilage are not included)
C30.0 Nasal cavity
C31.0 Maxillary sinus
C31.1 Ethmoid sinus

ANATOMY

Primary Sites. Cancer of the maxillary sinus is the most common of the sinonasal malignancies. Ethmoid sinus and nasal cavity cancers are equal in frequency, but considerably less common than maxillary sinus cancers. Tumours of the sphenoid and frontal sinuses are so rare as not to warrant staging.

The location, as well as the extent, of the mucosal lesion within the maxillary antrum has prognostic significance. Historically, Ohngren’s line, connecting the medial canthus of the eye to the angle of the mandible, is used to divide the maxillary antrum into an anteroinferior portion (infrastructure) which is associated with a good prognosis and a superoposterior portion (suprastructure) which has a poor prognosis (Fig. 1 a&b). The poorer outcome associated with superoposterior cancers reflects early access of these tumours to critical structures including the eye, skull base, pterygoids, and infratemporal fossa.

For the purpose of staging, the nasoethmoidal complex is divided into two sites, nasal cavity and ethmoid sinuses. The ethmoids are further subdivided two sub-sites, left and right, separated by the nasal septum. The nasal cavity is divided into four sub-sites, the septum, floor, lateral wall and vestibule.

<table>
<thead>
<tr>
<th>Site</th>
<th>Subsite</th>
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<tbody>
<tr>
<td>Nasal Cavity</td>
<td>Septum</td>
</tr>
<tr>
<td></td>
<td>Floor</td>
</tr>
<tr>
<td></td>
<td>Lateral wall</td>
</tr>
<tr>
<td></td>
<td>Vestibule</td>
</tr>
<tr>
<td>Ethmoid sinus</td>
<td>Left</td>
</tr>
<tr>
<td></td>
<td>Right</td>
</tr>
</tbody>
</table>
**Regional Lymph Nodes.** Regional lymph node spread from cancer of paranasal origin is relatively uncommon. Involvement of buccinator, submandibular, upper jugular and occasionally retropharyngeal nodes may occur with advanced maxillary sinus cancer, particularly those extending beyond the sinus walls to involve adjacent structures including soft tissues of the cheek, upper alveolus, palate, and buccal mucosa. Ethmoid sinus cancers are less prone to regional lymphatic spread. When only one side of the neck is involved, it should be considered ipsilateral. Bilateral spread may occur with advanced primary cancer, particularly with spread of the primary beyond the midline.

**Metastatic Sites.** Distant spread to lungs is most common; occasionally there is spread to bone.

**RULES FOR CLASSIFICATION**

**Clinical Staging.** The assessment of primary maxillary antrum, nasal cavity and ethmoid tumours is based on inspection and palpation including examination of the orbits, nasal and oral cavities, nasopharynx, and neurologic evaluation of the cranial nerves. Nasal endoscopy with rigid or fibroptic flexible instruments is recommended. Radiological assessment with magnetic resonance imaging (MRI) or computed tomography (CT) is mandatory for accurate pretreatment staging of malignant tumor of the sinuses. If available, MRI more accurately depicts skull base and intracranial involvement and differentiation of fluid from solid tumor. Neck nodes are assessed by palpation +/- imaging. Imaging for possible nodal metastases is probably unnecessary in the presence of a clinically negative neck. Examinations for distant metastases include appropriate radiographs, blood chemistries, blood count and other routine studies as indicated.

**Pathological Staging.** Pathologic staging requires the use of all information obtained in the clinical staging in addition to histologic study of the surgically resected specimen. The surgeon’s evaluation of gross unresected residual tumor must also be included. Specimens that are resected after radiation or chemotherapy need to be identified and considered in context. The pathologic description of the lymphadenectomy specimen should describe the size, number, position of the involved node(s), and the presence or absence of extracapsular extension.

**DEFINITION OF TNM**

**Maxillary Sinus**

**Primary Tumor (T)**

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>TX</td>
<td>Primary tumor cannot be assessed</td>
</tr>
<tr>
<td>T0</td>
<td>No evidence of primary tumor</td>
</tr>
<tr>
<td>Tis</td>
<td>Carcinoma in situ</td>
</tr>
<tr>
<td>T1</td>
<td>Tumor limited to the antral mucosa with no erosion or destruction of bone</td>
</tr>
<tr>
<td>T2</td>
<td>Tumor causing bone erosion or destruction including extension into the hard palate and/or middle nasal meatus, except extension to posterior antral wall of maxillary sinus and pterygoid plates</td>
</tr>
<tr>
<td>T3</td>
<td>Tumor invades any of the following: bone of the posterior wall of maxillary sinus, subcutaneous tissues, floor or medial wall of orbit, pterygoid fossa, ethmoid sinuses</td>
</tr>
<tr>
<td>T4a</td>
<td>Tumor invades anterior orbital contents, skin of cheek, pterygoid plates, infratemporal fossa, cribriform plate, sphenoid or frontal sinuses</td>
</tr>
<tr>
<td>T4b</td>
<td>Tumor invades any of the following: orbital apex, dura, brain, middle cranial fossa, cranial nerves other than maxillary division of trigeminal nerve, nasopharynx, and/or clivus</td>
</tr>
</tbody>
</table>
Nasal Cavity and Ethmoid Sinus

Primary Tumor (T)

T
- Primary tumor cannot be assessed

T0
- No evidence of primary tumor

Tis
- Carcinoma in situ

T1
- Tumor restricted to any one sub-site, with or without bony invasion

T2
- Tumor invading two sub-sites in a single region or extending to involve an adjacent region within the nasoethmoidal complex, with or without bony invasion

T3
- Tumor extends to invade the medial wall or floor of the orbit, maxillary sinus, palate, or cribriform plate

T4a
- Tumor invades any of the following: anterior orbital contents, skin of nose or cheek, minimal extension to anterior cranial fossa, pterygoid plates, sphenoid or frontal sinuses

T4b
- Tumor invades any of the following: orbital apex, dura, brain, middle cranial fossa, cranial nerves other than nasopharynx, or clivus

STAGE GROUPING - Identical to Larynx

HISTOPATHOLOGIC TYPE

The predominant cancer is squamous cell carcinoma. The staging guidelines are applicable to all forms of carcinoma. Nonepithelial tumours such as those of lymphoid tissue, soft tissue, bone and cartilage are not included. Histologic confirmation of diagnosis is required. Histopathologic grading of squamous carcinoma is recommended. The grade is subjective and uses descriptive as well as a numerical i.e., well differentiated, moderately differentiated, and poorly differentiated depending upon the degree of closeness to, or deviation from, squamous epithelium in mucosal sites. Also recommended where feasible is a quantitative evaluation of depth of invasion of the primary tumor and the presence or absence of vascular invasion and perineural invasion. Although the grade of the tumor does not enter into the staging of the tumor, it should be recorded. The pathologic description of any lymphadenectomy specimen should describe the size, number, position of the involved node(s), and the presence or absence of extracapsular extension.

HISTOPATHOLOGIC GRADE (G)

GX
- Grade cannot be assessed

G1
- Well differentiated

G2
- Moderately differentiated

G3
- Poorly differentiated

PROGNOSTIC FACTORS

In addition to the importance of the TNM factors outlined previously, the overall health of these patients clearly influences outcome. Comorbidity can be classified by more general measures, such as the Karnofsky performance score, or more specific measures such as the Kaplan-Feinstein Index or the Charlson Index, and increase in incidence and severity with increasing age. Continued exposure to carcinogens such as alcohol and tobacco smoke likely also affects patient’s outcomes adversely.
TNM Classification: AJCC 2002 –3
Pharynx
(Including base of tongue, soft palate and uvula)

(Nonepithelial tumours such as those of lymphoid tissue, soft tissue, bone, and cartilage are not included.)

C01.9 Base of tongue, NOS
C02.4 Lingual tonsil
C05.1 Soft palate, NOS
C05.2 Uvula
C09.0 Tonsillar fossa
C09.1 Tonsillar pillar
C09.8 Overlapping lesion
C09.9 Tonsil, NOS
C10.0 Vallecula
C10.2 Lateral wall of oropharynx
C10.4 Branchial cleft
C10.8 Overlapping lesion
C10.9 Oropharynx, NOS

ANATOMY

Primary Sites and Subsites. The pharynx (including base of tongue, soft palate, and uvula) is divided into three regions: nasopharynx, oropharynx and hypopharynx. Each region is further subdivided into specific sites as summarized in the following:

Nasopharynx. The nasopharynx begins anteriorly at the posterior choana and extends along the plane of the airway to the level of the free border of the soft palate. It includes the vault, the lateral walls including the fossae of Rosenmuller and the mucosa covering the torus tubaris forming the eustachian tube orifice, and the posterior wall. The floor is the superior surface of the soft palate. The posterior margins of the choanal orifices and of the nasal septum are included in the nasal fossa.

Parapharyngeal involvement denotes postero-lateral infiltration of tumour beyond the pharyngobasilar fascia. Involvement of the masticator space denotes extension of tumour beyond the anterior surface of the lateral pterygoid muscle, or lateral extension beyond the postero-lateral wall of the maxillary antrum, pterygo-maxillary fissure.

Oropharynx. The oropharynx is the portion of the continuity of the pharynx extending from the plane of the superior surface of the soft palate to the superior surface of the hyoid bone (or floor of the vallecula) and includes the base of tongue, the inferior surface of the soft palate and the uvula, the anterior and posterior tonsillar pillars, the glossotonsillar sulci, the pharyngeal tonsils; the lateral and posterior walls.

Hypopharynx. The hypopharynx is that portion of the pharynx extending from the plane of the superior border of the hyoid bone (or floor of the vallecula) to the plane corresponding to the lower border of the cricoid cartilage and includes the pyriform fossae (right and left), the lateral and posterior hypopharyngeal walls, and the postcricoid region.
The post cricoid area extends from the level of the arytenoid cartilages and connecting folds to the inferior border of the cricoid cartilage and connects the two pyriform sinuses thus forming the anterior wall of the hypopharynx.

The pyriform sinus extends from the pharyngoepiglottic fold to the upper end of the esophagus at the lower border of the cricoid cartilage and is bounded laterally by the lateral pharyngeal wall and medially by the lateral surface of the aryepiglottic fold, arytenoid and cricoid cartilages. The posterior pharyngeal wall extends from the level of the superior surface of the hyoid bone (or floor of the vallecula) to the inferior border of the cricoid cartilage and from the apex of one pyriform sinus to the other.

**Regional Lymph Nodes.** The risk of regional nodal spread from cancers of the pharynx is high. Primary nasopharyngeal tumours commonly spread to retropharyngeal, upper jugular, and spinal accessory nodes, often bilaterally. Oropharyngeal cancers involve upper and mid-jugular lymph nodes, less likely submental/submandibular nodes. Hypopharyngeal cancers spread to adjacent parapharyngeal, paratracheal and mid- and lower jugular nodes. Bilateral lymphatic drainage is common.

In clinical evaluation the maximum size of the nodal mass should be measured. It is recognized that most masses over 3 cm in diameter are not single nodes but are confluent nodes or tumour in soft tissues of the neck. There are three categories of clinically involved nodes for the nasopharynx, oropharynx, and hypopharynx: N1, N2, and N3. The use of subgroups a, b and c is not required, but is recommended.

For pN, a selective neck dissection will ordinarily include 6 or more lymph nodes and a radical or modified radical neck dissection will ordinarily include 10 or more lymph nodes. Negative pathologic examination of a lesser number of nodes still mandates a pN0 designation.

**Metastatic Sites.** The lungs are the commonest sites of distant metastases; skeletal or hepatic metastases occur less often. Mediastinal lymph node metastases are considered distant metastases.

**RULES FOR CLASSIFICATION**

**Clinical Staging.** Clinical staging is generally employed for squamous cell carcinomas of the pharynx. Assessment is based primarily on inspection, and by indirect and direct endoscopy. Palpation of sites (when feasible) and of neck nodes is essential. Neurologic evaluation of all cranial nerves is required. Imaging studies are essential in clinical staging of pharynx tumours.

Cross-sectional imaging in nasopharyngeal cancer is mandatory to complete the staging process. Magnetic resonance imaging (MRI) often is the study of choice because of its multiplanar capability, superior soft tissue contrast and its sensitivity to skull base and intracranial tumour spread. Computed tomography (CT) staging with axial and coronal thin section technique with contrast is alternative. Radiologic nodal staging should be done to assess adequately the retropharyngeal and cervical nodal status.

Cross-sectional imaging in oropharyngeal carcinoma is recommended when the deep tissue extent of the primary tumour is in question. CT or MRI may be employed. Radiologic nodal staging should also be done simultaneously. Cross-sectional imaging of hypopharyngeal carcinoma is recommended when the extent of the primary tumour is in doubt, particularly its deep extent in relationship to adjacent structures (i.e., larynx, thyroid, cervical vertebrae, and carotid sheath). CT is preferred currently because of less motion artifact than MRI. Radiologic nodal staging should be done simultaneously.

Complete endoscopy, usually under general anesthesia, is performed after completion of other staging studies, to accurately assess the surface extent of the tumour and to assess deep involvement by palpation for muscle resistance and to facilitate biopsy. A careful search for other primary tumours of the upper aerodigestive tract is indicated because of the incidence of multiple independent primary tumours occurring simultaneously.

**Pathologic Staging.** Pathologic staging requires the use of all information obtained in clinical staging in addition to histologic study of the surgically resected specimen. The surgeon’s evaluation of gross unresected residual tumour must also be included. The pathologic description of any lymphadenectomy specimen should describe the size, number and level of any involved nodes.
DEFINITION OF TNM

Primary Tumour (T)

TX  Primary tumour cannot be assessed
T0  No evidence of primary tumour
Tis  Carcinoma in situ

Nasopharynx

T1  Tumour confined to the nasopharynx
T2  Tumour extends to soft tissues
T2a Tumour extends to the oropharynx and/or nasal cavity without parapharyngeal extension *
T2b  Any tumour with parapharyngeal extension *
T3  Tumour involves bony structures and/or paranasal sinuses
T4  Tumour with intracranial extension and/or involvement of cranial nerves, infratemporal fossa, hypopharynx, orbit, or masticator space

*Note: Parapharyngeal extension denotes posterolateral infiltration of tumour beyond the pharyngobasilar fascia.

Oropharynx

T1  Tumour 2cm or less in greatest dimension
T2  Tumour more than 2cm but not more than 4cm in greatest dimension
T3  Tumour more than 4cm in greatest dimension
T4a Tumour invades larynx, deep/extrinsic muscle of tongue, medial pterygoid, hard palate, or mandible.
T4b Tumour invades lateral pterygoid muscle, pterygoid plates, lateral nasopharynx, skull base, or carotid artery.

Hypopharynx

T1  Tumour limited to one subsite of hypopharynx and 2 cm or less in greatest dimension
T2  Tumour invades more than one subsite of hypopharynx or an adjacent site, or measures more than 2 cm but not more than 4 cm in greatest diameter without fixation of hemilarynx
T3  Tumour measures more than 4 cm in greatest dimension or with fixation of hemilarynx
T4a Tumour invades thyroid/cricoid cartilage, hyoid bone, thyroid gland, esophagus or central compartment soft tissue. *
T4b Tumour invades prevertebral fascia, encases carotid artery, or invades mediastinal structures.

*Note: Central compartment soft tissue includes prelaryngeal strap muscles and subcutaneous fat.

Regional Lymph Nodes (N): Nasopharynx

The distribution and the prognostic impact of regional lymph node spread from nasopharynx cancer, particularly of the undifferentiated type, is different than that of other head and neck mucosal cancers and justifies use of a different N classification scheme.

NX  Regional lymph nodes cannot be assessed
N0  No regional lymph node metastasis
N1  Unilateral metastasis in lymph node(s). 6 cm or less in greatest dimension, above the supraclavicular fossa*
N2  Bilateral metastasis in lymph node(s), 6 cm or less in greatest dimension, above the supraclavicular fossa*
N3  Metastasis in a lymph node(s)
N3a greater than 6 cm in dimension
N3b extension to the supraclavicular fossa*

* Midline nodes are considered ipsilateral nodes.
Supraclavicular zone or fossa is relevant to the staging of nasopharyngeal carcinoma and is the triangular region originally described by Ho. It is defined by three points: (1) the superior margin of the sternal end of the clavicle; (2) the superior margin of the lateral end of the clavicle; (3) the point where the neck meets the shoulder (see Fig. 4-2). Note that this would include caudal portions of Levels IV and V. All cases with lymph nodes (whole or part) in the fossa are considered N3b.

**STAGE GROUPING: Nasopharynx**

<table>
<thead>
<tr>
<th>Stage</th>
<th>T</th>
<th>N</th>
<th>M</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>Tis</td>
<td>N0</td>
<td>M0</td>
</tr>
<tr>
<td>I</td>
<td>T1</td>
<td>N0</td>
<td>M0</td>
</tr>
<tr>
<td>IIA</td>
<td>T2</td>
<td>N0</td>
<td>M0</td>
</tr>
<tr>
<td>IIB</td>
<td>T2</td>
<td>N1</td>
<td>M0</td>
</tr>
<tr>
<td></td>
<td>T2a</td>
<td>N1</td>
<td>M0</td>
</tr>
<tr>
<td></td>
<td>T2b</td>
<td>N0</td>
<td>M0</td>
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<td></td>
<td>T2b</td>
<td>N1</td>
<td>M0</td>
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<td>N2</td>
<td>M0</td>
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<td>M0</td>
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<td></td>
<td>T3</td>
<td>N1</td>
<td>M0</td>
</tr>
<tr>
<td></td>
<td>T3</td>
<td>N2</td>
<td>M0</td>
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<td>IVA</td>
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<td>M0</td>
</tr>
<tr>
<td></td>
<td>T4</td>
<td>N1</td>
<td>M0</td>
</tr>
<tr>
<td></td>
<td>T4</td>
<td>N2</td>
<td>M0</td>
</tr>
<tr>
<td>IVB</td>
<td>Any T</td>
<td>N3</td>
<td>M0</td>
</tr>
<tr>
<td>IVC</td>
<td>Any T</td>
<td>Any N</td>
<td>M1</td>
</tr>
</tbody>
</table>

**STAGE GROUPING: Oropharynx, Hypopharynx: Identical to Larynx**

**HISTOPATHOLOGIC TYPE**

The predominant cancer type is squamous cell carcinoma for all pharyngeal sites. Nonepithelial tumours such as those of lymphoid tissue, soft tissue, bone and cartilage are not included in this system. For nasopharyngeal carcinomas it is recommended that the World Health Organization (WHO) Classification be used (Table 4.1). Histologic diagnosis is required to use this classification.

**HISTOPATHOLOGIC GRADE (G): Oropharynx, Hypopharynx**

<table>
<thead>
<tr>
<th>Grade</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>GX</td>
<td>Grade cannot be assessed</td>
</tr>
<tr>
<td>G1</td>
<td>Well differentiated</td>
</tr>
<tr>
<td>G2</td>
<td>Moderately differentiated</td>
</tr>
<tr>
<td>G3</td>
<td>Poorly differentiated</td>
</tr>
</tbody>
</table>

**PROGNOSTIC FACTORS**

In addition to the importance of the TNM factors outlined previously, the overall health of these patients clearly influences outcome. Comorbidity can be classified by more general measures, such as the Karnofsky performance score, or more specific measures, such as the Kaplan-Feinstein Index. Continued exposure to carcinogens, such as alcohol and tobacco smoke, likely also affects patients’ outcome adversely.
<table>
<thead>
<tr>
<th>WHO Classification</th>
<th>Former Terminology</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type 1. Squamous cell carcinoma</td>
<td>Squamous cell carcinoma</td>
</tr>
<tr>
<td>Type 2. Nonkeratinizing carcinoma with lymphoid stroma</td>
<td>Transitional cell carcinoma</td>
</tr>
<tr>
<td></td>
<td>Intermediate cell carcinoma</td>
</tr>
<tr>
<td></td>
<td>Lymphoepithelial cell carcinoma (Regaud)</td>
</tr>
<tr>
<td>Type 3. Undifferentiated carcinoma with lymphoid stroma</td>
<td>Anaplastic carcinoma, clear cell carcinoma</td>
</tr>
<tr>
<td></td>
<td>Lymphoepithelial carcinoma (Schminke)</td>
</tr>
</tbody>
</table>
TNM Classification: AJCC 2002 –3
Thyroid Gland

Although staging for cancers in other head and neck sites is based entirely on the anatomic extent of disease, it is not possible to follow this pattern for the unique group of malignant tumors that arise in the thyroid gland. Both the histologic diagnosis and the age of the patient are of such importance in the behaviour and prognosis of thyroid cancer that these factors are included in this staging system.

ANATOMY

Primary Site.
The thyroid gland ordinarily is composed of a right and a left lobe lying adjacent and lateral to the upper trachea and oesophagus.

An isthmus connects the two lobes and in some cases a pyramidal lobe is present extending upward anterior to the thyroid cartilage.

Regional Lymph Nodes.
Regional lymph node spread from thyroid cancer is common but of less prognostic significance in the generally well-differentiated tumors (papillary, follicular) than in medullary cancers. The first echelon of nodal metastasis is the paralaryngeal, paratracheal, and prelaryngeal (Delphian) nodes adjacent to the thyroid gland in the central compartment of the neck generally described as Level VI. Although involvement of these lymph nodes has minimal prognostic significance, it is usually observed in the older age group. Metastasis secondarily involves the mid and lower jugular, supraclavicular and, much less commonly, the upper deep jugular and spinal accessory lymph nodes. Lymph node metastasis to submandibular and submental lymph nodes is indeed very rare. Upper mediastinal (Level VII) nodal spread occurs frequently both anteriorly and posteriorly. Retropharyngeal nodal metastasis may be seen usually in the presence of extensive lateral cervical metastasis. Bilateral nodal spread is common. The components of the N category are described as first echelon (central compartment/Level VI), N1a and lateral cervical and/or superior mediastinal as N1b. The lymph node metastasis should also be described according to the Level of the neck that is involved. Nodal metastases from medullary thyroid cancer carry a much more ominous prognosis although they follow a similar pattern of spread.

Metastatic Sites.
Distant spread occurs by hematogenous routes, for example to lungs and bones, but many other sites may be involved.

RULES FOR CLASSIFICATION

Clinical Staging.
The assessment of a thyroid tumor depends on inspection and palpation of the thyroid gland and regional lymph nodes. Indirect laryngoscopy to evaluate vocal cord motion is essential. A variety of imaging procedures can provide additional useful information. These include radioisotope thyroid scans, ultrasonography, computed tomography scans (CT), and magnetic resonance imaging (MRI) scans. When cross-sectional imaging is utilized, MRI is recommended so as to avoid contamination of the body with the iodinated contrast medium generally used with CT. Iodinated contrast media will delay the possibility of administering radioactive iodine-131 postoperatively. The diagnosis of thyroid cancer must be confirmed by needle biopsy or open biopsy of the tumor. Further information for clinical staging may be obtained by biopsy of lymph nodes or other areas of suspected local or distant spread. All information available prior to first treatment should be used.
Pathologic Staging.
Pathologic staging requires the use of all information obtained in the clinical staging in addition to histologic study of the surgically resected specimen. The surgeon’s description of gross unresected residual tumor must also be included.

DEFINITION OF TNM

Primary Tumor (T)

Note: All categories may be subdivided: (a) solitary tumor, (b) multifocal tumor (the largest determines the classification).

TX Primary tumor cannot be assessed  
T0 No evidence of primary tumor  
T1 Tumor 2 cm or less in greatest dimension limited to the thyroid  
T2 Tumor more than 2 cm but not more than 4 cm in greatest dimension limited to the thyroid  
T3 Tumor more than 4 cm in greatest dimension limited to the thyroid or any tumor with minimal extrathyroid extension (e.g. extension to sternothyroid muscle or perithyroid soft tissues).  
T4a Tumor of any size extending beyond the thyroid capsule to invade subcutaneous soft tissues, larynx, trachea, oesophagus, or recurrent laryngeal nerve.  
T4b Tumor invades prevertebral fascia, carotid artery or mediastinal vessels

All anaplastic carcinomas are considered T4 tumors.

T4a Intrathyroidal anaplastic carcinoma – surgically resectable.  
T4b Extrathyroidal anaplastic carcinoma – surgically unresectable

Regional Lymph Nodes (N)

Regional lymph nodes are the central compartment, lateral cervical, and upper mediastinal lymph nodes.

NX Regional lymph nodes cannot be assessed.  
N0 No regional lymph node metastasis  
N1 Regional lymph node metastasis  
N1a Metastasis to Level VI (pretracheal, paratracheal, and prelaryngeal/Delphian lymph nodes)  
N1b Metastasis to unilateral, bilateral or contralateral cervical or superior mediastinal lymph nodes.

HISTOPATHOLOGIC TYPE

There are four major histopathologic types:

Papillary carcinoma (including follicular variant of papillary carcinoma)  
Follicular carcinoma (including Hurthle cell carcinoma)  
Medullary carcinoma  
Undifferentiated (anaplastic) carcinoma
STAGE GROUPING

Separate stage groupings are recommended for papillary follicular, medullary or undifferentiated (anaplastic) carcinoma.

**Papillary or Follicular**

Under 45 years
Stage I  Any T, Any N, M0
Stage II Any T, Any N, M1

**Papillary or Follicular**

45 years and older
Stage I   T1, N0, M0
Stage II  T2, N0, M0
Stage III T3, N0, M0
          T1, N1a, M0
          T2, N1a, M0
          T3, N1a, M0
Stage IV-A T4a, N0, M0
          T4a, N1a, M0
          T1, N1b, M0
          T2, N1b, M0
          T3, N1b, M0
          T4a, N1b, M0
Stage IV-B T4b, Any N, M0
Stage IV-C Any T, Any N, M1

**Medullary Carcinoma**

Stage I   T1, N0, M0
Stage II  T2, N0, M0
Stage III T3, N0, M0
          T1, N1a, M0
          T2, N1a, M0
          T3, N1a, M0
Stage IV-A T4a, N0, M0
          T4a, N1a, M0
          T1, N1b, M0
          T2, N1b, M0
          T3, N1b, M0
          T4a, N1b, M0
Stage IV-B T4b, Any N, M0
Stage IV-C Any T, Any N, M1

**Anaplastic Carcinoma**

All anaplastic carcinomas are considered Stage IV

Stage IV-A T4a, Any N, M0
Stage IV-B T4b, Any N, M0
Stage IV-C Any T, Any N, M1
Chapter 1  Role Of Imaging

INTRODUCTION

Within the head and neck there are requirements for imaging which are specific to each subsite and those which apply to evaluation of head and neck tumours in general. The common aims of baseline head and neck imaging are clarified and augmented with site specific protocols, taking into account the development of the Royal College of Radiologists guidelines/protocols. Squamous cell carcinoma is the commonest head and neck cancer and because it arises from mucosal surfaces is usually diagnosed by direct visualisation and biopsy. Clinical assessment of mucosal disease of the primary tumour is significantly superior to any radiological technique - the clinical estimate of the mucosal extent of the primary tumour exceeds the radiographic estimate. Conversely, cross sectional imaging is essential for evaluating the deep extent of the disease.

Imaging in patients with head and neck malignant disease is crucial for various reasons. It is important to ascertain the location and relationship to neighbouring structures of the primary disease. This is particularly important in surgical planning. The initial imaging will also indicate tumour size and the status of the rest of the neck. In order to stage certain head and neck tumours fully, information may also be required as to the metastatic status in other areas, notably the chest and the skeletal system/liver.

Imaging is important in monitoring the response to treatments, particularly in respect of recurrent disease, further spread or the development of a second primary. In some instances imaging can be useful in enabling optimal function to be obtained following treatment or as a baseline before treatment, e.g. the use of swallowing investigations and those associated with dental/maxillofacial function. The use of imaging therefore forms an important part of pre and post treatment work up and each unit should have access to CT/MRI scanning, ultrasonography, radio isotope imaging and more simple plain x-ray techniques such as a simple chest radiograph and an orthopantomogram (although the use of this form of radiograph is controversial and most now prefer CT scanning). Contrast examinations such as a barium/water soluble swallow may also be required. Ultrasonographic techniques can also be combined with the use of fine needle aspiration and this is considered further. Imaging is useful for detecting and monitoring post treatment complications, and rehabilitation.

Various reports in the world literature have shown a positive correlation between tumour volume and prognosis for all sites (1, 2, 3). Imaging has rendered T staging a lot more precise. Various methods of determining tumour volume from e.g. CT scans have been devised such as the summation of areas technique in various planes (4) and errors of volume calculations using this technique are within 5-10 % when compared with volumes determined by excision and examination. Most of the studies dealing with tumour volume and clinical progression arise from series of patients with laryngeal and oropharyngeal disorder and in the one by Gilbert et al (3) it was shown that the tumour volume determined from pre-treatment CT scans was the most important predictor for a successful outcome with radiotherapy.

Volume measurement, however, in other sites is probably less important. Olmi et al (5) and Cellai et al (6) showed that tumour volume in nasopharyngeal carcinoma has only a minor influence on prognosis. Pameijer et al (7) in a study investigating variability of tumour volumes in T3 staged head and neck tumours showed considerable variability of tumour volumes in conventionally staged T3 head and neck tumours and proposed incorporation of tumour volume data to the TNM classification system, in view of the potential as a prognostic indicator.
CT SCANNING

Although it is appreciated that all imaging departments are under a great deal of pressure imaging should be carried out ideally within two weeks of referral and preferably before biopsy. If imaging is performed after biopsy then this should ideally be carried out 7-10 days later as the biopsy may result in air pockets and possibly result in the disease being upstaged.

Computerised axial tomographic scanning or CT scanning uses conventional x-rays with computer processing of digital information obtained from layer or spiral tomographs. It is particularly useful in evaluating the relationship of tumours with bone and can also be useful for evaluating malignant disease in relation to cartilage infiltration.

Assessment of the tumour in relation to spread into cartilage is useful in, for example, laryngeal tumours where significant upstaging can result from invasion into or through the thyroid cartilage. Radiotherapy tends to lose its effectiveness if tumour invades cartilage.

CT PROTOCOL

3-5 mm collimation sections using intravenous contrast (100mls of 300mg/ml) should be obtained to include the skull base to sternal notch. Intravenous contrast helps increase the conspicuity of the primary tumour and is essential for evaluating cervical nodal metastases. The patient should lie supine with the neck comfortably extended and the head immobilised in a suitable support. Patients are told not to swallow during the examination and to breathe quietly. A lateral scout view should be obtained from which the examination is planned. Spiral or dynamic modes should be used if possible as this reduces motion artefact and optimises use of iv contrast. Direct coronal sections or reconstruction (if using spiral CT) may be necessary to assess tumour extension to the skull base and cranial fossa. Soft tissue and bone reconstruction algorithm should be used.

For the nasopharynx, sections parallel to the hard palate from the level of the cavernous sinus to sternal notch are obtained.

Proper gantry angle is essential in the oropharynx and oral cavity to avoid dental related artefacts and if necessary the gantry angle should be changed to optimise visualisation of the oral tongue and tongue base.

For the larynx and hypopharynx, sections must be obtained parallel to the true vocal cords and this is approximately parallel to a disc space or the plane of the hyoid bone.

The advantages of CT scanning over and above MRI is that the acquisition times are faster and thereby there is less motion artefact. Bone detail is better. It is cheaper than MRI scanning and non-claustrphobic. It is also noted to be easier to use.

Co-morbidity is very important and probably to date has not been fully addressed in head and neck practice. At least in some UK regions, the detection of unspecified pulmonary metastatic disease, even for small T stages is expensive and the rate not sufficiently high to warrant routine use. There are also specific sites [thyroid, sinus] and tumour types [high grade salivary] where chest CT is advisable in any case. For patients already in the scanning room, the marginal cost of additional chest CT is very little more than the previously "routine" chest x-ray, and can also be used by the anaesthetist. But if there is no spiral scanner, on the other hand, scanning the chest in addition to the primary site and neck drainage areas would significantly extend scanner time.

A screening chest CT following examination of the head and neck to detect pulmonary metastatic disease or synchronous primary in bronchogenic carcinoma has been advocated by some authors (8, 9, 10). Other authors have not found this to be justified (11,12).
This remains a controversial area, since one has to bear in mind the expense and the low pick up rate and the risk of added ionising radiation. CT guided needle biopsy of pulmonary lesions is also potentially useful.

**MAGNETIC RESONANCE IMAGING**

Magnetic resonance imaging uses the magnetic properties of biological materials and generates images without the use of ionising radiation.

**MRI PROTOCOL**

T1W and T2W or STIR sequences in at least two planes, preferably axial and coronal, and T1W FSE or Fat Suppressed images post gadolinium. The use of contrast is particularly useful in identifying perineural spread and leptomeningeal involvement. Although images of the nasopharynx can be obtained with a head coil, a combined head and neck coil is necessary to allow assessment of cervical lymph nodes. The ideal section thickness is 3-4mm with at most a 10% gap. Fat saturation images permit better identification of the tumour margins as the method helps separate the enhancing tumour margins from background fat.

MRI scanning has distinct advantages over CT. There is no ionising radiation, there is multiplanar capability and no dental amalgam artefact. Soft tissue contrast is superior and there is no requirement for iodinated contrast media. In addition MR angiography may also provide information about the relationship of the major vascular structures to the mass lesion.

**ULTRASONOGRAPHY**

Ultrasonography has proved useful in evaluation of lymphadenopathy and particularly useful in respect of accurate insertion of needles for biopsy. Differentiation between benign and malignant disease, however, has not otherwise been easy and CT/ MRI scanning is generally considered to be superior, being able to also stage the primary tumour. Ultrasonographic criteria have been evaluated and assessed to assist in this differentiation in the study by Vassallo et al 1992 (17). It is widely accepted that ultrasonography is more sensitive than simple palpation for the detection of enlarged lymph nodes (14, 15 & 16). High resolution ultrasound probes have been shown to differentiate between the hilus and cortex of a lymph node where changes in the usual relationship suggest the presence of malignant disease. It has also been useful in assessing infiltration of blood vessels by nodal metastasis (17). In the study by Vassallo et al 1992 (17) 204 patients with suspected lymphadenopathy underwent ultrasonography and a result obtained using various parameters concerning the shape and relationship of hilar/cortex.

This was compared with subsequent histological evaluation. Nodal size was found not to be a reliable criterion for differentiating benign from malignant nodes but of relevance in monitoring response to treatment.

Differences were found in the longitudinal-transverse diameter ratio and the presence of eccentric cortical widening was seen only in malignant nodes. The accuracy of ultrasonography can be increased by fine needle aspiration cytology performed at the same time. It is usually not possible to stage the primary tumour with ultrasonography, it is highly dependent upon operator experience and in general there are no significant advantages over CT/MRI scanning other than cost.

The aims therefore of imaging in patients with head and neck malignant disease can therefore be summarised in table 1. It is assumed that each unit has rapid access to CT scanning, orthopantography, ultrasonography and fine needle aspiration cytology.
RATIONALE FOR THE BASELINE HEAD AND NECK PROTOCOL

The key issue in tumour staging is the question of tumour volume, but there is as yet no standard volumetric method. The cross sectional area or maximum diameter of the primary tumour should be routinely reported.

The neck should be scanned 1] to assess the contralateral nodes, 2] because at the time of imaging the primary treatment is not known. If the neck is ultimately treated non surgically, no staging neck dissection information will be available.

(i) IMAGING OF NECK NODES

The diagnosis of pathological neck nodes in patients with squamous cell cancer of the head and neck is made either on size criteria or on the recognition of central nodal necrosis. This causes rim enhancement with central low density following IV contrast enhanced CT. If this is seen in a patient with squamous cell cancer of the head and neck, it is highly specific for the presence of a pathological neck node whatever the size of that node. It has been said to have an accuracy of 100% (18). The presence of central necrosis is recognised on MRI either by heterogeneity of signal intensity on T2 weighted scans or by rim enhancement following the use of IV Gadolinium. There is debate amongst authors whether IV Gadolinium is necessary or whether T2 weighted images alone will be sufficient (18,19). Different authors have used different size criteria. A minimal axial size of 10mm (11 mm in a subdigastric region) was felt to be the most effective (19). Groups of 3 or more borderline nodes also increases the sensitivity of detecting pathological neck nodes and does not significantly decrease the specificity (18,19). Other authors have used a minimal axial diameter of 8 mm as the cut off (20). Comparing CT with MRI some authors have found CT to perform better than MRI (21, 18).

A meta-analysis of CT versus physical examination using a 15 year Medline review with 647 neck dissections showed CT to have a sensitivity of 84%, specificity of 83% and accuracy of 83% with physical examination having a sensitivity of 74%, specificity of 81% and accuracy of 77%. It was found that the diagnostic modalities were additive with CT significantly enhancing detection rates of physical examination alone. Overall physical examination detected 75% but this increased to 91% with the use of CT (22).

Ultrasound guided fine needle aspiration cytology may also be used to detect pathological neck nodes. Some have found it to be more accurate than CT (23) but other authors have not found an advantage (24). Ultrasound alone does not have any advantage over CT and is unable to stage the primary tumour.

The role of positron emission tomography in staging the primary tumour and nodal disease in the initial staging is uncertain. It is expensive and lacks the spatial resolution of other cross-sectional imaging modalities. It is best reserved for problem patients or in the setting of recurrent disease.

(ii) SITE SPECIFIC ASPECTS

The radiological criteria used for diagnosing a lymph node as malignant on CT include:

1. nodal size, central necrosis
2. extracapsular extensions
3. obliteration of perivascular fat planes
The criteria for malignant node on MRI include:

1. Evidence of central necrosis.

2. Minimal axial diameter greater than 11 mm in the subdigastric area or greater than 10 mm in other areas.

3. Grouping of three or more borderline nodes in the lymph nodes draining region of the primary tumour.

With the above criteria, these results are an improvement on those reported in the literature for CT or MRI and of great importance in enabling the detection of occult metastatic disease in the neck. Intravenous contrast is important in the head and neck to allow recognition of pathological rim enhancement in nodes of normal size. Anatomical staging terms understood by clinician and radiologist should be used. The node number, size (CSA/max diameter) and levels should be reported on an agreed standard proforma, preferably with a diagram number.

**PARANASAL SINUSES AND NASOPHARYNX**

Tumours arising in the paranasal sinuses and nasopharynx often involve critical structures, their deep extent is not visible at nasoendoscopy and thus radiological imaging is essential in staging. The commonest tumours can be grouped as to their type and site and the imaging issues related to these are discussed.

**CT technique**

The complex nature of paranasal sinus anatomy means that imaging in more than one plane is often vital to an appreciation of disease extent, thus where helical CT is available it is recommended that the scan be performed with axial helical acquisition and reformats obtained for coronal images. This avoids patient repositioning and repeat scanning (plus dental artefact is minimised). Thin collimation (1mm) is required covering from the top of the frontal sinuses to the maxillary teeth and the image data should be reconstructed at 1mm increments (or less) in order to allow reformation of good quality true coronal images (perpendicular to the hard palate). Sagittal reformations may also be helpful to the surgeon for lesions involving the frontal recess region. Axial scanning has the further benefit that repositioning of the patient is not required in order to scan the neck to stage lymphatic spread and this should be performed as for any other head and neck neoplasm.

**MRI technique**

MRI of the paranasal sinuses must be performed with thin sections (3 - 4 mm), no gap, small field of view and high image matrix (512 reconstruction) in the head coil in order to maximise spatial resolution and SNR. Fast spin-echo imaging is helpful for T2 in order to reduce acquisition time; this is less of an advantage for T1 imaging. T1 images in both coronal and axial planes should be obtained. The choice of which plane is used for T2 is according to individual preference. Gadolinium contrast enhancement is strongly recommended to demonstrate skull base involvement and intracranial extension (25, 26) but must be performed with fat suppression and the same coverage/parameters as non-fat suppressed pre-contrast T1 imaging to enable direct comparison.

Additional sagittal imaging is helpful for posterior nasal and PNS lesions. If there is frank invasion of the skull base then a supplementary examination of the whole brain is prudent. As elsewhere an overview coronal STIR sequence of the neck will help in staging lymphadenopathy.
BENIGN PARANASAL SINUS TUMOURS

Benign tumours such as osteoma, ossifying fibroma and dentigerous cysts are usually adequately imaged by standard unenhanced CT sinus protocols though as imaging in both the axial and coronal planes is helpful then helical scanning is recommended where available.

Inverting papilloma requires more extensive imaging as there may be malignant degeneration and even with benign localised disease it is helpful to distinguish between the tumour mass and any retained secretions. Thus with CT, contrast enhancement should be employed and the neck scanned to exclude lymphadenopathy. MRI may be more productive in differentiating tumour from retained secretions and better help to determine local soft tissue extent (27).

Juvenile angiofibroma may be localised but often invades through the sphenopalatine foramen to the pterygopalatine fossa as well as to the infratemporal fossa. The extent at presentation must be known, as full excision is the key to minimising recurrence (28). MRI is recommended and the addition of sagittal imaging is suggested to show the anteroposterior extent of the lesion. It may also show sphenopalatine extension to advantage. Contrast enhanced MR angiography can usefully show the extent of major vessel vascular supply - this is best performed in the sagittal plane using the head coil with thin partitions and small field of view to maximise spatial resolution.

MALIGNANT PARANASAL SINUS TUMOURS -

Malignant paranasal sinus tumours are uncommon and include SCC (the commonest and usually maxillary sinus in origin), lymphoma and minor salivary gland tumours (including adenoid cystic and mucoepidermoid carcinomas). Tumours arising from bone and/or cartilage such as myeloma/plasmacytoma, osteosarcoma & chondrosarcoma also occur but are rare. Aside from the finding of calcification in the matrix of the sarcomas the features of these tumours are similar as they manifest as local soft tissue masses with local bone destruction. Malignant paranasal sinus tumours are best initially staged by MRI due to its superior sensitivity to the extent of tumour spread, particularly intracranial and perineural spread. Axial imaging best evaluates pterygopalatine and infratemporal fossa extension while coronal images are superior for evaluating orbital and intracranial extensions. Lymphatic spread of these tumours is initially to the retropharyngeal nodes and thence the upper deep cervical chain - these areas should thus be carefully evaluated.

TABLE 1 - IMAGING CHECK LIST FOR PARANASAL SINUS TUMOURS

- Identification of primary tumour, differentiation from retained secretions
- Assessment of extent of bone destruction
- Involvement of hard palate and middle meatus for maxillary tumours (i.e. the infrastructure as defined by Ohngren’s line)
- Extension to pterygoid plates, retroantral fat and pterygopalatine fossa
- Any orbital involvement
- Identification of perineural involvement, skull base invasion and extension to anterior or middle cranial fossa
- Nodal involvement looking first to retropharyngeal nodes.
NASOPHARYNGEAL SCC

The ideal protocol for the primary tumour and neck is MRI scanning and if there is any uncertainty in respect of disease involvement of the base of skull then CT may provide better bone detail. MRI is more accurate than CT in evaluating primary nasopharyngeal malignancy, primarily due to the relative immobility of the nasopharynx. Motion artefact is thereby minimised allowing MR to capitalise on its inherent superior soft tissue contrast relative to CT.

Deep infiltration of the tumour appears to be a more important prognostic factor in nasopharyngeal carcinoma than tumour volume, hence the importance of MRI. In recent studies MRI has proved better than CT in identifying obliteration of the pharyngobasilar fascia, invasion of the sinus of Morgagni, (through which the cartilaginous portion of the eustachian tube and the levator veli palatini muscle pass), metastases to lymph nodes in the carotid and retropharyngeal spaces (29, 30) and invasion of the skull base (29, 30, 31).

If nodal spread is confirmed the chest should be imaged, preferably by CT scan as there is a significant incidence of thoracic nodal and pulmonary parenchymal spread (32). Important radiological features are as shown in table 3.

TABLE 2 - IMAGING CHECK LIST FOR NASOPHARYNGEAL TUMOUR (33,34)

- Identification of primary tumour looking closely at any asymmetry in Fossa of Rosenmuller.
- Any involvement of the deep spaces (parapharyngeal, carotid, prevertebral and masticator) indicating violation of the pharyngobasilar fascia and other deep fascial layers.
- Identification of skull base invasion
- Any perineural tumour spread is critical as this upstages the tumour to T4 and requires an increase in radiotherapy dose.
- Any involvement of nasal cavity, maxillary, ethmoid or sphenoid sinuses.
- Any orbital involvement- direct involvement is rare but orbit may be invaded via the pterygopalatine fossa, inferior orbital fissure and ethmoid/sphenoid sinuses.
- Nodal involvement looking closely at retropharyngeal nodes.

IMAGING FOR SKULL BASED NEOPLASMS (32,36)

Aim
1. Diagnosis.
2. Define extent and size.
3. Perineural spread.
4. Pre-Operative staging
5. Surgical and DXT planning
6. Post-operative assessment

Modalities
1. CT Scan
2. MRI/MRA
3. Angiogram
4. PET scan

40
CT Scan
Axial/coronal scans.
Spiral CT with volume acquisition & multiplanar reformats.
Slice thickness 3mm or less.
Contrast. 50-100 cc IV.
Soft and Bone algorithms.

CT Scan better than MR at assessing bone involvement.

MRI
T1 axial & Coronals.
5mm slice or less.
STIR axial/coronals.
Pre and Gadolinium Post contrast T1 fat suppressed scans. 3-5mm slice thickness.
T2 and Proton density scans to assess sinus/mastoid secretions.

MR excellent at detecting
1. Intracranial involvement.
2. Dural/leptomeningeal involvement.
3. Perineural spread. This is associated with decreased survival.
4. Marrow infiltration.

Arteriography
2. For diagnosis and evaluation of vascularity.
3. For endovascular therapy.
4. Trial carotid occlusion.

PET Scans
FDG PET commonly used agent.
All Hospitals may not have access.
Post op. Assessment.
Differentiate scar/granulation tissue from recurrent/residual tumour.
Limitations of PET to be borne in mind.

ORAL CAVITY AND OROPHARYNX

Cross sectional imaging has significantly altered the treatment and management of these oral and oropharyngeal malignancies, but often underestimates the extent of gingival and hard palate tumours.

A comparison of CT and MRI in T and N staging of primary and recurrent SCC of the oral cavity and oropharynx was performed in 33 patients (51 episodes) by Leslie et al 1999 (37).

This showed that for T staging, MRI is overall more accurate than CT. If degraded images on CT due to dental amalgam and T1 tumours are however excluded, the techniques are comparable. MRI is oversensitive for recurrent disease in the post operative neck as non specific high signal from previous surgery and radiotherapy can mimic recurrent disease. For N staging all methods failed to detect small metastatic deposits. The presence of an irregular outline of nodes on MRI in this study suggested the presence of extracapsular spread but other authors (38) have found it of limited value. Wide et al 1999 (39) have also shown that MRI lacks sufficient sensitivity and specificity to replace elective neck dissection for both staging and prognostic purposes.
The oral cavity has an extensive lymphatic drainage system with metastasis most frequently to Levels I, II and III. The presence of cervical metastases in patients with SCC of the oral cavity is a vital prognostic indicator.

Positron Emission Tomography (PET) has been evaluated by Myers and Wax 1998 (40) in the N0 neck in patients with SCC of the oral cavity and correlated with pathologic examination of neck dissections. PET in 11 patients and 19 neck dissections was found to have an overall sensitivity, specificity, positive predictive value and accuracy of 100%. Although promising, the numbers in this study are small and PET scanning is not freely available. Thallous Chloride Tl201-Labelled Single Photon Emission Computed Tomography Scanning in Head and Neck Cancer was shown by Gregor et al(41) to be useful in detecting occult primary lesions in the oropharynx.

The most commonly performed surgical procedure for treatment of squamous cell carcinoma of the tongue is a partial glossectomy, a procedure that requires preservation of one lingual artery and hypoglossal nerve. Information from imaging that will directly affect the surgical approach includes whether the tumour involves the ipsilateral neurovascular bundle, submucosal involvement in adjacent areas including the floor of the mouth and whether the tumour crosses the midline. If the tumour crosses the midline, its relationship to the contralateral lingual neurovascular bundle precludes a partial glossectomy and requires a total glossectomy, which is often unacceptable. Treatment of tonsillar and soft palate carcinomas depends on lesion size and involvement of surrounding structures. Extension to involve the parapharyngeal and masticator spaces, requires resection of portions of the tongue base, mandible and maxilla. Important radiological features are as shown in tables 3 and 4.

**TABLE 3 - IMAGING CHECK LIST FOR OROPHARYNGEAL TUMOUR**

1. **Tongue base carcinoma**
   - Extension to floor of mouth and surrounding structures
   - Relationship to ipsilateral lingual neurovascular bundle
   - Extension across midline and relationship to contralateral neurovascular bundle

2. **Tonsil, soft palate and posterior pharyngeal wall carcinoma**
   - Submucosal extension into parapharyngeal space and nasopharynx
   - Tongue base invasion
   - Encasement of carotid artery
   - Bone erosion
   - Pre vertebral muscle invasion

**TABLE 4 - IMAGING CHECK LIST FOR ORAL CAVITY TUMOURS (other than nodal involvement)**

1. **Lip Carcinoma**
   - Bone erosion
   - Soft tissue invasion

2. **Floor of Mouth Carcinoma**
   - Extent of bone erosion
   - Deep invasion along the mylohyoid and hyoglossus muscles
   - Relationship to ipsilateral lingual neurovascular bundle
   - Extension across midline and relationship to contralateral neurovascular bundle
   - Tongue base invasion
   - Extension into the soft tissues of neck
(3) Oral Tongue carcinoma  
- Invasion of ipsilateral lingual neurovascular bundle  
- Extension across midline and relationship to contralateral neurovascular bundle  
- Invasion of floor of mouth and associated bone erosion

(4) Buccal carcinoma  
- Submucosal extension  
- Bone erosion

(5) Gingival and hard palate carcinoma  
- Bone erosion  
- Perineural invasion of the incisive canal and greater and lesser palatine foramen

(6) Retromolar trigone carcinoma  
- Bone erosion  
- Submucosal spread  
- Perineural invasion

LARYNX

The larynx is divided into the supraglottis, glottis and subglottis. The supraglottic larynx is located above the true vocal cords and extends from the tongue base and valleculae to a laryngeal ventricle.

Imaging of Laryngeal Tumours

The Royal College of Radiology is developing guidelines for all head and neck sites and these have been integrated into this section. They are evidence based and the level of evidence and recommendation are given below.

| Summarised results from each of the cited references | MRI is better than CT for T staging prior to partial laryngectomy (42,43,44) due to direct coronal imaging capability. MRI is more sensitive but is less specific than CT for cartilage involvement (45, 44) but both acceptable and may be complementary (46). MRI is preferred for T staging if the patient is cooperative (42) or tongue base is involved (43). CT is good at assessing anterior and posterior commissures and fat spaces when compared to histopathology, but is less good for assessing cartilage involvement, extralaryngeal spread and N stage (47). CT combined with clinical evaluation is better than either alone, particularly for supra- and trans-glottic tumours (48). CT is as good as MRI for N staging (44) or better (43). |
| Conclusion | MRI has advantages over CT for T staging in certain situations, and is otherwise equal, so MRI is preferable where available, though CT is acceptable. CT is as good as or better than MRI for N staging. US can be used for T and N staging and follow-up of laryngeal cancer by centres with appropriate expertise. |
| Evidence level classified I – IV | IIb – III for CT/MR and III for ultrasound |
| Investigation | CT or MRI. Ultrasound. |
| Recommendation * | CT or MRI are indicated but ultrasound is a specialised investigation. |
| Grade of Recommendation A – C | B |
| Comment | Where available, MRI preferable to CT for T staging. Either for N staging. Ultrasound can be used for T and N staging and follow-up in centres with appropriate expertise. |
CT of the neck should be performed with the gantry angled to the true cords. This may be approximated by using the position of the hyoid bone on a lateral scout view to determine the correct angling of the gantry. CT sections should be taken at least 5mm, preferably 3mm, intervals through the larynx. The area scanned should include nodal stations likely to be involved by the primary tumour, i.e. level 2 superiorly and scanned inferiorly to examine the subglottic region, i.e. to the root of the neck. Scans should be obtained following a bolus of intravenous iodinated contrast agent (at least 100 ml).

Intravenous contrast agent helps to increase the conspicuous of the primary tumour and is necessary for evaluating neck nodes. CT scans should be evaluated at soft tissue settings. Scans through the region of the laryngeal cartilage should also be evaluated using a bony algorithm and bony window settings.

MR imaging should be performed using a neck coil or other suitable surface coil. MR imaging should be performed in both the axial and coronal planes with a slice thickness of 4 mm or less. T1 and T2 weighted spin echo images should be obtained. STIR (Short Time Inversion Recovery) images may be useful for the evaluation of laryngeal cartilage involvement. The STIR sequence takes out the fat signal which appears as black with water as white. These scans however have poor spatial discrimination and are not used in isolation. If intravenous contrast agent is given, T1 weighted fat saturated scans may be useful following this. The role of intravenous Gadolinium enhanced MRI in the staging of the primary tumour and neck nodes in laryngeal carcinomas is less clear than that of intravenous iodinated contrast enhanced CT (18), (19). Important radiological features are as shown in table 5.

TABLE 5 - IMAGING CHECK LIST FOR LARYNGEAL TUMOURS

Key Imaging Findings
1. Transglottic extension of tumour.
2. Piriform sinus invasion.
3. Cartilage invasion.
4. Spread outside the larynx.

Glottic and Supraglottic Tumours
Transglottic extension.
Subglottic extension and relationship to cricoid cartilage.
Cartilage invasion.
Involvement of the anterior commissure.
Involvement of the crico-arytenoid joint.
Extension to the posterior commissure.
Deep invasion of the paraglottic fat.
Tumour volume.

Subglottic Tumours
Primary subglottic tumours are uncommon.
Inferior extent.
Cartilage invasion.
State of tracheal lumen, particularly in respect of ability to intubate.
Tumour volume.
HYPOPHARYNX

The hypopharynx extends from the hyoid bone to the inferior aspect of the cricoid cartilage and is anatomically divided into the pyriform sinuses, post cricoid region and posterior pharyngeal wall.

Imaging includes CT, MRI, barium swallow and PET scanning. There is little if any role for ultrasound to specifically assess the primary tumour but it may be of some use in detecting local tumour recurrence (49). Various authors (50, 51, 52) stress the importance of CT and MRI to assess deep tumour extension into the pre-epiglottic space, para-glottic space and laryngeal framework. The primary tumour is inaccurately staged by clinical examination in 35-47% of cases while CT/MRI upstages up to 40% of cases (53). PET scanning is at present of limited use in staging because of poor anatomical detail. CT has an accuracy of 65%-73% for hypopharyngeal cancer while a combination of clinical examination and CT results in accuracy rates for staging ranging from 73-88%. Comparison of conventional and single slice spiral CT by Kosling (54) showed no significant difference in the TMN staging of oro- and hypopharyngeal tumours. The addition of the valsalva manoeuvre in CT is a supportive method in evaluating pyriform fossa tumours (55). 3D and CT virtual endoscopy are promising techniques that are still being evaluated.

Zbaren (50) specifically comparing CT and MRI in the pretherapeutic staging of hypopharyngeal carcinoma concluded that there was no difference in the staging accuracy between CT and MRI. Held (56) comparing CT with ultrafast MR sequences in all pharyngeal tumours showed that functional imaging with phonation and valsalva manoeuvre was particularly useful in the case of hypopharyngeal tumours. MRI in this study was shown to have a higher false positive rate and CT a higher false negative rate. Held (57) showed improved tumour detection on MRI with provocative manoeuvres (accuracy 95.5%) when compared with MRI during quiet respiration (accuracy 80%). The use of intravenous gadolinium has also been shown to be more useful in assessing the extent of tumour and laryngeal cartilage invasion (58, 59).

Mukherji (60) and Pameijer (61) prefer CT for hypopharyngeal staging but in around 10% of cases feel that MRI is also needed to resolve issues such as prevertebral muscle invasion and submucosal spread towards the oesophageal verge. Various studies (65, 66, 67, 68) have shown that PET may be more accurate than CT or MRI for detecting local recurrence post treatment for a variety of head and neck cancers with sensitivity of 50-100% and specificity of 71-73%.

Follow up neck CT is particularly useful in detecting local failures in those patients initially identified as high risk (69). Thoracic CT is useful for symptomatic and asymptomatic post treatment patients with Stage 3 or 4 disease but only useful in Stage 1 or 2 patients who present with clinical recurrences (10).

Imaging is otherwise as for larynx but with the possible addition of barium swallow to assess the lower extent of the lesion and for detection of a synchronous oesophageal primary (62-64). This is important clinical information since some hypopharyngeal tumours are treated surgically with preservation of the cervical oesophagus while others require its removal.

**TABLE 6 - IMAGING CHECK LIST FOR HYPOPHARYNGEAL TUMOURS**

**Key Imaging Features**

1. Extension across the midline.
2. Apical involvement.
3. Extension into the oesophageal inlet.
4. Extension into chest.
5. Cartilage invasion.
6. Anterior extension into the para-glottic space.
PAROTID/SUBMANDIBULAR REGION

Ultrasound, CT and MRI are all useful in evaluating the salivary glands, particularly for neoplasia. MRI is probably better than CT. It should be pointed out that some clinicians do not image salivary gland tumours at all if they are superficial and mobile and particularly in the tail of the parotid gland.

i. CT Technique

5mm iv contrast enhanced images (75-100 mls of 300mg/ml contrast) from the level of the pinna to hyoid bone. The plane of section may need to be altered to avoid artefacts from dental amalgam. Patient is asked not to swallow and to breathe quietly. With this technique there is less claustrophobia than MRI scanning.

ii. MR Technique

Head and neck coil may be necessary for full coverage. Axial and coronal planes with 4-5mm slice thickness, T1W and T2W or STIR sequences. Intravenous contrast is rarely necessary unless adenoid cystic tumour is suspected because of the potential for perineural spread of disease.

| Summarised results from each of the cited references | Both CT and MRI can be used for staging parotid tumours (71, 72). Corr et al (73) reserve CT for staging deep lesions, using US to stage small, superficial ones. Soler et al (74) consider MRI important for staging and follow-up. Freling et al (75) found MRI good for staging malignant tumours, and Kamal et al (76) demonstrated MRI to be better than CT for staging parotid tumours. Corr et al (73) found US better than CT for the detection and characterisation of parotid masses and Shimizu et al (77) demonstrated that the shape and distribution of internal echoes can predict the nature of a parotid mass. Schick et al (10) found pulsed and colour Doppler sonography a useful adjunct to grey scale images for differentiating benign from malignant lesions. McGuirt et al (71), Banerjee et al (72) and Kamal et al (76) all found that FNAC improved the diagnostic accuracy of other methods of assessment - clinical or imaging. |
| Conclusion | MRI is preferred to CT for staging of parotid tumours, although either is acceptable, particularly for deep lesions. US +/- Doppler can suggest the nature of a parotid mass in addition to its detection. FNAC improves diagnostic accuracy further. |
| Evidence level classified I - IV | III for US and IIb – III for CT/MRI |
| Investigation | CT, MRI and US |
| Recommendation * | Indicated |
| Grade of Recommendation A – C | B |
| Comment | CT and MRI are particularly for deep lesions and prior to complex surgery. MRI preferred if available. Ultrasound is good for the detection of superficial lesions, and can often characterise them. FNAC improves diagnostic accuracy. |

Important radiological features are shown in table 7.
TABLE 7 - IMAGING CHECK LIST FOR PAROTID GLAND TUMOURS

Location of mass in superficial or deep lobe of parotid gland
Identify any occult mass in same gland or opposite gland
Identify relationship to facial nerve using styloid process, posterior belly of digastric and retromandibular vein as reference
Has mass smooth or infiltrative margins?
Is the mass solid or cystic?
Is the mass confined to gland?

EXTERNAL MEATUS AND MIDDLE EAR

Imaging should show primary site and metastatic fields. CT scan should include the neck with contrast. Raw data should be acquired to reconstruct relevant images on sharp bone algorithm.

MR scanning should include gadolinium enhancement to exclude intracranial involvement.

THYROID

Ultrasound, isotope scans, CT and MRI are all used for imaging the thyroid and guidelines are outlined here for diagnosis, staging and follow-up, taking into account the Royal College of Radiologists guidelines which are being drawn up.

Diagnosis

| Summarised results from each of the cited references | US alone is not reliable in differentiating benign from malignant nodules (79), but used to guide or in combination with FNAC, US-FNAC is the best available technique (80, 81, 82, 83, 84, 85, 86, 87), although it is unreliable for follicular and Hurthle-cell nodules (88). It is useful for the detection of residual/recurrent disease at the primary site and in locoregional lymph nodes after thyroidectomy (89, 79), and can again be combined with FNAC (87). |
| Statement (= the conclusion drawn from E) | US-FNAC for diagnosis, but unreliable for follicular and Hurthle-cell nodules. |
| Evidence level classified I – IV | IIb – III |
| Investigation | US |
| Recommendation * | Indicated |
| Grade of Recommendation A – C | B |
| Comment | Used in combination with or to guide FNAC. |
### Staging

| Summarised results from each of the cited references | US better than MRI for staging small primary tumours, multifocal disease and lymphadenopathy. (80,83,90). Hay and Klee (91) specify tumour extent and lymph nodes and James et al (87) concentrate on lymphadenopathy and recurrent disease. Shimamoto et al (92) obtained 82% accuracy for T staging, but only 48% accuracy for N stage due to a low sensitivity, and consider US limited in assessing extracapsular extent of tumour and lymph node involvement. |
| Conclusion | US useful for staging small primary tumours and lymph nodes. |
| Evidence level classified | I – IV IIb – III |
| Investigation | US |
| Grade of Recommendation A – C | B |
| Comment | Where appropriate expertise is available, the investigation of choice for locoregional staging. |

| Summarised results from each of the cited references | MRI (90), CT (80, 89), or either (82, 83, 84, 86, 79) for the demonstration extrathyroidal/extracapsular and mediastinal extension. Peritracheal disease (87) and tracheal compression (82) can be demonstrated by either modality. MRI to demonstrate lymphadenopathy if US is not available (82, 93), particularly for medullary thyroid carcinoma with 93% accuracy (81). CT or MRI are required for the staging of medullary thyroid carcinoma to look for other tumours in MEN syndromes (83, 89), and for assessment of recurrent disease (79). Distant metastases can be shown by CT for lung (80), MRI (89) or either (79). |
| Conclusion | The extent of large primary tumours, detection of distant metastases and staging of medullary thyroid carcinoma in MEN syndromes can be performed using CT or MRI |
| Evidence level classified | I – IV IIb – III |
| Investigation | CT or MRI |
| Recommendation * | Indicated |
| Grade of Recommendation A – C | B |
| Comment | To assess large primary tumours, detect distant metastases, and for medullary thyroid carcinoma in MEN syndromes. |
Follow up

| Summarised results from each of the cited references | NM mainly used for demonstration of residual/recurrent differentiated cancer after thyroidectomy (80, 88, 84, 89, 91, 94, 79), including medullary thyroid carcinoma. [18F]-2-deoxy-2-FDG PET has 82% sensitivity for demonstrating residual/recurrent differentiated carcinoma after thyroidectomy (79). |
| Conclusion | Follow-up of differentiated carcinomas after thyroidectomy. |
| Evidence level classified I – IV | Lib |
| Investigation | NM |
| Recommendation * | Indicated |
| Grade of Recommendation A – C | B |
| Comment | For the detection of residual/recurrent disease after thyroidectomy. See Cancer Section L05. |

POST TREATMENT IMAGING OF THE NECK

A potentially difficult area is imaging the post operative neck where surgery can lead to significant alteration in the normal anatomy. It is important therefore that the reporting radiologist has some familiarity with the various forms of neck dissection that are now performed from the original radical neck dissection described by Crile to the modified varieties (96,95). The situation may be complicated since reconstructive techniques can also produce anatomical confusion. This is principally by use of various skin flaps and/or visceral transposition. A useful review of these and typical and radiological representations are as described by Som et al (97).

This team recommends imaging 4-8 weeks post operatively, during which time a baseline can be obtained and periodic imaging thereafter over at least the first three years at 4-6 monthly intervals. Yearly intervals thereafter for at least two more years are also advised. These recommendations are probably not in accordance with most British practice which tends to centre around clinical suspicion of recurrence with appropriate investigation and imaging thereafter. Som et al (97) point out, however, that the imaging findings on this regime altered the post operative salvage plan in 25% of cases where recurrence occurred and that clinical occult disease was found at sectional imaging in 17%.

Both CT and MRI can be difficult to interpret and the imaging modality chosen depends on pre operative/radiotherapy work up. Other imaging such as nuclear medicine procedures have been tested. Although for staging purposes, PET is limited by its lack of anatomical detail, it compares favourably with CT and MRI in detecting recurrent/residual cancer (65, 98). Thallium 201 SPECT is particularly useful in assessing recurrences (41, 99). PET-FDG has also been shown to be more accurate than CT/MRI for identifying recurrent tumours. This area is reviewed more fully by Hanasono et al (68).

References:
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Pathology Services – Staffing: A Consultant Histopathologist and a Consultant Cytopathologist with special interest in head and neck diseases are essential members of the multidisciplinary clinical oncology team. Appropriate cover for periods of leave should be included in the service budget. The Cytopathologist may attend the early diagnosis clinics to provide a 'one-stop' service. Local circumstances may dictate that this is not an efficient use of the Consultant resource and cytological specimens may be sent to the laboratory for reporting. The turnaround time for reports will depend upon the nature of the diagnostic problem and the work required to reach a diagnosis. Consultant Pathologists should participate in an appropriate, accredited, diagnostic EQA scheme.

Laboratory Facilities: Space for the preparation and reporting of cytological specimens is required if a 'one-stop' clinic service is provided. The laboratory service should participate in technical EQA and should be accredited by Clinical Pathology Accreditation (UK) Ltd or to a similar standard.

A minimum dataset for histopathology reports on cases of head and neck squamous carcinomas has been published by The Royal College of Pathologists, and will be updated during 2002-3 to include a section on salivary neoplasms. The current document can be downloaded from College website [www.rcpath.org]. The aims of this document are to:

- enhance and make more consistent the information provided for patient management through the use of a proforma report or checklist
- facilitate the monitoring of changing pattern of disease
- allow equitable comparison of surgical units and of patients in clinical trials

The Royal College of Pathologists has commissioned minimum datasets for the histopathology reports on thyroid neoplasms and is considering salivary neoplasms; the broad outlines of their likely composition are included for guidance.

MINIMUM DATASET FOR SQUAMOUS CARCINOMAS

This minimum dataset is for squamous carcinomas, is evidence-based and the contents are summarised here.

The content of the histopathology report will include clinical information (that should be provided by the surgeon or oncologist), data on the primary carcinoma and data on the extent of nodal involvement (if dissected).

1. Clinical data
   1.1. Primary carcinoma
   - Site(s) and side(s) of the carcinoma(s) recorded according to the UICC nomenclature.
   - Type of resection specimen e.g., total or partial glossectomy, laryngectomy.
   - Clinical TNM stage. The final pathological T coding at some sites e.g., the larynx, will be determined by clinical features such as vocal cord mobility (2)
   - Previous radiotherapy or chemotherapy (this may influence the histological interpretation).
   - Illustrative diagrams of the primary resection specimen are invaluable to the pathologist in some situations, and their use is to be encouraged. Some centres provide pre-printed request forms that include such diagrams.
   - Where the orientation of the specimen is critical e.g. for assessment of marginal status, the surgeon and pathologist should agree a protocol for marking the specimens with clips or sutures.
For some sites e.g. mucosal resection and partial laryngectomy specimens, specimens may be pinned onto cork or polystyrene boards to preserve the anatomical relationships and minimise distortion during fixation (3).

1.2. Neck dissections
- The type of neck dissection should be specified as either comprehensive or selective.
- Node levels present in the specimen.
- Nodes in addition to the main groups e.g. parapharyngeal nodes, should be sent as separate specimens.
- Specimens should be pinned onto cork or polystyrene boards to preserve the anatomical relationships and minimise distortion during fixation. The surgeon and pathologist should agree a protocol for marking the specimens with clips or sutures to indicate node levels.

2. Pathological data for the primary carcinoma
- Maximum diameter of tumour (millimetres).
- Maximum depth of invasion (millimetres) below the luminal aspect of surface.
- Histological type of carcinoma. The guidelines specifically apply to typical squamous carcinomas. Subtypes of squamous carcinoma, such as papillary, verrucous, basaloid, adenosquamous and spindle cell carcinomas, should be recognised [4]
- Degree of differentiation. Grading of the most aggressive area is based on the WHO classification (4) into well, moderately or poorly-differentiated carcinomas. Anaplastic or undifferentiated carcinomas are also recognised. This system is prognostically useful, even though it suffers from inter-observer variability and sampling problems (5,6).
- Invasive front of the carcinoma. The pattern of invasion by the carcinoma at its deep margin is of proven prognostic value for oral carcinomas (7,8,9). The few published studies of tumours at other sites suggest that a similar approach may be of value (10). Scoring systems for histopathological features of squamous carcinomas have the potential to improve the consistency of reporting, but are not in widespread use and do not form part of the minimum dataset.
- Distance from invasive carcinoma to mucosal and deep margins (millimetres).
- Vascular invasion.
- Perineural invasion ahead of the invasive front of the carcinoma is especially important for carcinomas of the lip.
- Bone invasion.
- Severe dysplasia/in situ carcinoma is associated with a high risk of progression to carcinoma and its presence both adjacent to the primary carcinoma and at the resection margins should be recorded (11,12).

Other features that may be reported
It should be emphasised that this is a minimum dataset and individual centres may wish to record features in addition to those listed above. Some features should be included as part of a comprehensive description of a carcinoma and the surrounding tissues, but are considered to be of uncertain prognostic significance at most sites in the head and neck region.

1. Type and intensity of inflammatory infiltrate and desmoplastic stromal response.
2. Involvement of a tracheostomy (if present).
3. Response to previous therapy (if applicable).
4. Results of other investigations e.g. flow cytometry, molecular and immunohistochemical studies.

The diagnostic role of immunohistochemical techniques
A wide range of antibodies is available to help resolve diagnostic problems. Most antibodies lack a precise tissue of neoplastic specificity, so that a combination of appropriate results is required to make a diagnosis.
Molecular markers of prognosis or prediction of therapeutic response.
Markers including measures of cell proliferation and nuclear DNA content, the expression of involucrin, blood group antigens, cell adhesion molecules and oncogenes, and the intensity of neoangiogenesis have been investigated as potential prognostic factors. These features generally correlate with cellular differentiation but do not provide any consistent independent prognostic information (7,13,14).

The tumour suppressor genes p53 and p16 and the oncogene cylinD1 have been studied in squamous cell carcinoma of the head and neck. p53 mutation per se may not be of prognostic significance (15), but null mutations do appear to be of prognostic value (16). Likewise combinations of genetic alterations are being identified which carry prognostic significance (17). At present it is unclear as to whether or not molecular staging or classification of cancers are clinically relevant and multicentre trials are underway (18,19). In the UK, this type of staging may not be possible or practical with current funding of pathology laboratories. Thus, while molecular markers predictive of tumour behaviour or response to therapy may be required pathological data in the future, current surgical practice does not demand their inclusion in the minimum data set.

3. Pathological data from the neck dissection specimen
The extent of nodal involvement is a major prognostic factor for head and neck squamous carcinomas. The following data items should be recorded:
- At each anatomical level, record the total number of nodes identified and number of nodes involved by carcinoma.
- Size of largest metastatic deposit. The size of the largest metastasis is a determinant in the TNM staging (2).
- Presence or absence of extra-capsular rupture and the node level(s) showing this feature. Extracapsular spread is a manifestation of the aggressiveness of a carcinoma and is associated with a poor prognosis (20,21). Although the prognostic significance of micrometastases (<3mm. diameter) is not determined (22), their presence should be included in the number of involved nodes.

Other features that should be included as part of a comprehensive description, but are considered to be of uncertain prognostic significance at most sites in the head and neck region.
1. Presence of other pathology in cervical nodes.
2. Presence of evidence of response of tumour e.g. keratin debris, to previous therapy

Sentinel node biopsy
The clinical utility of sentinel node biopsy is controversial and the most appropriate method of handling such specimens has not been defined. The current guidelines from the ADASP (23) indicate that:

Intaoperative frozen section diagnosis is only appropriate where the result will change clinical management. Only routine, H&E stains should be used except for experimental situations. The whole node should be processed for paraffin sections and ‘more than one section’ taken from each block. There are no evidence-based guidelines to indicate how many sections should be used. It is unclear whether or not immunocytochemical staining adds clinically relevant information [false-positive results can occur].

4. Diagnostic coding of primary carcinomas and metastases
- pT and pN status should be recorded according to the UICC guidelines (2).
- SNOMED T and M codes should be recorded for both primary site(s) and for nodes.
MINIMUM DATASET FOR THYROID NEOPLASMS

A minimum dataset for pathology reports is being prepared for the Royal College of Pathologists (details may be obtained from Dr Ann Marie McNicol, University Department of Pathology, Glasgow Royal Infirmary, GLASGOW G4 0SF). The pathological features likely to be included in this document include:

- specimen type (thyroidectomy or lobectomy), size and weight
- histological type of neoplasm
- macroscopic size and degree of encapsulation of the neoplasm
- presence or absence of invasion of the thyroid capsule or surrounding tissues and distance from resection margin
- presence or absence of vascular invasion
- microscopic distance from excision margin
- extent of lymph node involvement
- diagnostic coding of primary carcinomas and metastases (UICC and SNOMED)

MINIMUM DATASET FOR SALIVARY TUMOURS

A minimum dataset for pathology reports will form part of the updated Head and Neck Carcinoma pathology.

The following features should be recorded for salivary neoplasms:

- size of tumour
- distance from tumour to resection margins [macroscopic distance confirmed histologically]
- histological type of neoplasm [and subtype for adenoid cystic carcinomas]
- grade of malignance
- presence of perineural or vascular invasion
- extent of lymph node involvement
- diagnostic coding of primary carcinomas and metastases (UICC and SNOMED).

Other features should be included as part of a comprehensive description, but are considered to be of uncertain prognostic significance:

- whether tumour is solitary or multifocal
- nature of tumour margin [encapsulated or ill-defined]
- solid or cystic tumour
- mitotic index
- microscopic changes in the macroscopically normal salivary tissue

References:

3 SERVICE PROVISION

Chapter 1 Prevention

Key Points

- Tobacco exposure is the important risk factor for the development of upper aerodigestive tract squamous cell carcinoma.
- Excessive alcohol has a synergistic effect with tobacco use.
- There is no evidence that screening leads to a reduction in mortality or morbidity.
- Preventative intervention must concentrate on modifying smoking habits.
- Optimal treatment for tobacco dependence consists of structural behavioural support involving multiple sessions combined with nicotine replacement or bupropion.
- Chemo prevention remains a promising area for future research, but the current evidence shows no benefit for the prevention of second primary cancers.

Introduction

The most important risk factor for the development of upper aerodigestive tract squamous cell carcinoma is tobacco exposure. Alcohol is also an independent risk factor although the exact inter-relationship is hard to quantify because intake of the two is so highly correlated. Tobacco and alcohol together, however, synergistically influence the development of head and neck cancer, with heavy smokers and heavy drinkers having 38 times the risk of oral cancer than abstainers of both (1).

Other risk factors are of lesser significance when controlling for alcohol and tobacco consumption and so preventative interventions for head and neck cancer must concentrate on modifying smoking habits. Other preventative interventions include screening and chemo prevention. The usefulness of general population screening is limited by the low prevalence and incidence of the disease, the potential for false positive diagnoses and the poor compliance with screening and referral (2). There is no evidence that screening of the general population or high risk groups leads to a reduction in mortality or morbidity. Chemo prevention is an attractive proposition. However, there is inadequate evidence to support the use of any chemo preventative drug or other agent.

Modifying tobacco and alcohol intake

Tobacco is clearly the most preventable cause of head and neck cancer. This was recognised for laryngeal cancer (3) and continues to be a constant finding with smokers of more than one packet of cigarettes per day having a risk of head and neck cancer that is 13 times higher than that of non smokers (4). Tobacco smoking and alcohol drinking each contribute to the risk of second cancers with the effects of smoking being more pronounced (5).

Tobacco use also adversely influences cancer patients’ survival – after controlling for age, race, alcohol use and histological grade, smokers had a lower rate of survival than non smokers with risk ratios of 1.55 for males and 1.43 for females (6).
Trigg et al (7) have also shown that smokers present with higher grades of cancers – patients diagnosed with stage III or IV laryngeal cancers smoked a significantly greater amount and were also more likely to be heavy drinkers.

Given these facts, the moderation of alcohol consumption and the cessation of smoking should be a high priority in head and neck cancer prevention.

Few medical interventions of any kind have the same potential as smoking cessation to both improve health and prolong life and arguably be cost effective. Smoking halves the chance of survival to 70 years and accounted for 120,000 deaths in the UK in 1995. In a group of 1445 male smokers aged 40 to 59, an intervention group received individual advice on the relationship between smoking and health and over a 20 year period total mortality was seven per cent lower in this group, fatal coronary artery disease 13 per cent lower and lung cancer 11 per cent lower (8). Similar studies in head and neck cancer have shown a substantial drop in oral and pharyngeal cancers.

Table 1

<table>
<thead>
<tr>
<th>Effect of smoking cessation on risk of oral and pharyngeal cancer in 1114 cases, 1268 controls (1)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Odds Ration</td>
</tr>
<tr>
<td>Current smoker</td>
</tr>
<tr>
<td>1-9y ex smoker</td>
</tr>
<tr>
<td>10-19y ex smoker</td>
</tr>
<tr>
<td>20y + ex smoker</td>
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</tbody>
</table>

An Italian study (9) confirms the reduction in Table 1 with an overall odds ration after allowance for age, sex, education and alcohol consumption of 8.4 for current smokers, 6.2 for those who had stopped smoking for less than two years, 4.5 for those who had stopped smoking for three to five years and 1.6 for those who had stopped for ten to 14 years. The risk of premalignant lesions such as leukoplakia is also found to be reduced following smoking cessation (10).

The effect of reducing alcohol intake is less clear and it is difficult to assess the effects of reducing alcohol intake independently of reducing smoking. Franceschi (11) does however show a reduced odds ratio for the development of oral and pharyngeal cancer from 11.6 for current drinkers to 1.9 for former drinkers.

Facilitating smoking cessation

Interest in smoking cessation is high in patients with head and neck cancer, and although the diagnosis of a tobacco related malignancy clearly represents a strong catalyst for stopping smoking, a sizeable subgroup of patients continue to smoke (12). More than 132,000 people used smoking cessation services in England between April 2000 and March 2001 with 49 per cent giving up for at least a month after setting a ‘quit’ date.

- Behavioural therapy

Data from 188 randomised controlled trials suggest that encouragement and advice given by a doctor during a single consultation resulted in two per cent of all smokers stopping without relapse for up to a year (13) Recent studies looking at intervention by ward nurses (14) or midwives (15) have no effect at six weeks or one year. Intervention may trigger an attempt to quit, but the subsequent treatment is important. A Cochrane review shows that behavioural support and counselling increases a smokers chances of achieving lasting abstinence (16).
• **Nicotine replacement**

Nicotine replacement attempts to replace nicotine in the body with gradual weaning to reduce the pharmacological sequelae of the chemical addiction. Nicotine is absorbed through the cheek from gum or lozenges, transdermally via patches or transmucosally via inhalers or sprays, though these latter may also become addictive. Patches are available on prescription and increase the odds ratio of abstinence to 1.71 (17).

• **Bupropion**

Available on prescription as Zyban this drug reduces the withdrawal symptoms and has been shown to improve abstinence rates over six months in people with cardiovascular disease (18). Optimal treatment for tobacco dependence consists of structural behavioural support involving multiple sessions combined with nicotine replacement or bupropion (19).

**Chemo prevention**

Chemo prevention may be defined as the use of chemical agents to prevent the development of cancer. The epithelium of the upper aerodigestive tract has a rapid turnover and is exposed to ingested and inhaled carcinogens. This, along with a genetic susceptibility predisposes to the development of premalignant lesions, cancers and second primary cancers. The idea that a chemo preventative drug could arrest the progression of premalignancy to invasion, or prevent the development of a metachronous primary is an attractive one. Laboratory and epidemiological studies suggest that dietary carotenoids are inhibitors of epithelial carcinogenesis with inverse associations between beta carotene intake and risk of laryngeal cancers (20). Low vitamin A and C intake also appears to increase the risk of oral cancer (21) and case controlled studies show that oral cancer patients have lower consumption of fruit and vegetables (22). In vitro studies have shown that retinoids block tumour promotion by inhibiting proliferation, inducing apoptosis and inducing differentiation (23). Work with nude mice suggests that retinoids and interferon alpha are synergistic anti-angiogenic agents (24).

Both vitamin A derivatives and N-acetyl cystine showed some early promise though toxicity was a significant concern with dose related dry skin, cheilitis, conjunctivitis and arthralgia being the main side effects. Disappointingly, however, the EUROSCAN multicentre trials of 2592 patients showed no statistically significant benefit for these interventions (25). Equally, topical vitamin A showed no benefit in the treatment of leukoplakia (26).

Chemo prevention remains a promising area for future research, but the current evidence shows no benefit for the prevention of second primary cancers. Vitamin A analogues may reduce the incidence of malignant change in premalignant lesions, but there remain considerable problems with toxicity therefore limiting their use at present.

**References:**


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Section 3  Chapter 2  Role of Multidisciplinary Team

All patients with a diagnosis of cancer of the head and neck must be seen in a multidisciplinary clinic. The recommended staffing structure is:

<table>
<thead>
<tr>
<th>Present in clinic</th>
<th>Available throughout clinic</th>
<th>Affiliated to clinic</th>
</tr>
</thead>
<tbody>
<tr>
<td>Head and Neck Surgeon (1 of 2)</td>
<td>Speech pathologist (dysphagia)</td>
<td>Dental hygienist</td>
</tr>
<tr>
<td>Clinical oncologist (1 of 2)</td>
<td>Restorative Dental Practitioner</td>
<td>Pathologist – head and neck focus</td>
</tr>
<tr>
<td>Reconstructive Surgeon (1 of 2)</td>
<td>Palliative Care Physician</td>
<td>Prosthodontist</td>
</tr>
<tr>
<td>Head and Neck Nurse Specialist</td>
<td></td>
<td>Cytologist</td>
</tr>
<tr>
<td>Dietitian</td>
<td></td>
<td>Radiologist – head and neck focus</td>
</tr>
<tr>
<td>Speech pathologist (voice)</td>
<td></td>
<td>GI surgeon/physician</td>
</tr>
<tr>
<td>MDT Co-ordinator</td>
<td></td>
<td>Physiotherapist</td>
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<tr>
<td></td>
<td></td>
<td>Social Worker</td>
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<tr>
<td></td>
<td></td>
<td>Psychologist</td>
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</table>

The particular skills of an Otolaryngologist which makes him/her a key member of the surgical team are in:
- diagnostic endoscopy/video documentation
- primary therapy
- management of acute and chronic upper airway compromise
- management of associated disorders/treatment complications e.g. otologic

The key to delivery of multidisciplinary care is the careful co-ordination of the combined oncology clinic as well as its consistent delivery throughout the year. The Clinic co-ordinator is responsible for preparatory work for the clinic including assembly of results, as well as keeping an attendance register, ensuring liaison between members, supporting data capture and facilitating further appointments and treatments.

Head and neck cancer patients represent a significant challenge in achieving and maintaining optimal nutritional status, with around two thirds of diagnosed patients experiencing moderate to severe malnutrition. (1,2,3,4). Early intervention is essential to prevent deterioration of nutritional status in this high risk group. (5) The Dietician is a key attendee at the combined oncology clinic to provide nutritional assessment, advice and support from diagnosis, and regular monitoring through the patients cancer journey and in liaising with relevant professionals across the secondary care and community. See section Beyond Primary Treatment

Most patients benefit from the service of a Speech and Language Therapist (SALT)(Pathologist). The SALT, as a permanent member of the multidisciplinary team provides assessment and ongoing advice on swallowing, speech and overall communication, which may include advice and/or provision of appropriate communication aids. Their patient contact will normally have begun preoperatively, and supports the patients’ journey involving radiotherapy, and major head and neck surgery including surgical voice restoration. The multidisciplinary clinic provides an appropriate environment for regular patient review and monitoring of progress/change.

The role of nurse specialists is multifaceted and is a core function of the multidisciplinary team (6,7), in co-ordinating professionals across secondary and primary care to give seamless care. The nurse specialist shares information with the patient and relative at the multidisciplinary clinic. Patients fail to retain half of medical information given(8): therefore nurse specialists can help amplify the consultation outcome, and provide contact telephone numbers to each patient and relative for future use.
Their skills encompass expertise in pain and symptom management and the ability to discuss treatment options. Specialist nurses utilise counselling skills and act as the patient’s advocate from diagnosis and throughout the cancer journey, independent of treatment outcome.

All patients should have a general dental/prosthodontic assessment because of the impact of both radiotherapy and surgery on dentition. Continued support through treatment and in palliative care is important. The dental hygienist educates patients in self care, and prevents possible post treatment problems by improving oral hygiene, as well as encouraging visits to their own dentist or Community dental scheme if appropriate.

Some patients, especially after radical neck dissection, or with coexistent chest disease, will benefit from physiotherapy. Close liaison with specialist head and neck pathology and radiology services, and an affiliated gastrointestinal team with an interest in gastric mobilisation and jejunal harvest, and where necessary, percutaneous gastrostomy, is desirable.

The advantages of working from a multiprofessional combined oncology clinic for both patient and professional are as follows:-

**Advantages of Centralised Multidisciplinary Clinics**
- Avoids attendances at multiple appointments with different professionals
- Avoids reduplication of information from various members of the MDT
- Ensure professionals work “together”, rather than “alongside” each other
- Increases knowledge, development and understanding among professionals
- Potential sharing of certain advisory roles and thus provide some cover for staff absence
- Peer support
- Continuity of care
- Cost effective for both time and resources

These outweigh the potential disadvantages:-

**Disadvantages of Centralised Interdisciplinary Clinics**
- Sub optimal use of specialist time
- Intimidating atmosphere from team size
- Pressure on room space
- Brief consultation times, especially if patient well
- Increased complexity to run clinic to schedule
- Longer distances to travel
- Appointment dates potentially inflexible

Many of these drawbacks can be addressed by increased resources, in particular having multiple rooms available and a surplus of personnel. To assist in the smooth running of the clinic, some groups find it helpful to divide new and follow up patients into quite separate clinic times, minimising extended waits for the patient and reducing the number of clinicians needed throughout the clinic.

Continuous engagement of patient views on the configuration of the multidisciplinary clinic should be sought.
References:
The “NHS Cancer Plan” (1) was presented to Parliament by the Secretary of State for Health in September 2000. The plan applies to England only but similar plans have been published in Scotland, Wales and Northern Ireland (2). The “NHS Cancer Plan” builds on previous government publications including “The New NHS: modern, dependable” (Dec.1997) (3), “The NHS Plan” (July 2000) (4) and its main features are summarised in the short guide for patients/users “Your guide to the NHS” (Feb 2001) (5). It sets ambitious targets for waiting times for investigation and treatment of patients with cancer or symptoms suggestive of cancer. It also states that these targets can only be achieved with an expansion of capacity and investment in new facilities, equipment and staff; and reform of cancer delivery services.

“The New NHS” target, to be achieved by December 2000, was that all patients with suspected cancer should be seen by a specialist, within 2 weeks of their GP deciding that they need to be seen urgently and requesting an appointment (in Wales, within 10 working days of receipt of the referral by the hospital). By 2005 the maximum wait from diagnosis to treatment should be one month. The maximum wait from GP referral to treatment should be two months with an ultimate goal of one month (which is currently the standard for all paediatric and testicular cancers).

Ambitious targets have also been set for routine referrals. By 2005 the maximum wait for routine outpatients should be 3 months (average 5 weeks) and the maximum wait for routine inpatient procedures should be 6 months (average 7 weeks). “Your Guide to the NHS” states that “waiting lists for all hospital appointments and admissions will be replaced by booking systems that allow you to choose a convenient date and time with a guaranteed maximum waiting time.”

The concept of QED (quick and early diagnosis) was presented in the Lancet in 1996 (6) outlining the procedures in the QED unit in Birmingham. The unit provides centralised facilities and administration for a variety of specialities and/or symptoms. The actual work of the unit is no different to that of conventional clinics but the process is efficient and streamlined. The most important features are:

- Neck lumps should be referred to head and neck clinicians who can adequately examine the upper aerodigestive tract before proceeding to further investigations
- Referral by telephone with immediate provision of date and time of appointment to patient and GP
- One-stop approach for examination, endoscopy, radiology and pathology
- ‘Real time’ computer documentation of the whole process
- Regular demographic and clinical audit.

Can ‘QED/rapid access/one-stop’, be successful in the head and neck cancer setting?

Three UK departments have published initial results of such services (7,8,9). Fine needle aspiration cytology and fibreoptic endoscopy have been known to be diagnostically reliable and reproducible in the outpatient setting for over 17 years in the UK (10,11) and 26 years in the USA. The major difficulties for most departments will be:

- providing experienced and skilled staff for regular, rapid access clinics, every week of the year
- the number of referrals

This second point is highlighted in the recent Commission for Health Improvement, National Service Framework Assessment Number 1, (12), which states “…more resources are probably spent on checking patients who do not have cancer than treating those who do.”
The second edition of this document (13) estimated 50,000 new dysphonia referrals to UK otolaryngology clinics with a cancer detection rate of 5%. Our own district audit, if extrapolated, would suggest 111,000 referrals with a cancer detection rate of only 2% for laryngeal cancer. Resouly et al (Dec 2001) (9) suggest much stricter referral criteria to their dysphonia clinic than those published in the Department of Health “Referral Guidelines for Suspected Cancer” (14). Even so, their cancer rate was 1 in 34 referrals (3%). The Glasgow experience (15) shows that, even with referral by fax, it is not possible to achieve the 2 week waiting time target with current staffing levels.

**QED: the future**

There are many possible reasons for setting up QED services.

1) Early diagnosis leads to improved survival and/or quality of life.
2) Rapid access to diagnostic and treatment services reduces anxiety and uncertainty in patients with symptoms suspicious of cancer and is the preferred choice of patients.
3) The Patients Charter requires that patients with a possible diagnosis of cancer be seen within 2 weeks of referral.

The first point, while clinically the most important, lacks evidence. Early stage disease has a better prognosis than late stage and it is tempting to extrapolate from this that survival from head and neck cancer will be improved with the introduction of rapid access services. Unfortunately, no such evidence has been reported in head and neck cancer. In breast cancer, where high quality screening and symptomatic services are available (and funded), mortality has fallen with the associated service improvements. The Eurocare Working Group report (16), suggests that the UK has lower survival rates for cancers: “unexpectedly lower than rates of nearest nations, often below the European average”. Reducing the outpatient waiting time for cancer symptoms from the current average of 4 to 6 weeks to the target of 2 weeks on the face of it may only produce a small improvement in survival. However, small improvements at every stage of the cancer patients journey may have an impact. A lack of direct evidence should not prevent common sense being used as an argument for funding an improved service that most multidisciplinary staff as well as patients and politicians would like available.

The second point is obvious in that all patients with worrying symptoms would like to be seen promptly. One stop and other rapid access services are popular with patients and score well on satisfaction surveys.

The third point is the most easily dealt with. The NHS plan is absolutely clear that cancer patients or patients with suspicious symptoms must be seen quickly, whether or not evidence exists that this will improve survival. Funding is currently available to allow the development of innovative cancer services and cancer is at the moment the first priority for booked appointments.

**Developing a QED service**

Major differences in facilities and staffing in different hospitals make it difficult to advise on the exact structure of a QED service for head and neck cancer symptoms.

The cancer network multidisciplinary head and neck team must approve and be part of the model. The MDT should agree how patients are managed and treated by the most appropriate specialities.

A recent report in the British Medical Journal suggests the use of the collaborative improvement model to enhance services and reduce delays in cancer treatment in the UK (17). National Service Framework documents and NICE “improving outcomes in cancer” documents, specifically targeting head and neck cancer, should be published in 2003 and are available on the internet (18,19).

A possible ‘gold standard’ model is described, providing an integrated service.

- Plan to deliver at least one clinic each week (typical population covered 500,000)
- At least 2 consultant head and neck surgeons and specialist registrars to allow for leave
- Cytopathology service including technicians and consultant pathologists (within clinic)
• Consultant radiologists providing ultrasound scanning of the neck, with guided FNA where necessary (within clinic)
• Central office receiving telephoned or faxed requests for appointments
• Full documentation of both process and clinical details of the consultation, ultimately ‘real-time’ on computer
• Other members of the MDT as deemed appropriate: nurse specialist, speech therapist, dental hygienist, to support the clinic.

References:
2. NHS Wales Quality Care and Clinical Excellence: Putting patients first. Welsh Office, Jan 1998 and HMSO (CM 3841)
4. The NHS Plan (CM 4818-I) HMSO July 2000
12. NHS Cancer Care in England and Wales, National Service Framework Assessment Number 1 (Commission for Health Improvement December 2001) (also at www.chi.nhs.uk/cancer/index.htm)
18. NICE “Improving Outcomes in Cancer” series (available at www.doh.gov.uk/cancer)

Search Strategy for Chapter

Section 3  Chapter 4  Quality Assurance

In April, 2001, the NHS sponsored a ‘proposal generating event’ in Harrogate consisting of multidisciplinary groups from all over the UK working to determine what an ideal head and neck service would look like. Since then, a team at NICE has been using this as a basis for new COG guidelines which should include much that would be covered by this section in future years. However, at the time of writing, the emergence of the COG document has been delayed until at least September 2003. Therefore, we feel that the standards below are likely to remain relevant until the 4th Edition of this book in 2004.

Quality
This is a ‘degree of excellence’. The use of this term has different implications in different contexts. In industry, it is associated with a philosophical viewpoint and the use of a wide range of tools and techniques, not all of which are of use in the service sector. In the service sector, including health care, quality assurance has moved from being an ill-defined process of assessing client satisfaction, to a comprehensive and, where possible, quantitative process.

Standards
Standards are the cornerstone of quality assurance. They are precise and authoritative statements of the criteria necessary to ensure that a process is fit for the purpose for which it is intended. Standards are drawn up with the co-operation of and consensus, or general approval of, interested parties. They are based on the consolidated results of science and experience, aimed at the promotion of optimum community benefits and approved by a body recognised on a national (in this case, the BAO-HNS), regional or international level. Standardisation is an activity to improve efficiency by bringing consistency to services. In the present context, this could be seen as an attempt to reduce so called ‘postcode’ variation. Standards are essential for audit, bench-marking and accreditation/certification/designation of cancer centres.

In preparing this section on quality assurance, we do not seek to limit the judgement of individual clinicians or teams in how to manage individual patients. Rather, we hope the standards provided will form a common framework for the delivery of the head and neck care process and a mechanism for continuous improvement, including providing evidence for better service provision by health authorities and hospital Trusts.

Accreditation
The accreditation process is now widespread in head and neck cancer care, with each region (and nationally, in Scotland) producing its own set of standards. Third-party accreditation has been used as part of this process in some regions. It is hoped that the present standards (themselves subject to regular review), will assist in the continuing audit and re-accreditation cycles in these regions, as well as assisting those regions wishing to embark on such a process.

Derivation of standards
The quality section of the first edition of Effective Head and Neck Cancer Management (1998) was used as the template. This was modified using recent research, as well as by comparison with other national and regional standards. The most important modifier was the result of an extensive patient and carer exercise. Where professional and patient/carer standards disagreed significantly, the patient/carer viewpoint has been taken as the default position.
References:

1. Effective Head and Neck Cancer Management. 1st Ed. London; BAO-HNS. Royal College of Surgeons of England, 1998. This is the first edition of the present document and used widespread national consensus within Otolaryngology.


3. Head and neck cancer: consensus standards for the process of care. Bristol; South and West Regional Cancer Organisation, 1997. This regional document used an extensive formal multidisciplinary consensus process to define detailed standards for all parts of the process of head and neck cancer care. Updating is overdue, however.


MINIMUM TEMPORAL STANDARDS IN THE PROCESS OF HEAD AND NECK CANCER CARE

<table>
<thead>
<tr>
<th>Interval</th>
<th>Standard</th>
</tr>
</thead>
<tbody>
<tr>
<td>GP/GDP to first outpatients</td>
<td>14 days</td>
</tr>
<tr>
<td>clinic note to GP/GDP</td>
<td>7 days</td>
</tr>
<tr>
<td>Fine-needle or other biopsy arrival in pathology</td>
<td>no wait</td>
</tr>
<tr>
<td>time for frozen section result</td>
<td>30 minutes for one, 45 for multiple</td>
</tr>
<tr>
<td>biopsy to report issue</td>
<td>7 days</td>
</tr>
<tr>
<td>Patient to be seen in MHNC from being told of diagnosis of cancer</td>
<td>7 days</td>
</tr>
<tr>
<td>general clinic to MHNC</td>
<td>7 days</td>
</tr>
<tr>
<td>MHNC to EUA/panendoscopy/dental/prosthetic</td>
<td>7 days</td>
</tr>
<tr>
<td>MHNC to radiotherapy, chemotherapy (curative intent)</td>
<td>14 days to planning for radiotherapy</td>
</tr>
<tr>
<td>All patients notified of start and finishing dates of their treatment</td>
<td>14 days to start chemotherapy</td>
</tr>
<tr>
<td>Radiotherapy start from planning</td>
<td>Within 48 hours</td>
</tr>
<tr>
<td>MHNC to radiotherapy, chemotherapy (palliative intent)</td>
<td>7 days to planning</td>
</tr>
<tr>
<td>MHNC to ablative surgery</td>
<td>14 days</td>
</tr>
<tr>
<td>Within radiotherapy/chemotherapy course</td>
<td>as planned and documented</td>
</tr>
<tr>
<td>primary treatment to rehabilitation (speech, swallowing, needs assessment)</td>
<td>no delay</td>
</tr>
<tr>
<td>treatment to first follow-up clinic</td>
<td>one month</td>
</tr>
</tbody>
</table>

GP = General Practitioner
GDP = General Dental Practitioner
MHNC = Multidisciplinary head and neck clinic

STANDARDS FOR AUDIT AND ACCREDITATION

<table>
<thead>
<tr>
<th>Activity</th>
<th>Task</th>
<th>Standard</th>
</tr>
</thead>
</table>
| All activities | co-ordination | 100% of centres should have a named head and neck specialist responsible for co-ordinating the local provision of care.  
100% of centres should have a 100% specialist head and neck liaison nurse (e.g. Macmillan), whose contact details should be provided to all patients at the earliest opportunity in all cases.  
100% of centres should have a list of consultants accredited to provide head and neck cancer care at that centre |

70
<table>
<thead>
<tr>
<th>Communication</th>
<th>Issue referral letters</th>
<th>should arrive at hospital within 48 hours of dictation [e.g. fax]</th>
</tr>
</thead>
<tbody>
<tr>
<td>issuing of discharge summaries</td>
<td>Centres need a written protocol and or/checklist to include: • BRIEF overview of the inpatient care received • name and contact details of head and neck specialist liaison nurse for GP and patient/carers • diagnosis, procedures, complications, with dates • follow-up arrangements</td>
<td></td>
</tr>
<tr>
<td>Information</td>
<td>documentation of patient and carer information</td>
<td>100% of patient notes document what the patient and his/her carers have been told about the diagnosis, prognosis and treatment</td>
</tr>
<tr>
<td></td>
<td>patient support information</td>
<td>two leaflets available in 100% head and neck clinics and wards: 1. a general one on head and neck cancer; 2. a list of support organisations (national and local), and details of where to get extra information. One document on the list must be the current set of national standards (in lay terms). Register of patients willing to visit should be offered to all patients</td>
</tr>
<tr>
<td></td>
<td>Pre treatment advice</td>
<td>100 patients should be given the opportunity to meet a previous patient who has similar treatment (including radiotherapy). All specialist nurses should keep a register of such persons locally.</td>
</tr>
<tr>
<td>Information technology</td>
<td></td>
<td>100% centres have a computerised database containing the BAHNO minimum data-set. 95% patients’ entries must be updated a minimum of annually / until death</td>
</tr>
<tr>
<td>First hospital outpatient visit</td>
<td>prioritisation</td>
<td>95% by a Consultant</td>
</tr>
<tr>
<td></td>
<td>consultation</td>
<td>90% with a consultant, the remainder to have a consultant available in the building for advice</td>
</tr>
<tr>
<td>Interdisciplinary head and neck clinic</td>
<td>time-tabling</td>
<td>These should be held weekly in each centre</td>
</tr>
<tr>
<td></td>
<td>consultation</td>
<td>100% with a consultant</td>
</tr>
<tr>
<td></td>
<td>environment</td>
<td>100% with a 100% specialist head and neck liaison nurse (e.g. Macmillan)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>there should be no talking in the background during consultations</td>
</tr>
<tr>
<td></td>
<td></td>
<td>bad news should be broken in a manner consistent with King’s Fund guidelines 100%*</td>
</tr>
<tr>
<td></td>
<td>staffing</td>
<td>Head and Neck specialist surgeon(s) with skills in endoscopy and dental assessment 100%</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Oncologist with a special interest in Head and Neck Oncology 100%</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Plastic and Reconstructive surgeon 50%</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Palliative care team representative 50%</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Specialist head and neck nurse 100%</td>
</tr>
<tr>
<td></td>
<td>treatment planning</td>
<td>both a surgeon and an oncologist involved in the consultation and planning in 100% of cases</td>
</tr>
<tr>
<td></td>
<td></td>
<td>the team should use a written protocol and a bench mark to manage co-ordinated care</td>
</tr>
<tr>
<td></td>
<td></td>
<td>the aim of treatment (cure/palliation) should be documented at the first combined clinic in 80% of cases</td>
</tr>
<tr>
<td></td>
<td></td>
<td>the treatment plan should be communicated to the patient and carers verbally, and to the GP in writing, within 3 days in 75% of cases.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>80% of plans should include actual dates of admission and radiotherapy</td>
</tr>
<tr>
<td></td>
<td>patient selection</td>
<td>95% of all new head of neck cancer patients seen in a combined head and neck clinic prior to treatment</td>
</tr>
</tbody>
</table>
**Diagnosis and staging**

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Responsibility</th>
</tr>
</thead>
<tbody>
<tr>
<td>Examination under anaesthesia</td>
<td>90% of all tumours</td>
</tr>
<tr>
<td>Radiology</td>
<td>a standard imaging protocol should be applied by a radiologist with special expertise in H &amp; N</td>
</tr>
<tr>
<td>Biopsy</td>
<td>100% of new cases require a histological diagnosis of cancer pre planning</td>
</tr>
<tr>
<td>Fine-needle aspiration biopsy</td>
<td>95% of neck masses and parotids</td>
</tr>
</tbody>
</table>

**Primary Therapy guidelines**

<table>
<thead>
<tr>
<th>Surgery</th>
<th>Should include clear surgical margins in radical [non-palliative] surgery in 95% of cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Oncology</td>
<td>Standards have still to be developed, but are expected to include:</td>
</tr>
<tr>
<td></td>
<td>- delay prior to onset of treatment</td>
</tr>
<tr>
<td></td>
<td>- treatment morbidity</td>
</tr>
<tr>
<td></td>
<td>- nutritional input / gastrostomy usage</td>
</tr>
<tr>
<td></td>
<td>- functional outcomes</td>
</tr>
</tbody>
</table>

**Outcome**

30 day and 3 year relative survival to be recorded in 98% of patients

**Follow up**

GP’s should be faxed a typed discharge summary within 48 hours of discharge from hospital in 90% cases

---

**Rehabilitation of speech and swallowing**

<table>
<thead>
<tr>
<th>Staffing</th>
<th>Centres must have a named speech therapist with at least 25% of time dedicated to head and neck cancer work and with specialist laryngeotomy skills</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Videofluoroscopy must be available in all centres</td>
</tr>
</tbody>
</table>

**Dental and prosthodontic care**

<table>
<thead>
<tr>
<th>Pre-treatment assessment and treatment</th>
<th>95% of patients with maxilla/orbital tumours should see a restorative/prosthetic team member before surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>a dental practitioner and or hygienist should see 100% of patients</td>
</tr>
</tbody>
</table>

**Follow-up scheduling**

Monthly year one, tailing to annually by year 5

At year 5, patients to be given the option of annual visits

**Research and audit clinical trials**

All centres should be encouraged to take part in national trials. A comprehensive directory of local centre Network trial participation should be available on-site.

Notification of and recruitment to clinical trials should take place as part of the treatment-planning process, with the same 3-day standard.

**Clinical audit**

There should be 100% participation in process and outcome audit, with at least one active head and neck project at all times

Participation in national head and neck audit (e.g. Royal College of Surgeons/Radiologists) in 100% of centres

The results of audit (local and national) should be published and made available to purchasers [Cancer Commissioning Groups]
| Palliative care medicine * | hospital medical and nursing staffing | all centres have written guidelines agreed with the local palliative care consultant  
100% of staff should know route of referral to palliative care (person, place, method) |
<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Breaking bad news</td>
<td>This should be done in accordance with Kings Fund guidance</td>
<td></td>
</tr>
</tbody>
</table>
| crisis planning          | 100% of centres and hospices dealing with head and neck cancer patients should have protocols for tracheostomy blockage and major vessel ‘blow-out’  
90% of ward nurses should know the protocols  
Patients at risk of these crises and their carers to be made aware of warning signs 100% |
| living with cancer       | *Patients and carers request that new standards be established to include:*  
- contact with specialist nurses, clinical psychologists and support groups  
- provision for returning to primary care as main source of support  
- financial and social support |
| notification of death    | For patients dying in hospital, the patient’s GP should be informed within one working day in 100% of cases.  
For patients dying at home or in a hospice, the patient’s Consultant should be informed of the date and location of death within 10 working days in 80% of cases |

Radiotherapy for head & neck cancer has, like most radiotherapy, traditionally been delivered in daily fractions, five days a week. Fractionation practices vary considerably between UK centres, whereas the majority of North American and European radiotherapy (and many British centres) have used single fractions of 2Gy per day (for brevity, we can call this “conventional” fractionation). A number of clinical trials in recent years have suggested that using more, smaller fractions, without reducing total dose (hyperfractionation), and with some reduction in overall time (acceleration), or simply employing either strategy, leads to better results than “conventional” fractionation\(^1\)\(^3\). At the same time (\textit{vide infra}), results from combining chemotherapy with “conventional” fractionation have also yielded benefits\(^5\). The strategy of combining modified fractionation with chemotherapy, perhaps the next logical step, remains inadequately tested, but “conventional” fractionation alone can no longer be regarded as “state of the art” except, perhaps, for very early lesions.

Radiotherapy for head & neck cancer requires great precision and this must be reproduced daily for several weeks. A complex process of immobilisation (usually using individually-made thermoplastic shells) and imaging (usually CT, performed with the immobilisation device in place) is thus necessary as a preliminary to the actual radiotherapy planning process, which in itself is quite time-consuming.

Technical advances in imaging, mechanics and computing have resulted in the real possibility of improving radiotherapy techniques by more precisely defining which areas to irradiate, which to spare, and to be able to achieve this with complex three-dimensional (“conformal”) planning. This is a rapidly-evolving field, one of the most exciting areas currently being the use of intensity-modulated radiotherapy (“IMRT”), where the radiation fluence across an individual beam is actually varied during treatment.

Interstitial radiotherapy (the insertion of radioactive sources directly into tumours), also often referred to as “brachytherapy”, is an effective form of treatment for small accessible tumours. Usually in treating head & neck cancer it is performed using wires of iridium-192, which are left in place for approximately a week. This entails significant risks of radiation exposure for staff, and comprehensive radiation protection procedures must be rigorously enforced. Its place in relation to other forms of local treatment – surgery or external beam radiotherapy – has however, never been properly assessed. A recent publication of the Royal College of Radiologists advises that this treatment should only be performed in centres with adequate volume of work\(^5\).

Radiotherapy is also a very effective treatment for palliation of certain symptoms in incurable patients, notably bone pain associated with metastases, but the place of radiotherapy as a palliative measure for incurable, locally-advanced disease is debatable.

Radiotherapy should be delivered in an accredited department. This will have undergone assessment according to the national radiotherapy standards defined in the Manual of Cancer Services\(^6\).

These ensure that radiotherapy services are of a high quality through:

1) Clearly defined leadership and organisational arrangements.

2) Provision of adequate professional staffing and equipment.

Treatment should be planned by a specialist Clinical Oncologist with access to a mould room, 3D planning facilities, beam blocking techniques and portal imaging so as to ensure to geometric accuracy and reproducibility of planning, with minimisation of dose to neighbouring normal but radiosensitive structures.
3) Minimising delays for treatment and breaks in treatment. This is particularly important for squamous head and neck cancer where unscheduled prolongation of overall treatment time reduces the cure rate by at least 1% per extra day. Not more than 10% of radical courses should be prolonged by unscheduled breaks. Where treatment gaps are unavoidable they should be managed using the Royal College of Radiologist guidelines.

4) Use of standardised processes for prescribing and checking radiotherapy treatments.

5) Use of standard principles for the delivery of radiotherapy.

6) Clear documentation of treatments delivered.

7) Quality assurance processes for radiotherapy treatments. An externally reviewed quality assurance programme is likely to be in place to ensure that these principles are covered. An example of this is ISO 9000:2002.

8) Implementation of protocols to minimise treatment toxicity. These should:
   a) adequately inform patients of common toxicities.
   b) ensure that dentate patients are assessed pre-radiotherapy by a restorative dentist/hygienist.
   c) ensure weekly review of all cases by clinician/specialist health care professional and dietician.

9) Participation in network agreed clinical trials, especially those involving quality assurance and peer review.

The Joint Council for Clinical Oncology recommended that for radical courses of radiotherapy the delay from deciding to treat to the actual start of treatment should not exceed four weeks, and this recommendation has been adopted by the Manual of Cancer Services. To complete the preparatory work and planning within this time scale is challenging, but must be the standard that radiotherapy strives to achieve.

Some of the more pressing gaps in evidence-based knowledge in the field of head & neck cancer are:

- What are the relative roles of radiotherapy (including brachytherapy) and conservative surgery in the management of early disease?
- Can the strategies of chemoradiation and modified fractionation, both of which have been shown independently to be superior to “conventional” fractionation alone, be combined?
- In combining chemotherapy and radiotherapy, what are the best drugs, what is the best route (arterial or venous), and what is the best scheduling of drugs in respect to radiation?
- To what extent do the improved results obtained with altered fractionation and chemoradiation make organ preserving strategies (i.e. non-surgical primary approaches to advanced disease) more widely applicable?
- Is radiotherapy useful as a palliative tool for incurable, locally-advanced disease, and if so, for which patients?
References:


Chemoradiation

Chemotherapy is likely to play an increasing role in the management of advanced head and neck cancer. Three meta-analyses have shown that chemotherapy can improve survival when added to radiotherapy (1-3). The improvement is modest, and at the cost of increased acute toxicity.

What meta-analysis cannot identify is the “optimal” chemoradiation schedule. The combination of cisplatin and fluorouracil is the most widely-accepted “standard” chemotherapy, but evidence to support its use above other drugs or combinations is not conclusive.

The best timing of drug administration in relation to radiotherapy is also an unresolved issue. Chemotherapy can be combined with surgery and radiotherapy in three main ways:

I. Neoadjuvant (induction) chemotherapy – given prior to radiotherapy or surgery. This improves local control but does not have a significant effect on overall survival. It does however allow greater organ preservation.

II. Concurrent chemoradiotherapy – only concurrent chemoradiotherapy has been shown to be consistently associated with a significant survival advantage. The most recent meta-analysis showed an 8% absolute survival benefit from chemotherapy (3).

III. Adjuvant chemotherapy – given after radiotherapy or surgery. This should only be given in the context of a clinical trial for patients at very high risk of recurrence.

Although the trend is moving from neoadjuvant (induction) chemotherapy towards concurrent chemoradiotherapy, delays in starting radiotherapy treatments in the UK at the moment mean that good performance patients may benefit from both neoadjuvant and concurrent chemotherapy.

(a) LOCALLY ADVANCED UNRESECTABLE DISEASE

Four recently published randomised trials comparing concurrent chemoradiotherapy with radiotherapy alone have shown significant differences in both loco-regional control and survival (4-7).

The absolute survival benefit was between 9% and 24% for chemoradiotherapy.

Concurrent chemoradiotherapy should be considered for good performance status patients with locally advanced unresectable disease. Poor performance status patients should receive radiotherapy alone.

(b) LOCALLY ADVANCED RESECTABLE DISEASE

A recent study in advanced laryngeal cancer demonstrated that 88% of patients preserved their larynx at 2 years with concurrent chemoradiotherapy compared to 69% in patients treated with radiotherapy alone (8). Another study in advanced oropharyngeal cancer reported a 20-25% absolute improvement in local control and survival when chemotherapy was given concurrently with radiotherapy (9). Concurrent chemoradiotherapy is an alternative for good performance status patients who would prefer to avoid surgery in stage III and IV cancer of the larynx and oropharynx (10-12).

(c) NASOPHARYNGEAL CARCINOMA

Concurrent chemoradiotherapy has proved superior to radiotherapy alone for advanced disease (13). Al-Sarraf et al (14) demonstrated an increase in 5-year survival from 47% to 78% when chemotherapy was added to radiotherapy.
(d) CHOICE OF REGIMEN

The choice of cytotoxic and the optimum regimen are not well-defined. The most commonly used drugs are Cisplatin, 5-FU and Methotrexate. Recent very promising results have been seen with the addition of newer drugs such as Taxanes to Cisplatin and 5-FU (15-19).

DELIVERING CHEMOTHERAPY

Chemotherapy should only be delivered by centres/units fulfilling the national chemotherapy standards. These ensure that chemotherapy services are of a high quality through:

1. Clearly defined leadership and organisational arrangements.
2. Provision of dedicated and suitably equipped areas for the administration of chemotherapy.
3. Co-ordination and control over the use of specified chemotherapy regimens within a network.
4. Supervision of chemotherapy by appropriate specialists (clinicians and pharmacists).
5. Administration of chemotherapy by appropriately trained staff.
6. Use of guidelines for the prevention and treatment of side effects and implications of chemotherapy.
7. Minimising delays in starting treatments.
9. Clear and comprehensive documentation of chemotherapy delivery.

Photodynamic Therapy

Requires the systemic administration or topical application of a photosensitizing agent, which is activated by a specific wavelength of light related to the physical properties of the sensitizing agent.

The photosensitiser acts as a vehicle to allow the transfer of energy to produce highly reactive singlet oxygen, which causes cellular damage and vascular shut down resulting in tissue necrosis.

Second generation systemic photosensitisers are highly efficient generators of singlet oxygen and can be activated by simple to use computer controlled diode lasers. Light may be administered directly onto the tumour surface where the degree of tissue penetrance is related to the wavelength of the administered light. The longer the wavelength of light the greater the tissue penetrance. Red light of 652nm will penetrate soft tissue reliably up to 1cm. Light may also be delivered to a tumour via interstitial methods and accurate calculations of tumour necrosis made.

Photodynamic therapy may be used for management of low volume disease of the head and neck as well as palliation of end stage disease.

Currently Photofrin and Foscan are licensed for photodynamic therapy for squamous cell carcinoma in the head and neck.

References:


Chapter 1 Quality of Life Assessment

The evaluation of the quality of life (QOL) in people with head and neck cancer is integral to best patient care (1). Survival may be the initial primary concern of patients (2), (3) however after treatment there tends to be a shift towards maintaining or improving their quality of life (4). Health-related quality of life (HRQOL) is an important measure of outcome as different treatments sometimes can confer relatively small gains in survival but at the cost of disrupting the most basic of human functions such as speech, appearance, swallowing, chewing and social interaction. Health-related quality of life will be considered in the following sections:

1. What is quality of life?
2. Why should we measure quality of life?
3. Who should measure quality of life?
4. How should it be measured?
5. When should it be measured?
6. What are the key issues?

WHAT IS QUALITY OF LIFE?

The available literature supports the difficulty in defining and measuring quality of life. (5), (6) It is a broad concept that incorporates all aspects of an individual’s existence. Aaronson et al describe QOL as essentially having four core dimensions functional status, physical complaints (such as symptoms of disease), psychological distress, and social interaction. (7) The term ‘health-related quality of life’ is more focused and points to those aspects of life that are affected by healthcare interventions. (8).

WHY SHOULD WE MEASURE QUALITY OF LIFE?

The inclusion of ‘quality of life’ into treatment planning and appraisal has many potential benefits. It encourages multidisciplinary team working, helps identify poor outcome groups, gives better information for the patient and their carers and the opportunity to identify problem areas and so target support and intervention. (9).

When two methods of treatment produce equivalent cure rates one of the major factors determining which treatment should be chosen is the post-treatment quality of life. The principle of HRQOL measurement forms part of the national head and neck advisory datasets. (The British Association of Head and Neck Oncologists and the British Association of Otorhinolaryngologists Head Neck Surgeons).

WHO SHOULD MEASURE QUALITY OF LIFE?

The literature supports the view that HRQOL should be recorded from the patient’s individual perspective. There are significant differences when health care professionals rather than patients measure quality of life (10). The simplest way to measure HRQOL is by patient self-completed questionnaire although other methods include open and semi-structured interview. One member of the multidisciplinary team should have clear and agreed responsibility to collect HRQOL data.

The results for an individual and for groups of patients should be communicated to the rest of the team in order for appropriate action to be taken such as psychosocial support, treatment for depression and the adjustment of treatment protocols. An interesting development is the inclusion of the patients rating of the ‘most important’ domains (11).
It may be that the combination of changes in individual domain scores linked with the importance rating will serve to highlight the complex functional alterations that occur with treatment and provide an indication for targeted therapeutic interventions.

**HOW SHOULD IT BE MEASURED?**

Questionnaires give a structured insight into the patients’ perspective. The choice of questionnaire depends on many factors including the aim of the study, its design and the available resources. If the measure is for patient self-completion it needs to be easily understood and take less than 10 minutes to complete (12). A different questionnaire would be appropriate to use in routine practice as a basic indicator of HRQOL as compared to that in a research setting. Although there are at least 14 validated head and neck questionnaires most workers in this field recognise that there is no one instrument ideal for all purposes (13), (8). They are each different in style and content and should be selected after consultation with the multi-disciplinary team so that a consensus is reached. If the intention is for data to be compared with other units or nationally it is important that there is a common questionnaire in use.

The questionnaires most commonly in use are:

**University of Washington Head and Neck cancer Questionnaire (UW-QOL)**

The UW-QOL has undergone three major revisions since it was first published. In version 2, an importance-rating scale was added. In version 3 two new domains (taste, saliva) were added and the employment domain dropped and in the latest version mood and anxiety has been included. There is no charge for use and it is in the published domain.

The questionnaire is available at the University of Washington website. [www.depts.washington.edu/soar/projects/dxcat/hnca/qol_uw.htm](http://www.depts.washington.edu/soar/projects/dxcat/hnca/qol_uw.htm)

Postal address: Surgical Outcomes & Research (SOAR)
1660 South Columbian Way
HS 112 OTO
Seattle WA 98108
USA

**European Organization for Research and Treatment of Cancer (EORTC)**

This is a modular approach with assessment of generic aspects of QOL (30 questions) and a module specific to head and neck (35 items). Both the QLQ-C30 and the QLQ-H&N35 are copyrighted instruments and require the completion of a User's Agreement form. There is no charge for academic use but if the study is sponsored there is a royalty fee.

Further information is available at the following web address: [www.eortc.be/home/qol/](http://www.eortc.be/home/qol/)

Postal address: EORTC Data Centre
QOL Unit, Ave. E. Mounier 83, B-11
1200 Brussels, Belgium

**Functional Assessment of Chronic Illness Therapy (FACIT)**

The FACIT scales are modular with the general version (FACT-G, 27 questions) used with patients of any tumour type and a head and neck cancer component (FACT-H&N, 11 questions).

Translations in a range of languages are available. All collaborators must complete a collaborators’ project information form prior to use of any FACIT scales. There is no charge.

Further information is available at the following web address: [www.facit.org/facit_questionnair.htm](http://www.facit.org/facit_questionnair.htm)
WHEN SHOULD IT BE MEASURED?

Questionnaires can be administered as part of either a cross sectional or longitudinal study. Longitudinal data from pre-treatment has the distinct advantage of allowing the measurement of change and also recording HRQOL during the different phases of treatment. The HRQOL at baseline gives an indication of outcome in different clinic-demographic groups such as size of tumour, combine treatments (surgery +/- adjuvant radiotherapy) (14).

It is a logistical challenge to ensure patients self-complete questionnaires before treatment and at regular intervals subsequently. However, prospective study allows for comparison over time for the same individual and this can be used to trigger interventions. The poorest scores are seen at 3 months and 6 months following primary treatment (15), whilst HRQOL at 1 year gives an indication of long-term outcome (16).

The frequency of data collection will depend on how the data is being used in clinical practice and the resources available. It is recommended that where possible questionnaires are completed at pre-treatment, 6 months and 1 year.

WHAT ARE THE KEY ISSUES?

There are a large number of articles published each year reporting HRQOL in patients with head and cancer. Key issues relate to tumour site, treatment options, and a variety of other HRQOL issues.

TUMOUR SITES

Published studies have demonstrated a difference in HRQOL profile between cancer sites. Generally the worst groups are oropharynx and hypopharynx as many domains are affected. (15)(17), (18), (19), (20).

TREATMENT OPTIONS

There are very few studies that have attempted to compare surgery and radiotherapy in matched groups (21), (22), (23) however in general HRQOL outcome has been found to be much better following single modality treatment (24). Combined treatments involving both surgery and radiotherapy confer much poorer HRQOL than single modality treatment. (15), (25), (26).

Some of the major themes are summarised in the list below and for brevity there are only two references cited:

Coping (27), (28)
Dental status (29), (30)
Disfigurement (31), (32)
Emotion (33), (34)
Speech (35), (36)
Oral Rehab (37), (38)
Swallowing (39), (40)
Shoulder (41), (42)
Social support (43), (44)
CONCLUSION

To date very few centres in the UK are routinely measuring HRQOL in their caseload of patients either by cross sectional or prospective study. The reasons are multifactorial and include the lack of guidance on which measure to use, emphasis on other measures of outcome such as survival and complication rates, time constraints and manpower restrictions. There is a need for clinicians to derive positive gain from acquiring HRQOL data in order to inform clinical practice and have direct relevance to improving patient care. It is only by overcoming these problems that progress can be made. The adoption of a common questionnaire would allow for multicentre HRQOL data collection and facilitate both specific sub-site and treatment analysis which is not possible with single site data collection.

References:

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INTRODUCTION

There have been major advances in speech, swallowing and voice rehabilitation for patients with head and neck cancer in recent years (1-5). Effective rehabilitation allows the patient to minimise the disability of the surgical and or radiotherapy/chemoradiation treatment by finding alternative or compensatory strategies. Speech, swallowing and voice rehabilitation should be available to patients with head and neck cancer. Rehabilitation should be team orientated, with the specialist speech and language therapist assigned the key worker role, in these aspects of head and neck cancer care.

Active rehabilitation should aim to:

- assess and manage speech and swallowing difficulties, ensuring treatment is specifically aimed at maximising residual function and introducing compensatory strategies where appropriate

- facilitate neuromuscular recovery wherever possible (i.e. post-radiotherapy)

- provide compensatory aids to assist or replace function

- teach patients and carers how to manage speech/voice/swallowing difficulties within the realistic confines of their disability

- help the patient and their carers to cope with the consequences of a major life-changing situation.

- help the patient and their immediate family/carer to adjust to the huge changes from their pre-morbid function, and where realistic assist in restoring as much of the pre-morbid lifestyle as possible

- promote choice and autonomy in partnership with patients and carers

Service Delivery

- The lead specialist speech and language therapist in head and neck cancer should be highly skilled, have significant clinical experience in the field and have appropriate post graduate training. The specialist speech & language therapist should be responsible for setting standards, devising local policies and developing pathways of care. The therapist will adhere to Clinical Guidelines in Head & Neck Cancer published by the Royal College of Speech & Language Therapists (RCSLT).

- Appropriate staffing levels are essential to delivering a quality rehabilitation service that allows for intensive therapy, multidisciplinary liaison, audit etc. (1-6).

- Speech and language therapists working in the field, will participate in peer review to ensure best practice and adherence to guidelines. This may be done locally or regionally.

- In cases of transfer to local speech and language therapy services the clinician should have an adequate level of skill and access to specialist advice and training. Communication should be two-way with close liaison to ensure continuity of care within the extended MDT. Discussion of this complex issue is beyond the scope of this document (see future directives section at the end of this chapter).

- Time and funding should be made available for the continuing professional development.
There should be an active teaching and training policy within the Unit to achieve a cohesive multidisciplinary approach. This continuing education programme should be available to all members of the team and they should be familiar with how to access it.

There should be a commitment from speech and language therapy to training non-specialist speech and language therapists and other health professionals in the community.

RESOURCES

Sufficient funding should be available for all communication aids, SVR and swallowing equipment prescribed by the speech and language therapist.

The specialist speech & language therapist will have adequate access to equipment and expertise for the assessment of speech, voice & swallowing e.g. Fibreoptic Evaluation of Swallowing, Videofluoroscopy. (7, 8, 9, 10).

Tracheostoma valves should be offered to all laryngectomy patients who undergo SVR and are assessed as suitable candidates. A choice of tracheostoma valves should be available.

THE REHABILITATION PROCESS

a. Pre-treatment period

The speech and language therapist should be present at the combined clinic treatment planning meeting in order to discuss rehabilitation issues. All patients who are likely to experience significant speech swallowing/voice problems should be seen by speech and language therapy.

The speech & language therapist requires information regarding the extent of the resection as this may be correlated to the extent of speech, voice & swallowing impairment (11, 12, 13, 14, 15, 16) The nature of the surgical reconstruction may also affect the patient’s functional ability postoperatively (12,17).

The aims of the initial appointments are to:
1. Carry out appropriate assessments of speech, swallowing and voice within a psychosocial context.
2. Provide information on speech, swallowing and voice to the patient/carer.
3. Specific communication needs should be taken into account, e.g. people with literacy/visual problems will have access to video/audio information.

Where ever possible realistic aims and goals are discussed with the patient and family.

b. Early post-treatment period

All patients require a post-treatment re-evaluation of baseline rehabilitation needs – e.g. swallowing, voice and speech status.

In the surgically treated patient, speech and language therapy begins when the surgeon indicates that the patient’s healing has progressed to the point where there is no danger to the suture lines.

It is important to recognise that the rehabilitation options should be tailored to meet the needs of the patient.
• This stage is often the optimum time for speech & language therapists to direct swallowing management techniques and range of motion exercises in order to achieve the goal of re-establishing safe oral feeding, or if this is not possible to determine whether a patient needs total or supplementary non-oral feeding. (Ref. 15,18) Refer also to NICE Guidelines. These factors should be taken into consideration when planning discharge and early discharge should be avoided at the cost of compromising optimal swallowing and nutritional care.

• Therapeutic work should be commenced prior to sutures healing e.g. work on intelligibility with silent mouthing or work with carers and patients to assist them in coping with the frustration of communication breakdown.

c. In-patient discharge

• Discharge is potentially the most difficult time for the patient (6). Discharge should be planned and agreed by all members of the rehabilitation team. Care should be made to prepare the patients for discharge and appropriate support networks should be in place.

• The timing of the discharge should be made known to all the relevant parties (i.e. GP. Social worker etc).

• There should be liaison between the rehabilitation team with Community staff and Support Services including Hospice care, to ensure a seamless continuity of care (2).

• The patient should be quite clear of how and where to get further advice before the follow-up visit.

• or patients whose long term communication remains poor, family members may find lip reading skills and training helpful. Consideration should be given to methods of telephone communication, summoning emergency aid etc.

d. On going care

• Out patient therapy will be tailored to patient’s individual need.

• The specialist speech & language therapist will be aware of the potential side affects (temporary and chronic) from radiation and chemoradiation treatments for head & neck cancer.

  The speech & language therapist and patient will remain in contact throughout the period of treatment and thereafter. If the patient suffers with side affects that prevent regular rehabilitation management, progress needs to be monitored and active treatment reinstated as soon as the patient is able to comply (19, 20, 21, 22).

• The multidisciplinary and community team will be kept updated regarding the patient’s progress and prognosis in therapy.

• Patients should be made aware that there is open access after discharge and know how to re-refer.
SPECIFIC CONSIDERATIONS FOR LARYNGECTOMY PATIENTS

- The surgeon and speech & language therapist should plan the most appropriate vocal rehabilitation - oesophageal voice, the use of tracheo-oesophageal voice prosthesis, an artificial larynx or a combination. It is important to respect the patient’s preference in the planning process.

- The speech and language therapist should be the key worker with laryngectomy patients in terms of rehabilitation of voice and swallowing and prosthesis selection. Nurses also have an important role, especially in out of hours cover for patients with voice prostheses.

- An active programme of surgical voice rehabilitation is essential in every centre where laryngectomy is performed (1,3,7,8). Primary SVR, i.e. a puncture undertaken at the time of primary surgery is now accepted as the gold standard for rehabilitation, if the patient is judged as a suitable candidate by both the ENT Consultant and the Speech and Language Therapist (23, 24, 25,). Patients should only be deselected for primary puncture when there are clear contraindications, these should be assessed by both the ENT Consultant and Speech and Language Therapist.

- Considerable expertise is required from both the surgeon and speech and language therapist to provide a SVR service appropriately and there should be a commitment to continuing education and clinical audit.

- A choice of indwelling and exdwelling valves and valve brands should be offered at all units in order that valve selection can be tailored to patient’s needs and preference. Only a small number of studies have been carried out comparing voice prostheses and their pros and cons. Further studies are needed (26, 27).

- A 24-hour SVR service is essential for all units, as urgent problems with valves are inevitable. A rolling training programme should be offered for ward nurses and junior medical staff as they are first in line for out of hours emergencies.

- All units should have access to videofluoroscopy with audio recording to assess pe segment function. This may be in house or as a tertiary referral. This should include access to botulinum toxin injections under videofluoroscopic guidance to treat spasm or marked hypertonicity of the p-e segment (28, 29).

- Primary myotomy or neurectomy is recommended as a routine part of surgery to maximise the chance of successful voice outcome.

This includes patients deselected for SVR as it creates optimal conditions for the acquisition of functional oesophageal voice (30).

- Manometry (to measure tracheal air pressure) should be available to assess patient suitability for a “hands free” tracheostoma valve and to train patients to minimise back pressure when voicing with this type of valve (31).

- Patients should be retained on an inactive caseload once routine rehabilitation and review ceases. SVR patients should attend for annual review. Electrolarynx and communication aid users should return their aid for an annual safety check to be carried out by the electronics department at each unit.
Specific Considerations for Tumours of the Oral / Oropharynx and Laryngeal Conservation Treatment

- The precise functional effects of the treatment are generally difficult to predict generally cannot be provided in detail for the individual during preoperative counselling. Counselling focuses on informing the individual that there are likely to be changes in speech and swallowing after treatment. (15, 33). Patients are informed about the rehabilitation process and that it will be their responsibility to practise and follow rehabilitation strategies.

- A multi-disciplinary team discussion regarding alternative/augmentative feeding is required when patients are anticipated to have considerable problems maintaining adequate oral intake either because of the tumour or the expected outcome of their treatment (34).

- There is evidence to demonstrate the benefits of speech & language therapy interventions to improve function for both speech and swallowing post radiotherapy (15). Currently there is under resourcing of specialist speech & language therapy services nationally, with particular limitations in radiotherapy units.

- Therapy continues until a patients swallow has reached a point where the speech & language therapist and the patient agree that maximum goals have been attained. Often the ultimate functional outcome cannot be determined until several months post treatment and then involves discussions between the various members of the team seeing the patient (15).

SPECIFIC CONSIDERATIONS FOR PALLIATIVE CARE

Orally feeding the dysphagic patient at the end stage of life
The Speech and Language Therapist has a role in ensuring that appropriate information is given about the choices and risks facing dysphagic patients on their caseload, who may wish to continue orally feeding in the end stage of life. Decision making and reviews should be made in the context of the Multidisciplinary Team. In terminal care this team could be the Cancer Centre Multidisciplinary Team, community GP/Macmillan Nursing team, or hospice team. Community Speech and Language Therapists should agree shared goals with their Primary care team, and have access to cancer centre specialist Speech and Language Therapists, if necessary, for support and advice where shared care, or onward referral has taken place. More consensus needs to be reached by Royal College of Speech & Language Therapists on this issue.

OUTCOME MEASURES

Currently there is no guidance or agreement nationally, regarding which parameters should be measured for both clinical or research purposes, or indeed, who should be collecting the data and at what time intervals. Many units do not have adequate speech & language therapy staff to enable them to collect outcomes routinely, even for clinical practice. A number of the quality of life tools such as the SF36, Washington and EORTC also include measurements of some of these parameters and this section should be read in conjunction with the quality of life chapter. In addition to these tools there is a widely known speech & language therapy outcome measurement package – the Enderby Therapy Outcome Measures (TOMS) (35) which rate Impairment, Handicap, Disability and Distress. There are sections for dysarthria, dysphonia, dysphagia and laryngectomy. A number of clinicians are also using outcome measurement packages that have been devised locally or that are not specifically designed for head and neck cancer. Others may be using modified versions of standardised tools, which affects the validity of any information collected. There are currently no assessments for dysphagia post- laryngectomy or consensus on a “normal” versus disordered swallowing after this type of surgery.
This is one of many areas that require more research to develop evidence based practice. A range of speech, laryngeal voice, alaryngeal voice and swallowing parameters, and perceptual and instrumental tools used to measure them are listed below:

<table>
<thead>
<tr>
<th>Speech Parameters</th>
<th>Perceptual measures</th>
<th>Instrumental measures</th>
</tr>
</thead>
<tbody>
<tr>
<td>Speech intelligibility</td>
<td>Assessment of Intelligibility of Dysarthric Speech</td>
<td>Spectography</td>
</tr>
<tr>
<td>Range, Strength, accuracy of articulation</td>
<td>SF 36 Frenchay Dysarthria Assessment</td>
<td>Spectography Electro-palatography</td>
</tr>
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<table>
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<tr>
<th>Laryngeal voice Parameters</th>
<th>Perceptual measures</th>
<th>Instrumental measures</th>
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</thead>
<tbody>
<tr>
<td>Voice quality</td>
<td>GRBAS Vocal Profile Analysis Buffalo</td>
<td>Acoustic analysis Laryngograph</td>
</tr>
<tr>
<td>Patient ratings of function / acceptability</td>
<td>SF 36 Vocal handicap Index Patient questionnaire of vocal performance</td>
<td></td>
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<tr>
<td>Laryngeal function</td>
<td>Fibreoptic profiles Stroboscopy profiles</td>
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<tr>
<th>Laryngeal voice Parameters</th>
<th>Perceptual measures</th>
<th>Instrumental measures</th>
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<tbody>
<tr>
<td>Voice quality / intelligibility</td>
<td>Wepman Semantic 7 point scale</td>
<td>Spectrography</td>
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<tr>
<td>Patient ratings of Function / acceptability</td>
<td>Vocal handicap Index Acceptability ratings</td>
<td></td>
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<tr>
<td>P.E. Segment function</td>
<td></td>
<td>Videofluoroscopy Manometry</td>
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<tr>
<th>Swallowing Parameters</th>
<th>Perceptual measures</th>
<th>Instrumental measures</th>
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<tr>
<td>Swallowing Function</td>
<td>Penetration-Aspiration Scale</td>
<td>Videofluoroscopy (VFSS) Fibreoptic Endoscopic Evaluation of Swallow Study (FEES)</td>
</tr>
<tr>
<td>Patient ratings of Function/ acceptability</td>
<td>SWAL-QOL</td>
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</tr>
</tbody>
</table>

A full reference list is available from Speech and Language Therapy Departments at Sunderland Royal Hospital or Freeman Road Hospital, Newcastle-upon-Tyne. Email: anne.hurren@chs.northy.nhs.uk paul.carding@ncl.ac.uk
IDEAS FOR FUTURE DIRECTIONS

1. There is a need for collecting information concerning functional outcomes nationally for Head & Neck Cancer patients. There needs to be more consensus regarding appropriate outcome measures and methods of collection and analysis. This can be a time consuming process and can only be achieved with adequate staffing. This information can be used for establishing best practice.

2. There is a need for a national strategy for prioritising and coordinating a research and audit plan in speech, voice and swallowing in head and neck cancer.

3. There are currently differences in levels of service provision and skill amongst non-specialist SaLTs across the UK. The degree of responsibility for patient care differs according to local agreement. As a profession we need to define skill level, competencies, guidelines for education and training as outlined by the National Cancer Plan.

We also need a consensus on access to appropriate support, clinical supervision and peer review. Roles, responsibilities and standards for provision of service between hospital-based specialist speech and language therapists and non-specialist community-based therapists needs clarification. This will require planning at a local and national level in keeping with RCSLT and NICE guidelines.

4. Patient and carer involvement should remain at the core of planning speech & language therapy services.

Core References:
2. Communicating Quality II (1997), Royal College of Speech and Language Therapists, London.

References:
**Section 4  Chapter  3  Nutritional Aspects**

**Introduction**

The role of nutrition in both the amelioration and treatment of the head and neck cancer patient has become increasingly recognised in recent years. Most of the data linking symptom control and therapy have been largely epidemiological however recent advances in molecular biology and genetics are now allowing a greater focus on “cause and effect”.\(^1\) It is possible that as many as 80% of cancer cases may be lifestyle related. In this context diet, tobacco use and/or alcohol consumption are likely to be significant risk factors in the development of head and neck cancer.

Advances in technology enhance the ability for earlier diagnosis and treatment with reduced morbidity. In addition increased survival rates mean that patients are now living long enough for deleterious long term effects of therapy to occur including the development of secondary malignancies. These present new challenges in the provision of nutritional support and a greater understanding the pathophysiology of cancer cachexia.

These patients, due to the position of tumours, are particularly susceptible to malnutrition and which if untreated can progress to cachexia. Although the syndrome of cachexia results in a profound loss of both adipose tissue and skeletal muscle mass, it is the latter which determines both the quality of life and overall survival.

Nutrition has become an increasingly important quality of life issue surrounding the psychosocial and rehabilitative needs of many head and neck cancer patients. Quality of appetite and the ability to eat are often regarded as crucial components in patients’ definitions of their physical well-being and symptom control.\(^2\) Nutrition should be considered an integral part of treatment irrespective of whether the therapeutic intent is cure or palliation. It is essential to remember that intervention is likely to be of most benefit and arguably most cost effective when concurrent clinical treatment is being undertaken. The moribund patient is unlikely to benefit to any significant extent from nutritional support. Professional carers “have a duty to prolong life, but not to inappropriately prolong dying”.\(^3\)

**Effects of Treatment on Nutritional Status**

**Surgery**

Although multimodal treatment approaches have evolved and improved markedly over recent years surgery remains the primary and often the only cure for some tumours. Any surgery results in varying degrees of catabolic stress and increases the risk of compromising nutritional status. When surgery is undertaken for non systemic pathological conditions in otherwise healthy, well nourished patients, nutrition is usually not a risk factor, at least not in the immediate post-operative period. However when a patient is malnourished and debilitated by an ongoing disease process, complications or associated therapy nutritional status becomes a significant risk factor in clinical outcome.\(^4\)

Even minor oropharyngeal surgery may lead to the development of impaired taste and smell and impaired chewing and swallowing which can temporarily, or in some cases, permanently render patients unable to maintain an adequate dietary intake.

The majority of patients undergoing a radical resection are likely to require enteral or occasional parenteral nutrition certainly in the immediate post operative period and often for several weeks thereafter.\(^5\) It is important to ensure that appropriate protocols are planned in advance and in place so that the nutritional requirements of patients can be met to best effect.\(^6\)
This will involve a co-ordinated interdisciplinary approach, otherwise there is a risk of patients being overlooked, particularly in the current environment of shorter hospital admissions and out patient follow up. Long term changes in nutritional requirements may be necessary as a result of surgery. Wound healing and the integrity of grafts are markedly affected by nutritional status. This is an important consideration in the rehabilitation process. While surgery for the purposes of reducing tumour bulk or the resection of metastatic disease does not in itself present nutritional risks which are different from those for other similar surgical procedures; nevertheless nutritional requirements will be affected to a certain extent. For example head and neck surgery and its effects may lead to changes in activity levels and in body composition. Some patients will expend increased levels of energy during rehabilitation and adaptation while others may become relatively sedentary.

Additionally one of the main aims of reconstructive and rehabilitative head and neck surgery will wherever possible be to restore the patient to normal or near normal oral function. Even if this is not entirely possible any symptomatic relief will enhance a patient’s quality of life and significantly improve the chances of an adequate nutritional intake being achieved. When food can be eaten normally the psychological benefits of improved self-image are considerable.

**Radiotherapy**
The negative effects of radiotherapy on nutritional status and oral feeding are well documented. These can be ameliorated by establishing nutritional support at the beginning of (or preferably before) irradiation. Early and ongoing nutritional support improves the chances of patients completing full courses of therapy and may reduce morbidity in the head and neck group.

Much of the effect of radiotherapy on nutritional status is related to the location of the tumour which can also determine the type and amount of radiation delivered (or possible).

Brachytherapy may result in local complications such as dysphagia and odynophagia particularly in the oropharynx.

The effect of any radiation injury can be acute or chronic and generally depends on whether single or cumulative doses are being administered. The common effects of radiotherapy and nutritional status in head and neck cancer are listed in Table 1.

**Table 1 - Effects of radiotherapy in the head & neck region on nutritional status**

- **Oropharyngeal Irradiation**
  - Impaired taste and smell accompanied by food aversions
  - Stomatitis
  - Xerostomia
  - Mucositis
  - Trismus
  - Dental caries
  - Mandibular osteonecrosis

- **Oesophageal Irradiation**
  - Dysphagia
  - Oesophagitis
  - Oesophageal stricture
  - Odynophagia

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Xerostomia is usually the most immediate and lasting side effect of radiotherapy to the head and neck. It can be induced by as little as 6-7 Gray (Gy) units given in two or three doses. If the parotid glands (or at least one gland) can be spared during irradiation and parotid function preserved there is evidence to suggest that xerostomia may be prevented or minimised; and that consequently oral intake and body weight can be maintained particularly with early stage tumours.\textsuperscript{12}

At a cumulative dose of about 20 Gy mucositis and loss of taste usually occur. At this stage oral intake becomes not only mechanically difficult and painful but also pleasureless. Additional problems such as alterations in the viscosity of saliva and in oral pH with an increased oral bacterial concentration may predispose to a greater risk of dental caries. Any oesophageal involvement is likely to result in significant dysphagia.\textsuperscript{13}

**Chemotherapy**

The gastrointestinal epithelium is particularly susceptible to the effects of chemotherapeutic agents.\textsuperscript{14} The most common and immediate side effects of chemotherapy are nausea, vomiting and anorexia. If untreated this will lead inevitably to a decreased oral intake, fluid and electrolyte imbalances, general malaise and weight loss. The level of toxicity of the chemotherapy depends on the agent used, the single and cumulative doses, drug metabolism and excretion rates, other concurrent treatments and individual patient tolerance. For example combined use of carboplatin and cisplatin in the treatment of head and neck tumours presents a number of challenges in the nutritional management of patients. Although carboplatin has a spectrum of activity similar to that of cisplatin it is less toxic and better tolerated with reduced levels of nausea and vomiting and nephrotoxicity which may afford greater ability and desire to achieve an enhanced nutritional intake.

Common side effects associated with cisplatin and 5-fluorouracil (5-FU) additionally include mucositis and anaemia.

The side effects of newer chemotherapeutic drugs, such as taxanes and retinoids, on nutritional status have yet to be fully established.\textsuperscript{15,16} However, it is likely that these will be similar to other agents and will be dose related. Mucosal toxicity (particularly of the oral mucosa) stomatitis, ulceration, cheilosis, glossitis and pharyngitis will all interfere with the ability to maintain an adequate oral intake. Dehydration and malnutrition will often occur rapidly as a result.

Agents that induce transient xerostomia and acidification of the saliva may have implications on dental health and consequently nutritional status.

**Combined-Modality Therapy**

Although chemotherapy appears to be most effective when it is administered concurrently with radiotherapy it also exacerbates the radiation reaction. Adelstein et al have reported the concurrent use of 5-FU, cisplatin and radiation therapy in a study involving squamous cell head and neck cancer patients resulted in a significant increase in the incidence of acute mucositis, tube feeding and greater weight loss.\textsuperscript{17}

Generally acute mucositis is significantly increased when more than one chemotherapeutic agent is used in addition to radiotherapy.\textsuperscript{18} Radioprotective agents (e.g. amifostine), growth factors and cytokines which can reduce the toxicity of combined modality therapy may play an important future role in ameliorating the side effects of treatment which interfere with nutritional intake.
Practical aspects of nutrition in the head and neck cancer patient

Nutritional Assessment

It is essential for all head and neck cancer patients to undergo nutritional screening in order to identify those individuals who have, or are at risk of developing malnutrition\(^\text{19}\). There is, however, no single anthropometric or biochemical marker that can identify the nutritional status of an individual\(^\text{20}\).

Head and Neck Cancer patients frequently will have already encountered periods of malnutrition prior to Hospital admission\(^\text{21}\). Nutritional assessment is likely to be most effective if carried out by means of a team approach.

Nutritional screening is the first step in the process of identifying those who would benefit from specialised dietetic assessment. Screening may be carried out by nursing or medical staff and should be conducted on admission, to facilitate early nutritional intervention. To date there is no valid screening tool for cancer patients,\(^\text{22}\) however, there are a number of tools in use, mainly based on the Subjective Global Assessment tool. Recent weight loss, dietary intake, body composition, anthropometry, biochemical status and functional ability should all be considered. Any identified chewing and swallowing problems must be taken into consideration in order that the most appropriate and effective dietary programme is designed.

Patients who have unintentionally lost more than 10\% of their weight in 6 months, or more than 5\% in the one month before admission to hospital are likely to be clinically at risk\(^\text{20, 23}\). Early nutritional screening and counselling from a Dietitian, are essential to provide nutritional support tailored to the patients' needs\(^\text{24}\). This will improve the patient life outcome prospects.

Table 2 – Recommended energy & protein requirements

<table>
<thead>
<tr>
<th>Energy</th>
<th>Maintenance</th>
<th>Repletion</th>
</tr>
</thead>
<tbody>
<tr>
<td>25-35 Kcal/Kg/24 hours</td>
<td>maintenance</td>
<td>repletion</td>
</tr>
<tr>
<td>35-45 Kcal/Kg/24 hours</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Protein</th>
<th>Maintenance</th>
<th>Repletion</th>
</tr>
</thead>
<tbody>
<tr>
<td>0.8-1.2 g/Kg/24 hours</td>
<td>maintenance</td>
<td>repletion</td>
</tr>
<tr>
<td>1.2-1.5 g/Kg /24 hours</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Table 3 – Recommended micronutrient requirements

<table>
<thead>
<tr>
<th>Vitamins</th>
<th>A complete range of water and fat soluble vitamins especially vitamin K, B1 and B6 Antioxidants (vitamins A, C and E)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Trace Elements</td>
<td>A complete range of trace elements especially zinc and selenium</td>
</tr>
</tbody>
</table>

As previously stated many head and neck cancer patients have a poor nutritional state on the initial presentation of their diagnosis, thus early assessment and nutritional support will aim to prevent further weight loss and/or improve nutritional state.

Nutritional Monitoring

Monitoring is an essential step to ensuring the patient does not develop a compromised nutritional state, contributing to outcome\(^\text{25}\). Re-screening of patients on a weekly basis or on each admission as an in-patient is essential.
As part of the re-screening, patients should have their weight checked weekly, or on admission. Any individual's weight which has decreased by more than 2 kg over a two week period must be re-referred to the dietetic team.

The biochemical state must be closely monitored. This is especially important at the early states of enteral feeding, where re-feeding syndrome may occur.\(^2\)

During the treatment, whether that is radiotherapy, chemotherapy or combined, it is vital to support the patient nutritionally, allowing adaptation of nutritional advice depending on the side effects of the treatment. This adaptation of advice should involve the following.

**First line intervention:**
meal fortification – allowing an increase in energy (Kcal) and protein through food consumption and snacks.

**Second line intervention:**
the use of oral nutritional supplements.

**Third line intervention:**
non-oral feeding. This is ideally enteral feeding whether that is nasogastric feeding or gastrostomy feeding.

**Nutritional Management**

**Oral Nutritional Support**
Where possible patients should be encouraged and laser assisted to take food orally. As previously outlined, maintaining a normal eating pattern is important psychologically to both patients and their carers.\(^9\)

An early evaluation of current dietary intake should be undertaken. Any identified chewing and swallowing problems must be taken into consideration in order that the most appropriate and effective dietary programme is designed.

**Nutritional Support**
Nutritional support should achieve and maintain a desirable body weight within a targeted weight range and correct or prevent nutritional deficiency.\(^1\) It must be tailored to individual patient requirements.

**Dysphagia**
The primary aim in dysphagia management, once the underlying cause has been identified, is to utilise strategies and postures to ensure patients can maintain an intake of as much of a normal food consistency as possible. A full swallowing assessment should be carried out by a Speech and Language Therapist who will determine the most appropriate food consistencies to use as part of ongoing therapy.
**Table 4 – Clinical symptoms affecting oral intake**

1. Loss of teeth
2. Dry mouth
3. Poor dentition and caries
4. Excess saliva
5. Sore mouth (mucositis, oesophagitis)
6. Food Disinterest and Aversion
7. Anorexia
8. Weight Loss
9. Diarrhoea
10. Constipation
11. Aversion to Taste
12. Preference for Cold Foods
13. Nausea
14. Pain
15. Swallowing Difficulties
16. Fatigue
17. Loneliness

**Enteral Feeding**
In situations where oral intake is inadequate or not possible enteral feeding should be considered without undue delay, otherwise compromised nutritional status will rapidly occur. If it is envisaged that the effects of surgery may lead to ongoing feeding difficulties careful planning of appropriate nutritional management should be carried out preoperatively.

**Nasogastric Feeding**
In the short term (≤ 14 days) most patients requiring enteral nutrition will receive this by means of a nasogastric tube. The choice of tube is likely to depend on individual circumstances, personal preference and experience. Fine bore feeding tubes improve patient comfort and reduce tube associated complications such as pharyngitis and oesophagitis. On the other hand wide bore tubes may facilitate
bolus as opposed to continuous feeding regimens and allow for easier administration of drugs if this is necessary.

Although there is a perceived increased risk of pulmonary aspiration when wide bore tubes are used this is not borne out in clinical practice.\textsuperscript{21} The risk of aspiration can be reduced by ensuring the patient is fed in an upright position or at least having the head and shoulders raised by 30-40 degrees. In addition to problems with mucosal erosion and nasopharyngeal irritation, the likelihood of tube displacement and the resultant difficulties in replacement in the head and neck patient ultimately limit the nasogastric route as a long term feeding option.

**Gastrostomy 29-37**

When enteral feeding is likely to be needed for periods of more than 14 days, there is an inability to tolerate a naso-gastric tube or the tube itself is contraindicated due to the presence of an oesophageal abnormality, the placement of a PEG tube should be considered. The assessment process, ideally carried out by a specialist team, must also take into account the acceptability of the PEG to the patient and his/her family and carers and the patient’s long term prognosis.

The advantages of gastrostomy over prolonged NGT feeding are broadly speaking two-fold. It provides the patient with a more socially acceptable facial appearance and it avoids problems caused by the NGT itself. These include the impediment of swallowing rehabilitation, arytenoid oedema, aspiration, the prolongation of radiation mucositis, sinonasal infection and discomfort.

There are three methods of gastrostomy tube placement:

1. **Percutaneous Endoscopic Gastrostomy (PEG).** Using the usual "pull" technique, the endoscope is used to introduce a pull cord into the stomach percutaneously. This is then used to pull the PEG catheter perorally into the stomach and out percutaneously. Variations exist whereby the PEG catheter is pushed out or the stomach directly punctured but all methods involve the endoscope being passed through the upper aerodigestive tract.

2. **Percutaneous Fluoroscopic (or Radiologic) Gastrostomy.** Under fluoroscopic control, a guidewire and dilator is introduced directly into the stomach. Some methods involve gastropexy using two or three T-fasteners. It is preferable to pass a nasogastric tube to use this method, but this can be circumvented.

In the case of head and neck tumours the placement of a **percutaneous radiological gastrostomy (PRG)** is preferred and recommended for two principal reasons. Firstly the radiological procedure does not involve endoscopy which can be difficult due to access and secondly there is a high risk of transferring tumour cells from the oral maxillary region to the stoma site which can then colonise in/or adjacent to the stoma, if an endoscope is used.\textsuperscript{38} If an nasogastric tube is already in situ this can normally be used to administer barium contrast (approx 200 ml) to aid radiological visualisation. Administration of a prophylatic antibiotic regimen prior to the procedure is now standard practice in most units.\textsuperscript{39}

In most cases full feeding using proprietary enteral feeds can be established within 24-48 hours post PRG placement.

Surgical gastrostomy. This can be achieved either laparoscopically or by an open laparotomy under general anaesthesia. It is usually reserved for failed percutaneous gastrostomy or in patients with a contraindication to a percutaneous technique.

The clinical situations in which a gastrostomy should be considered are:
Surgical patients: patients in whom oral diet is not possible in the medium or long term because of post-operative dysphagia, aspiration, odonophagia or limited/painful mastication, particularly if post-operative radiotherapy is planned.

A low threshold for gastrostomy should be maintained after open partial laryngeal/pharyngeal surgery because its placement greatly assists swallowing rehabilitation and reduces the psychological pressure and frustration by inferring that the expectation of its use is medium term. A further post-operative indication is the development of a fistula necessitating a prolonged period of NBM.

Radiotherapy patients: patients due to have high intensity radiotherapy or chemo-radiotherapy for locally advanced tumours are particularly likely to have swallowing difficulties, and patients who are likely to need a gastostomy can be predicted by virtue of their tumour site, stage and comorbidities. Ideally, such patients should be identified before treatment so arrangements can be made for gastrostomy, in order to avoid interruptions in radiotherapy or chemoradiotherapy.

Palliation: gastrostomy placement is an integral component of a comprehensive palliative care package for patients with no or little oral nutrition.

Minor complications of percutaneous gastrostomy tube placement occur both in the short and long term and are relatively common (10-15%). These include skin leakage, wound infection, blockage or dislodgment of tubes. It is essential for patients outside hospital to have liaison with a head and neck nurse who can deal with long term minor complications, which usually necessitate a tube change. Major complications include aspiration and cardio-pulmonary events during the procedure. In general terms, most series report a major complication rate of 2-9%. The definition of a complication as “major” is not fixed and accounts for some variation. The mortality due to the procedure is around 0.5-1%.

No matter whichever method is used and who the operator is (radiologist, gastroenterologist, general surgeon, head and neck surgeon), agreed protocols and responsibilities need to developed to detect and deal with complications that arise in hospital.

Areas of controversy:

Which method of gastrostomy?

Gastrostomy should be performed percutaneously where possible. The choice between radiologic and endoscopic methods is determined by the following issues:

- Situations in which it is impossible or not desirable to pass an endoscope: examples of this are patients with severe dysphagia, a post-operative fistula or in a patient with either recent or delicate reconstruction. In these situations, radiologic methods are preferred.

- Metastatic deposition. There are now over 20 case reports of stomal seeding of head and neck cancers. This complication is well recognised now and many cases probably therefore not reported. The most accepted school of thought is that the tumour is seeded directly using endoscopic techniques, especially the “pull” technique, before tumour resection or treatment. Some authors have suggested either using a radiologic technique or using an alternative to the “pull” technique for PEG placement in this situation.
• Safety issues: Complication rates for either percutaneous method vary widely and probably reflect local expertise and strict adherence to a thought-out protocol rather than the technique per se. Therefore, when either percutaneous method is applicable, the one with the lowest complication rate in the unit concerned should be used. Regular audit of complication rates should take place.

The timing of gastrostomy

As a general rule, if a patient will require a gastrostomy tube, it should be done as early as possible. It is certainly better to adopt a prophylactic policy for radiotherapy patients to avoid interruptions in treatment. On the other hand, adopting an expectant policy will reduce the number of gastrostomies performed. For surgical patients, the timing may be influenced by the particular method used. Radiologic percutaneous gastrostomies are more amenable to an expectant policy. PEG’s, on the other hand, may be difficult to perform in the early post-operative period and be associated with more complications. If a PEG is to be performed prophylactically, it is more practical and safer to perform it per-operatively after tumour resection and before reconstruction. This also avoids possible tumour stomal seeding.

In conclusion, the provision of a gastrostomy service is essential in the treatment of patients with head and neck cancer. Head and neck surgeons and oncologists need to have close liaison with endoscopists, radiologists and general surgeons. Nursing support in the community is required for longer term problems associated with gastrostomy tubes. Finally, it should be noted that this procedure carries an associated mortality of 0.5-1% and protocols to deal with major complications need to be developed and audited.

Home Enteral Nutrition (HEN)

Careful discharge planning incorporating regular follow up is essential if feeding problems are to be minimised and compliance achieved. Ideally ongoing close liaison with patients and their carers should be arranged through inter-disciplinary teams. For the majority of patients where gastrointestinal function is satisfactory and once feeding has been fully established, a bolus feeding regimen will be the preferred option. This prevents the need for periods of continuous pump feeding and may facilitate the transition to oral feeding and regulate appetite.

Immunonutrition

There has been a growing interest in the concept that qualitative modulation of specific nutritional substrates may influence nutritional and immunological status without enhancing tumour growth in head and neck cancer. Ideally substrate supplementation should enhance immunogenic response, replete protein stores and possibly sensitise tumours to specific treatments. Glutamine due to its role as the preferential amino acid substrate for rapidly dividing cells such as lymphocytes, macrophages and intestinal epithelial cells has been explored in several studies investigating chemotherapy induced toxicity.

Jebb et al found no benefit in administering a low dose of glutamine (16g/day) on the degree of mucositis in patients with gastrointestinal cancer receiving 5-FU/calcium folinate (Leucovorin) combination therapy. However, in a similar cross over study Skubitz and Anderson demonstrated a significant decrease in mucositis when glutamine (8g/day) was given orally over a 28 period of chemotherapy.

Similarly arginine is known to exhibit a number of immunomodulatory actions although its benefits in head and neck cancer have not been documented.

The most promising developments involve the use of new lipid formulations containing Omega-3 fatty acids in particular eicosapentaenoic acid derived from fish oils. Omega-3 polyunsaturated fatty acids (PUFA) appear to have protective effects on the development of carcinogen-induced tumours, the growth of solid tumours, cachexia and metastatic disease.
Dietary supplementation has been tested in several clinical trials. The principal findings include significant weight gain, improved performance status and enhanced appetite.\textsuperscript{46-48}

Additionally the immunomodulating effect of Omega 3 PUFA may prolong survival rates.\textsuperscript{49} Although these studies have been carried out predominately in pancreatic cancer there are encouraging early indications that similar effects of Omega 3 PUFA are applicable across a range of different cancers.

**Summary**

Malnutrition is prevalent in head and neck cancer patients and is associated with an impaired quality of life and a reduced survival in many cases. It results from a number of multifactorial events such as an inadequate dietary intake, alterations of taste and smell, metabolic disturbances, inflammatory responses and the side effects of concurrent anti-cancer treatments.

Nutritional assessment will determine the initial nutrition therapy and on-going management. Early dietetic and speech therapy intervention is essential to ensure that the oral nutrition plan is both appropriate and safe and that regular monitoring of nutritional status is facilitated effectively. If oral feeding is not possible nutritional requirements can be adequately met by means of enteral tube feeding.

Current research is focusing on the effects of immunonutrition on the host-tumour relationship and how it may influence metabolic response. Further studies are needed to determine the efficacy of specific nutrients in terms of overall nutritional and oncological objectives and whether there are real cost benefits, in outcome, to be gained on the length and quality of life.

**References:**

3. Lennard-Jones JE. Legal and Ethical Aspects of Hydration and Nutrition. BAPEN PO Box 422, Maidenhead, Berks. 1998; 6
Principal Patient Concerns

Patients priorities are that all patients, carers and families should be supported throughout the patient journey.

a) Planning for discharge should begin at admission – all patients should be assessed by a multidisciplinary team, fully conversant with all aspects of head & neck care, professionals will begin the journey along the route which will produce the most patient centred outcome.

b) Patients feel that to ensure good outcomes co-ordination between secondary, primary, social Care and the principal carer is essential. This hopefully should ensure that support (for patient and carer) is available at the time of need, this is particularly true in the benefits scenario. Patients should be encouraged to seek support of a trained benefits advisor conversant with head and neck issues (see National Association of Laryngectomee Club’s benefits pack as a possible role model). It is extremely important that all professionals recognise that patients may have social problems not always obvious in the medical setting.

c) Patients must be given information in a timely manner and in an accessible format appropriate to their needs. To enable this there should be screening/ recognition of existing disabilities or language barriers.

d) All Health Care Trusts must ensure that all aspects of pre and post registration staff training are inclusive of all aspects of head and neck care, particularly around resuscitation of neck breathers. To support this aim it is essential that new (neck breather inclusive) resuscitation mannequins are developed and that all National Health Service staff, carers and families receive training in resuscitation techniques.

e) There is a clear need for ongoing training to include speech disability awareness and skills. This could perhaps be rolled out to include carers and families of head and neck patients.

f) Patients feel that the British Association of Otolaryngologists - Head and Neck Surgeons should be encouraged to consult with National Patient Support Associations to enable appropriate protocols for head and neck care to be established nationally.

g) The availability of pre treatment counselling for new head and neck patients is felt to be highly advisable. Wherever possible, befriending partnerships should be offered to both the patient and family. It is, however, essential for the well being of both parties that befrienders are supported, and whenever possible trained to undertake this role.

h) Concern has been expressed that patients undergoing radiotherapy may not receive adequate levels of support at that time. The National Laryngectomee Club recognises its limitations in this role, given the potential risks of non-Laryngectomee patients feeling that treatment could fail at a later stage, were Laryngectomees to be involved in support at this time.

It is recognised that there may well be an enhanced role at this time for the head and neck specialist nurse, particularly in stoma care.
i) Patients look to advances in treatment regimes to reduce the side effects of radiotherapy. However they do feel that greater use of complimentary/alternative therapies may reduce the effect upon the patient.

j) Patients should all have access to equipment appropriate to their medical/social needs – i.e. nebulisers, suction and humidifier machines, bathing aids, Servox, voice enhancer and telephonic equipment.

k) Follow up – directly as a result of the relative rarity of head and neck cancer it is felt that specialist follow up should be open ended. Patients do, however, feel that where non specialist Consultant clinics are held in district hospitals, those facilities could be used for this purpose for the 5 year+ post operative patient.

l) Patients feel strongly that no Laryngectomy should be undertaken without the patient being enabled to access, together with professionals a full choice, of voice restoration methodology. Similarly all head and neck patients must have access to a full range of cosmetic surgery and speech therapy as appropriate. Contracts between commissioners and providers must be fully inclusive of these items.

m) Research should be promoted into new treatments/therapies, and into the potential risk to laryngeal health of the papilloma virus.

n) Serious consideration should be given to an effective screening methodology for high risk groups; this may well result in a need to update primary care practitioners’ information of potential risk factors. Patients and carers recognise that for progress in this area there is a need for further research into all possible causes of head and neck cancers.

o) Patients and their carers are particularly aware of the risk factors for patients with speech impairment. The Social Services Inspectorates have produced standard guidelines for communication support for deaf, hard of hearing and deaf/blind people. Research has indicated quite clearly that no similar support is available for those with speech impairment. In fact great difficulty is being experienced in finding researchers to undertake the initial research required. Speech support groups recognise that this matter is now urgent given the requirements of the Disability Discrimination Act and Human Rights Acts and the responsibilities placed on the statutory sector, in particular, as a result.

p) Difficulties with communication should, patients feel, be taken into account whenever staffing issues are reviewed. Laryngectomees, older patients in particular, do appear to experience difficulties when staff have poor English skills, with stressful episodes being more difficult where there is an additional language barrier.

q) Specialist Care needs and communication problems would indicate that head and neck patients’ care may require unit staffing to include at least one head and neck specialist nurse practitioner, with wards being organised to allow for all head and neckpatients to share that care even where admissions are for other specialist areas.

r) The role of the specialist nurse practitioner, supported in many cases by at least one head and neck specialist nurse, is seen by patients to be vital to ensuring that ongoing care in both secondary (centre and district) and primary settings is of the standard required by the National Support Federation guidelines. Patients envisage an enlarged training role for these specialist staff members.

s) Patients continue to face difficulties with direct ambulance admission. Many feel that patient’s medical conditions may be compromised long term by these difficulties.
Patients and their carers feel that new protocols should be in place to ensure assessment by head and neck clinicians as soon as possible after arrival at Accident and Emergency departments.

Whilst we recognise that for many units this may prove difficult in practice, we do feel that paramedic units attending emergency calls are sufficiently skilled to ensure appropriate screening of patients.

Useful Addresses

Macmillan Cancer Relief  
89 Albert Embankment  
London SE1 7UQ  
Tel 0207-840-7840  
information_line@macmillan.org.uk

Marie Curie Cancer Care  
28 Belgrave Sq.  
London SW1X 8QG  
Tel 0207-235-3325  
www.mariecurie.org.uk

Cancer BACUP  
3 Bath Place  
Rivington St.  
London EC2A 3JR  
Tel. 0207 696 9003  
www.cancerbacup.org.uk

NALC (National Association of Laryngectomee Clubs)  
6 Ground Floor  
Rickett St.  
Fulham  
London  
Tel:0207 381 9993  
Fax: 0207 381 0025  
www.communicationsforum.org.uk/organisations/nalc.shtm
Section 4  Chapter 5  Supportive and Palliative Care

Definitions

Although head and neck tumours can be curable if caught early, many patients present with advanced malignancies. Supportive and palliative care becomes appropriate in these patients and also in patients who have failed treatment to their head and neck tumours.

Supportive and palliative care is the care of patients with life-threatening illness, their partners and carers. Its focus is the quality of their lives with the aim of maintaining comfort and dignity from diagnosis, through treatment to the end of the patient’s life, and of the carer’s comfort and dignity throughout this time and into bereavement.

Effective supportive care, backed by palliative care, has become a key feature of the Calman-Hine commissioning of cancer services.

- **Supportive care**
  = good quality communication and symptom control that is the right of every person and the duty of every professional

- **Palliative interventions**
  = treatments used to palliate symptoms (eg. surgery, radiotherapy)

- **Palliative care**
  = care delivered by clinicians with specialist training as part of an interdisciplinary care for patients with severe or complex problems.

**Supportive and palliative care start as soon as they are needed.**

**Supportive care**

All care professionals should offer effective supportive care, but palliative care help will be needed for some patients, partners and relatives. The nurses, family, surgeons and oncologists will give most of the supportive care.

**Palliative interventions**

Palliative interventions are not palliative care. Surgery, radiotherapy and chemotherapy all have the potential for palliation. A neck dissection to control nodes in the neck and stop them fungating is an acceptable palliative procedure. In contrast, major resections when cure is not possible can cause great morbidity and distress at the end of life, resulting in a poor quality of life. Palliative radiotherapy

**Palliative Care**

Some patients have severe or complex physical or psychological symptoms, and some partners and relatives and difficult psychosocial problems. These are likely to need help from palliative care which has the following key elements:

- Provided by accredited clinicians
- Life-threatening illness
- Interdisciplinary working (medical, nursing, physio, OT, social worker)
- Rehabilitation (readaptation)
- Evaluation, research, education

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Palliative care services are available in hospital, hospice and community

- specialist medical advice (all settings)
- specialist nursing support (all settings)
- day hospice
- home care
- inpatient care
- telephone advice
- education and research
- Partnerships in care

Establishing principles of palliative care in Head and Neck Cancer

The effects of cancer invade patients, partners and carers alike. Although some will grow from the experience, it remains a distressing experience for all. For the patient, everyday activities become a source of distress and irritation, while the partners and family feel shut out by the patient and unprepared for the ordeal.

The sense of isolation in patients and carers is reflected in professionals who, faced with multiple physical, psychological, spiritual and social issues can find it difficult to find logical and appropriate treatments with consequent distress to themselves.

Although such distress can seem insurmountable, supportive and palliative care is well established as an effective means of managing this distress. It cannot remove all distress, but provides a path through the distress and confusion, helping achieve a worthwhile quality of life for patient, family, partner and professional alike.

Information sources and learning materials


For clinical information and advice contact your local specialist palliative care service.

For information on palliative care services contact St. Christopher’s Hospice Information Service on www.hospiceinformation.info
Symptom management key clinical decisions

**Checklist ‘CueCards’**
These summarise the key clinical decisions required in a range of situations. More details on management details are to be found in the information sources mentioned previously.
Effective palliative care is the right of every patient and partner, and the duty of every professional.

Ensure adequate team skills, knowledge, attitudes and communication.

Create a ‘safe place to suffer’

Establish a partnership with the patient, the partner and family.

Do not wait for a patient to complain- ask and observe.

Accurately diagnose the cause of the problem.

Clinical estimates of symptom severity are highly subjective and are a poor basis for choosing a treatment.

Do not delay starting treatment.

Administer drugs regularly in doses titrated to each individual, that ensure the symptom does not return.

Set realistic goals.

Re-assess repeatedly and regularly.

Treat concurrent symptoms.

Empathy, understanding, diversion and elevation of mood are essential adjuncts.

These Cue Cards are designed to help you follow these principles in a logical way, while adapting your approach to the individual person.
Resources and Reading

Local Palliative Care Team
Contact your local hospice, or community/hospital palliative care team for advice and help.

Introductory text
Supportive and Palliative Care: an Introduction

Guides
PCF2: Palliative Care Formulary, 2nd ed.
Symptom Management in Advanced Cancer, 3rd edition.

Learning materials
CLiP (Current Learning in Palliative Care) 15 minute worksheets.

Reference text

Further Resources

Palliative care internet/web sites

www.dundee.ac.uk/MedEd/welcome.htm
Dundee palliative care program

www.hospiceinformation.info
Information on palliative care services in the UK and overseas.

www.jr2.ox.ac.uk/Cochrane
PaPaS (Pain, Palliative Care and Supportive Care): special interest group of the Cochrane Collaboration

www.mailbase.org.uk/lists/palliative-medicine
Mailbase: a site for discussion groups and now containing a palliative medicine mailbase

www.palliativedrugs.com
Palliative Drugs: advice on drugs relevant to palliative care.

www.palliative-medicine.org
Association for Palliative Medicine
1: Notes on breaking difficult news

Difficult news: You cannot assume that news is good or bad until you have spoken to the person to find out what they know and whether they want to know more.

Setting: Corridors are not appropriate!

Understanding: Check that problems like deafness or severe anxiety will not prevent understanding.

What do they know? Many people have already guessed the seriousness of the situation.

Knowing more: Most people are clear whether or not they want to know more. If a person does not want to know or is unsure, acknowledge this and offer to answer their questions if they ask again in the future.

Warn – Pause – Check: If they want to know more
Warn eg. "The results were more serious than we hoped"
Pause to see response and give time for the person to respond.
Check if person wants more information.
Repeat as needed.

Denial is a good coping mechanism for many people. It is unusual for it to be difficult to obtain informed consent for treatment.

More help: Ask for help if a person is very distressed, strongly colluding, or if they are so unrealistic that they are having difficulty accepting treatment.


See over for notes
2: Handling difficult questions

- Have you done these first?
  - accepted the setting may not be ideal
  - acknowledged the importance of the question
  - checked why the question is being asked

- Is the person reluctant to pursue the question?
  - have they misunderstood?
  - were you unprepared?
  - were you unwilling to listen?
  - are you uncomfortable with the question?
  - does the person want to stop the interview?

- Is a clear answer difficult?

- Is the answer difficult news?

NB. Difficult questions have to be answered, but clear answers are not always possible- being honest about uncertainty is acceptable!

See over for notes
3: Notes on diagnosing pain in advanced disease

- **Breakthrough pain**
  This is pain occurring despite regular analgesia.

- **Pain related to movement**
  - Fracture: movement of the affected part by the examiner will usually result in severe pain on the slightest movement.
  - Bone metastases or infection: best picked up on bone scan (myeloma and renal carcinoma may show better up on X-rays).
  - Muscle pain: Myofascial pain: has a tender spot when pressed with band of muscle in spasm beneath (= trigger point). Skeletal muscle strain usually occurs suddenly during exertion.
  - Severe soft tissue inflammation: overlying skin is red and swollen (deep infections or in AIDS may have few signs).

- **Other causes**
  Structures that are inflamed, infected or distended.

- **Pain present at rest**
  - Pleuritic pain: due to local inflammation. Check for a rub.
  - Periodic pain (colic): regular episodes of pain lasting a few minutes. Bowel the commonest, followed by bladder and ureter.
  - Related to eating: oral, pharyngeal, gastric or duodenal problems.
  - Unpleasant sensory changes at rest: neuropathic pain (eg. neuralgia)
  - Supplied by a peripheral nerve: nerve compression

- **Pain persisting**
  - Is there unresolved fear, anger or depression?
  - Is the compliance poor?
  - Is this a new pain?
  - Is the analgesic inappropriate or incorrectly administered?

See over for notes

4: Pain treatments in advanced disease (cont)

**Pain during a procedure**
Change technique. Consider: 4-hourly dose of usual analgesic or sedation with midazolam (1-5mg titrated IV or 2.5 - 5mg SC).

**Pain related to eating**
Mucosal pain in mouth or oesophagus: treat infection. Consider benzydamine (Difflam) mouthwash or benzocaine lozenge. Gastritis: ranitidine or omeprazole.

**Skin ulcer pain**
Use non-adherent, silicone-dressings (eg. Mepitel).

**Neuropathic pain**
Amitriptyline 10mg at night, titrated up to 50mg if tolerated. If no better, add gabapentin 100mg 8-hourly, titrated daily to control pain (up to 900mg 8-hourly).

**Nerve compression pain**
Start and titrate a strong opioid. Exclude skeletal instability (eg. vertebral collapse). Treat bone metastases. Consider dexamethasone 8mg daily.

**Persistent pain**
A complete reassessment is necessary.

**NB. Contact pain or palliative care specialist if pain persists.**

5: Notes on using opioids

The WHO analgesic ladder
This stepped approach only applies to choosing an analgesic for opioid responsive pain. Weak opioids are a useful step if strong opioids are difficult to obtain, or patients are too fearful of strong opioids. These steps should be used in conjunction with co-analgesics such as NSAIDs.

Choosing the right opioid
In renal failure the active metabolites of morphine accumulate- this is less of a problem with hydromorphone and does not seem to be a problem with fentanyl.

Doses
There are no fixed dose ranges for strong opioids, but most patients are managed on 30 - 600mg oral morphine / 24 hours. Elderly patients should start on low doses eg. some people over 75 years will not tolerate a morphine starting dose over 20mg daily.

Titrating strong opioids
The aim is to prevent the pain returning by prescribing the opioid regularly and titrating the dose until the patient is comfortable throughout the dosing interval. Opioids are normally increased by 50% every other day. They can be increased faster in severe pain, but at the expense of adverse effects such as drowsiness.

Breakthrough doses
‘As required’ or ‘PRN’ prescribing for pain control must be used with regular prescribing, never on its own (‘PRN’ = Pain Relief Nil). For most opioids, using the 4-hourly equivalent is a suitable dose. For breakthrough pain whilst on fentanyl use dextromoramide, hydromorphone, morphine or diamorphine in equivalent of 4-hourly doses (see Conversion Card).

### 6: Notes on changing opioids

- Calculate the 24 hour dose of opioid
- Find the starting opioid and route in the columns at the TOP
- Find the opioid and route you are changing to on the RIGHT
- Where the column and line crosses, read the conversion

- **x** multiply by this factor
- **÷** divide by this factor
- **nr** this conversion is not recommended

### Rules of opioid conversions:
Conversion factors are only approximations so:
1. Know your opioid
2. Use a conversion factor with which you are familiar.
3. Be prepared to re-titrate the dose.

More potent opioids or routes DO NOT provide greater efficacy. eg. a pain that is not responsive to titrated oral morphine, will not respond to injectable diamorphine either, even though this route and drug are 3 times as potent.

Conversions from weak opioids to potent opioids (and vice versa) are not recommended eg. codeine to fentanyl.

For fentanyl: check manufacturer’s conversion tables
(quick conversion: oral morphine in mg/24hrs ÷ 3 
≈ SC or TD fentanyl in microg/hr)

### If in doubt, contact pain or palliative care specialist


### Example
Oral morphine to diamorphine infusion: conversion factor is [÷ 3]
So, 60mg/24 hrs oral morphine = 20mg /24 hrs SC diamorphine

(see over for more information)
7: Managing opioid adverse effects

**Constipation:** Regular laxative (eg. senna + docusate)

**Nausea and vomiting:**
- If gastric stasis (large volume vomiting):
  - Metoclopramide or domperidone
- In other cases:
  - Haloperidol SC or PO 1.5-3mg once at night for 2 weeks.

**Dry mouth:** Local measures or pilocarpine PO 5mg 8-hrly

**Sedation:** Usually mild and self-limiting (2-5 days). If troublesome, exclude other causes (drugs, hypercalcaemia). If due to opioid consider hydromorphone or fentanyl.

**Fear of opioid:** Information (psychological dependence, respiratory depression and tolerance are rare, the ‘double-effect’ is not seen with good palliative care).

**Confusion / nightmares:**
- If due to sedation: treat as above.
- If persistent or troublesome: reduce dose, or change to hydromorphone or fentanyl.

**Hallucination:** reduce dose, or change opioid.

**Urinary retention:** Rare. Reduce dose, or change opioid.

**Myoclonus:** Reduce dose, or change opioid.

See over for notes

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**Constipation** occurs in nearly all patients on morphine and diamorphine. Fentanyl may be less constipating.

**Nausea and vomiting:** Gastric stasis causes large volume vomiting with brief nausea, and may be accompanied by fullness, heartburn, early satiation and hiccups. Other cases are caused by stimulation of the area postrema and respond to the dopamine antagonist haloperidol (in low doses it is an effective antiemetic for chemical causes of nausea and vomiting with few adverse effects).

**Sedation:** tolerance develops within a few days. Persistent sedation usually has other causes. In patients on morphine or diamorphine, check the renal function, since their active metabolites are renally excreted.

Opioids are poor sedatives: tolerance develops rapidly and there is a risk of increased agitation and myoclonus with inappropriately increased doses.

**Fear of opioid:** addiction is rare because psychological dependence and tolerance to analgesia are rare. The belief in the ‘double effect’ is due to ignorance of analgesic use- it is not an issue with good palliative care.

**Confusion:** confusion in advanced disease is often due to infection, other drugs or biochemical disturbances (eg. hypercalcaemia). If the opioid is the cause, it is usually due to sedation and this will wear off in a few days. Persistent and troublesome confusion due to opioids requires an opioid or dose change.

**Myoclonus:** a sign of opioid toxicity (along with pin-point pupils, drowsiness, confusion).

**Respiratory depression:** this is rare with chronic opioid usage.

8: Reduced hydration and feeding

- **Is hydration or feeding necessary?**
  - is deterioration day-to-day?
  - does the patient want to continue hydration and feeding?
  - does the partner, family and staff agree with patient?

- **Exclude the following as causes:**
  - anxiety or depression
  - swallowing problems
  - weakness or disability preventing intake
  - constipation
  - nausea and vomiting
  - infection or odour
  - appetite suppressant drugs
  - drugs causing nausea, mucosal irritation or gastric stasis.
  - poor food presentation

- **Can anorexia be helped by corticosteroids?**

- **Can thirst be helped by non-oral routes?**

See over for notes

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**Necessity of hydration and feeding:**
Check the patient’s wishes

Usually unnecessary in coma or semi-coma due to advanced disease in the context of day-to-day deterioration. No distress results from this approach if good nursing care continues.

Hydration is usually necessary in most other situations.

Feeding is necessary for comfort, pleasure or if further treatment is awaited.

If the partner, family or staff disagree with patient arrange meeting with all parties (including patient). If necessary ask help from palliative care team.

Discussions should be around the need for hydration and feeding as comfort and health measures, not as life-prolonging measures.

**Exclude causes of reduced hydration and feeding:** a wide range of causes can reduce intake. Check these before prescribing appetite stimulants.

**Corticosteroids:** useful if brief (1-4 week) effect is wanted. Dexamethasone 2 – 4mg once in the morning, has the fewest adverse effects and is convenient.

**Non-oral routes:** as an alternative to IV consider:
For hydration: SC infusions in amounts of 1 – 3 l/24 hours.
For nutrition a gastrostomy (endoscopic or under X-ray control) are the best long-term solutions. NG routes are not well tolerated for more than 1-2 weeks (even with fine bore tubes) and may increase the risk of aspiration.

9: Nausea and vomiting

- Is patient mainly vomiting?
  - Is this regurgitation?
  - Is there reduced gastric emptying which would respond to a prokinetic agent?
  - Is there raised intracranial pressure?

- Is there a drug or toxic cause that would respond to very low dose haloperidol?

- Could there be vagal stimulation that would respond to cyclizine?

- Is this bowel obstruction?

- Is the nausea and vomiting persisting?
  - Is fear or anxiety present?
  - Is gastritis present?
  - Would low dose levomepromazine be helpful?
  - Is specialist help needed?

NB. Treatments used in chemotherapy emesis are often ineffective in palliative care emesis.

See over for notes
### 10: Confusion

- Is memory failure present that would suggest a dementia?
- Has alertness changed due to an acute confusional state?
- Is concentration impaired due to anxiety, pain or depression?
- Is patient misinterpreting external events due to altered alertness?
- Is patient hallucinating due to a toxic state or psychiatric illness?
- Has behaviour altered?
- Is control of disturbance urgent?
  - Are abnormal experiences present that would respond to haloperidol or levomepromazine?
  - Is fear the main feature that would respond to lorazepam or midazolam?

**NB. Add new drugs only if there is no other option**

See over for notes

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### Memory failure:

Dementia: suggested by little fluctuation in confusion, no change in alertness and history lasting months.

Cerebral tumour: uncommon cause, which may respond to dexamethasone and cranial irradiation.

### Change in alertness:

Typical of acute confusional states (alertness may be increased or decreased). Exclude/treat: infection (chest, urine), drugs (started or stopped), chemical withdrawal, biochemical causes (hypercalcaemia, uraemia), cardiac disease, hypoxia, trauma (long bone fracture, subdural).

### Impaired concentration:

Consider anxiety state, depression, pain.

### Misinterpretations:

Common in causes of altered alertness.

### Hallucinations:

Consider drugs, chemical withdrawal, and psychiatric illness.

### Altered behaviour:

Eg. paranoid, euphoria, dysphoria

Exclude: causes of memory failure, drugs (especially corticosteroids) and psychiatric illness.

### Urgent control of disturbance:

See CueCard 15 on Severe Agitation.

11: Notes on the withdrawn patient

**Usual behaviour:** some people are normally introverted or quiet. Offer time to establish trust.

**Refusing help:** This is their right. Acknowledge the refusal and offer willingness to help in the future.

**Confusional state:** manage as in CueCard 10.

**Organic causes:** consider Parkinson’s, drugs causing Parkinson-like symptoms, or severe fatigue due to the illness. If this is the only cause these patients will not express a low mood.

**Fears, guilt or shame:** clarify their concerns and check if these feelings a realistic. Ask for specialist help if necessary.

**Clinical depression:** indicated by
- persistent low mood for > 2 weeks, for > 50% of time
- unlike their usual mood
- difficult to distract them out of mood
- 4 other depressive symptoms (eg. early morning wakening, diurnal variation, loss of enjoyment, undue guilt, hopelessness.)

Start lofepramine 70mg at night, increasing to 70mg 12 hrly or if necessary 70mg mane, 140mg nocte. Alternatively consider venlafaxine or citalopram.

Responses can occur within 2 weeks.

**Persisting low mood:** ask for specialist help. Referral for cognitive behavioural therapy reduces relapses.

12: The anxious person

- Is this a drug-induced dyskinesia mimicking anxiety?
  
  Acknowledge the anxiety
  ('You seem to be worried.')

- Would simple supportive measures help?

- Is this an anxiety state?

- Is the person functioning poorly?
  - Is sedation needed?

- Are panics or phobias a feature?

- Is there an underlying depression?

- Is specialist help needed?

NB. Anxiety and depression commonly co-exist

See over for notes

Drugs: drug-induced dyskinesia causing restlessness can seem like anxiety, but if this is the only cause, patients are clear that anxiety is not the main problem.

Simple measures: empathy, offering information if wanted, exploring causes of concern, reviewing procedures causing anxiety.

Anxiety state: indicated by
  - persistent apprehension for > 2 weeks for > 50% of time
  - unlike their usual mood
  - 4 other anxiety related symptoms (eg. rumination, irritability, insomnia, tremor, tachycardia, sweating)

Poor functioning:
  Mild disorganisation (able to care for self):
  - during day lorazepam 0.5mg – 1mg 6-hourly
  - at bedtime temazepam 10-30mg
  Severe disorganisation (unable to care for self):
  - haloperidol 5-20mg at night.
  - for more sedation change to levomepromazine (Nozinan) 10-50mg at night.

Panics and phobias: consider clomipramine 25mg at bedtime
  (10mg if > 70 years)

Persistent anxiety:
  - consider depression (the two often co-exist). See CueCard 11.
  - ask for specialist help. The response to cognitive behavioural therapy can be rapid (six sessions or less).

13: The angry person

- Do you know how anger affects you?
  Acknowledge the anger
  (‘I can see you’re angry, how can I help?)

- Is the person controlled and contained?

- Is the anger correctly directed?

- Is the anger escalating?
  - can the person contain their anger? (If not, leave the room)

- Is the person depressed?

- Is their anger causing isolation?

- Is the anger persisting
  - is this normal behaviour?
  - is specialist help needed?

NB. If you acknowledge the anger and are clear in your willingness to help, anger should start to defuse in the first few minutes

See over for notes

13: Notes on the angry person

Anger and you: when facing an angry person
- if you get angry: you need to be more restrained
- if you tend to withdraw: you need to be more assertive

Patient is controlled and contained:
- if you’re the only angry person, you need to withdraw.
- some patients express passive anger: acknowledge the anger, discuss the cause and encourage its expression.

Appropriateness of anger:
- misdirected: check this and explore the causes.
- correctly directed: show understanding without being defensive. Apologise if it is your fault, but do not apologise for others.

Escalating anger: if anger is not defusing or is worsening
- position yourself near exit door
- set limits (‘I can only continue if you can control your anger’)
If person cannot accept limits = pathological anger
Stop interview and leave room immediately.

Depression: anger can be a feature. See CueCard 11.

Isolation: acknowledge anger and explore the effects on relationships.

Persisting anger: this may be normal behaviour or due to causes unconnected to illness. Consider specialist help.

From: A Clinical Decision Guide to Symptom Relief in Palliative Care.
Radcliffe Medical Press, 2002
The most immediate goal is to reduce pain at rest and to allow the patient to settle sufficiently to allow adequate assessment.

Agitation that is driven by fear and pain will ease with titrated midazolam—some patients will require no more than 2.5mg (eg. ill patients or those who have slept little in the previous 24 hours), others will require more than 10mg (eg. younger patients or those previously on benzodiazepines).

Fractures: simple treatments for some pathological fractures are possible eg. splinting for a humeral neck fracture, or intercostal nerve block for a rib fracture. An intercostal block of bupivacaine and methylprednisolone (as DepoMedrone) gives analgesia for as long as radiotherapy. Other fractures (eg. vertebra, femoral shaft) may need referral for local nerve blocks or spinal analgesia.

In situations when a procedure is to be delayed (eg. in the night), or where a patient is deteriorating rapidly, titrated sedation is appropriate.

The advice of palliative care, pain and oncology colleagues can be invaluable.

Psychological issues: although the agitation may settle, low mood, anxiety and exhaustion may persist. This persistence of psychological problems will delay the resolution of the pain for several weeks and to avoid disappointment this needs to be understood by patient, partner and staff.

15: Notes on managing severe agitation

- Is control of the agitation urgent?
  eg. irreversible haemorrhage, prevention of injury

- Is hypoxia present?

- Would simple measures help?

- Is a confusional state present?
  - have the clinical decisions for confusion been considered?

- Is control of disturbance necessary?
  - are abnormal experiences present that would respond to haloperidol or levomepromazine?
  - is fear the main feature that would respond to lorazepam or midazolam?

NB. Do not use opioids to control agitation- they can make agitation worse

See over for notes

Urgent control:
  eg. irreversible haemorrhage, prevention of injury
  - midazolam 2-10mg titrated IV, or 5mg IM
    (repeated until settled)
  - ensure environment is safe
  - do not leave patient unattended

Do not use opioids to treat agitation
(They can make agitation worse)

Hypoxia: can cause or worsen agitation. Give 100% through facemask
(NB. if there is a history of pulmonary disease with CO2 retention give no more than 24%).

Simple measures: try gentle explanation of the situation. Many confused patient’s fear about what is happening can be eased by explaining what is happening.

Confusional state: see CueCard 10.

Control of disturbance:
Ensure well lit, quiet, constant environment.
In absence of abnormal experience or behaviour:
  - minimal sedation use lorazepam 0.5mg PO or SL
  - for sedation use midazolam 2-10mg SC, IM or PR (or 20 – 120mg SC infusion per 24 hours)
In presence of abnormal experience or behaviour:
  - minimal sedation use haloperidol 2.5 – 10mg once at night PO or SC
  - for sedation use thioridazine 25 – 50mg PO at night or levomepromazine 25 – 100mg at night PO or SC
    (or 50 – 200mg SC infusion per 24 hours)

16: Unexpected deterioration - deciding about treatment

- Are drugs the cause?

- Is the treatment indicated for comfort only?
  - Is sedation required?
  - Is analgesia required?

- Is a delay in treatment likely?

- Is the need for treatment uncertain?
  - Would it help to consult with partner or family?
  - Would it help to consult with the care team?
  - Would it help to wait a few hours or days before deciding?

NB. Clarity about the cause of deterioration is important - do not assume it is irreversible

See over for notes

Drugs as the cause: reduce dose, but if ventilation has been seriously compromised (<5 resps/min) do the following:
Opioid: dilute 400microg naloxone in 10mls 0.9% saline and titrate IV at 2ml/min until improved, followed by naloxone infusion (do not fully reverse all the opioid or severe pain and agitation will result).
Benzodiazepine: flumazenil IV 200 microg over 15 seconds followed by 100 microg/min up to 1000 microg (1mg)

Comfort only: eg. rapid deterioration with irreversible cause, very short prognosis (hour by hour deterioration), or patient refusing treatment.
  - Sedation if agitated. See CueCard 15
  - Analgesia if in severe pain. See CueCard 14
Support patient, partner, family and staff (including you!).

Delay in treatment: if distressed give sedation. See CueCard 15.

Uncertainty about treatment:
Consult with the partner or family about patient’s previous wishes. Consult with care team about history, rate of deterioration, availability of treatment and previous quality of life.
If need for treatment is still uncertain use rule of 3:
  - Hour by hour deterioration, review in 3 hours
  - Day by day deterioration, review in 3 days.
If further deterioration has occurred, treat for comfort only.
If no further deterioration, or improvement, consider treating.

17. ‘Do Not Attempt Resuscitation’ (DNAR)

1. Decide if it is possible to offer CPR: CPR is not an option in a terminally ill patient where CPR is not expected to succeed. A DNAR decision needs to be documented, but permission from partners or relatives is not needed for a treatment that cannot succeed.

2. Consider the consequences of discussion about CPR with the patient and family: if the patient’s intent is clear and they are competent for this decision, document their decision in the notes. If the patient agrees, explain the decision with partner, family and multidisciplinary staff (including advocate and keyworker). If patient’s intent is unclear for this decision (ie. they are not competent or unwilling to discuss matter), talk to family and staff to develop clear consensus on the DNAR order.

3. Document the DNAR order in the notes.

4. Assume that any patient without a DNAR order is for resuscitation and act accordingly in the presence of collapse or a major, life-threatening bleed.

5. Reassess the wish for resuscitation regularly
While this does not mean burdening the patient and family with a difficult decision every day, it does require staff to be sensitive in picking up any change of views during discussions with the patient, partner or family. The frequency will depend on the clinical situation, eg. week-by-week deterioration needs a weekly review.

6. Any change in decision needs a new DNAR form

See over for notes

17: Notes on deciding a ‘DNR’ order

Uncertainty can result in distressing attempts to resuscitate someone who has collapsed, or inappropriate withdrawal of treatment.

CPR cannot be offered if it is not expected to succeed.

These are resuscitation measures: Cardiac massage and artificial respiration (CPR). These measures are instituted immediately and in full following an unexpected collapse, and in the absence of a DNAR order.

These are comfort and palliative treatment measures: Analgesia, antibiotics, drugs for symptom control, feeding, hydration, oxygen, seizure/status control, suction, treatment for choking. Comfort and treatment measures are instituted after assessment, consultation with patient and family, and on the basis of clinical need. Competency: competent patients are able to understand the results of their decisions, and are free from depression or undue influence.

Advice points
The patient’s view is paramount, ie. decisions cannot be made without them. The only exceptions are if the patient is not competent, death is an expected and inevitable outcome, or the patient does not want to discuss the matter.

Both offering and withholding resuscitation can have immediate and long-term benefits, ie do not assume one is good and the other is bad.

The consultant responsible for the patient has the final decision, but it is wise to reach a consensus with staff and relatives.

The only exceptions are if the patient is not competent, death is an expected and inevitable outcome, or the patient does not want to discuss the matter.

Both offering and withholding resuscitation can have immediate and long term benefits, ie do not assume one is good and the other is bad.

The consultant responsible for the patient has the final decision, but it is wise to reach a consensus with staff and relatives.

Providing resuscitation can be less distressing than withholding it against the wishes of patient and relatives.

Asking patients and relatives is uncomfortable but is easier if they have the information they want about the situation and they are allowed time to make their decision.

Advice from outside the team is invaluable eg. primary health care team, palliative care team, chaplain, social worker, BMA and UKCC advice lines, local ethics committee.

This section has been updated and modified taking into account the conclusions reached at the 3rd Evidence Based Consensus Conference on Head & Neck Cancer, entitled ‘The Management of the Neck in Squamous Head & Neck Cancer’, held at the Freeman hospital, Newcastle upon Tyne on 24th November 1999 (see www.noto.org), and again following a further literature review in January 2002.

Key points
The available level of evidence for managing nodal disease in the neck is poor with an almost complete lack of good randomised controlled clinical trials.

- The status of the cervical lymph nodes is the single most important prognostic factor.
- When a single nodal metastasis exists at presentation or subsequently develops, the cure rate halves\(^1\).
- Prognosis is affected by the number of metastases, the level in the neck, the tumour load, the presence of extracapsular spread, the presence of perineural and/or vascular invasion, previous treatment by surgery or radiotherapy and resectability\(^2,3\).
- A large number of malignant nodes will measure less than 10 mm in diameter (micrometastases) and extranodal spread will occur in a substantial percentage of smaller nodes, as small as 2mm\(^4\).
- The incidence of nodal metastases depends mainly on the site and the size of the primary tumour. This figure may be as low as 1% for early glottic tumours or as high as 80% for nasopharyngeal carcinomas\(^5\).
- The majority of tumours will metastasise in a predictable manner to certain nodal groups but it should be remembered that certain tumours will fast track to remote sites (i.e. nasopharyngeal cancers to level V, tongue cancers to jugulo-omohyoid nodes) and the pattern of spread will be disrupted by previous surgery or radiotherapy.
- The possibility of bilateral nodal disease should be considered especially when the primary site involves the tongue base, nasopharynx or supraglottic larynx.
- There is no evidence that control of metastatic disease in the neck leads to improved survival and issues of function and quality of life have to be considered in the management.
- Standardised reporting of neck dissection specimens according to the Royal College of Pathologists is essential.

Assessment
Clinical Palpation is generally regarded as inaccurate (sensitivity and specificity 60% – 70%), due to a number of factors including shape of neck and the recognised incidence of micrometastases.

CT scanning has a higher sensitivity in detecting metastatic disease [69-93%] than the 70% sensitivity of physical examination\(^6,7\). Overall the accuracy of CT is slightly better than MRI in the N0 neck.
Ultrasound Guided Fine Needle Aspiration Cytology: US-guided FNAC requires both expertise and experience, and is a useful technique when available. It has a sensitivity of 76% and a specificity of 100% in necks that were clinically negative by palpation alone.

TREATMENT OPTIONS
Treatment of cervical lymph nodes is either elective (in the clinically negative neck) or therapeutic (in the clinically positive neck).

Clinically Negative Neck (N0)
The rationale for elective treatment of the N0 neck is based on the following:
Occult disease in the neck will inevitably develop into clinically manifest disease. Despite close regular follow up, some patients will develop inoperable disease in the neck with a wait and see policy.
Untreated occult disease in the neck may predispose to the development of distant metastases.
- Although a survival advantage from elective treatment of the N0 neck in squamous carcinoma has not so far been demonstrated by controlled trials, some multivariate analyses of retrospective data suggest a significant benefit in N0 tongue tumours.
- Elective neck dissection provides useful histological information on which to base future research as well as prognostic information for the patient. Weiss et al used a decision analysis model to propose that tumours that carry a risk of occult metastases of >20% should undergo elective treatment of the neck. This paper was based on data of variable quality. It is now a widely held view that the neck should be treated in cases with a high probability of micrometastases. The evidence to support this is stronger in oral cavity primaries than in laryngeal or hypopharyngeal primaries.

The arguments against elective treatment of the neck are:
- Where the risk of occult neck disease is low, a large number of patients will undergo unnecessary treatment.
- Such treatment destroys a barrier to cancer spread which may be important if recurrence in the primary site occurs at a later date.
- A close follow-up regime to ensure early detection of metastatic disease will improve salvage rates if a ‘wait and see’ policy is adopted. Adding US-guided FNAC to clinical follow-up may provide an alternative strategy to elective neck dissection.
- There is some morbidity and mortality associated with elective treatment.
- The evidence to support elective neck treatment is retrospective and of variable quality.

Indications for elective treatment of the N0 neck
- High likelihood of occult nodal metastases (over 20%). Most sites and stages of squamous cell carcinoma in the head and neck, specifically excluding lip, early glottic cancers and lower alveolar ridge SCC, qualify for elective treatment because the incidence of occult nodal metastases is over 20%. Whilst this is generally accepted practice, it is not supported by strong evidence and should be the subject of clinical trials.
- The status of the cervical lymph nodes cannot be adequately assessed.
- The patient will be unavailable for regular follow up.
- The neck needs to be entered for surgical access to the primary tumour and/or for microvascular anastomosis.

There is good evidence that elective irradiation of the neck is as effective as elective surgical treatment. The choice of neck treatment should, therefore, be influenced by the mode of treatment for the primary.
**Elective Neck Dissection**

The committee of Head and Neck Surgery and Oncology of the American Academy of Otolaryngology and Head and Neck Surgery has produced a standardised neck dissection terminology (Table 1)15.

<table>
<thead>
<tr>
<th>Table 1 Classification of neck dissection techniques</th>
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<tr>
<td>Radical neck dissection</td>
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<tr>
<td>Modified radical neck dissection</td>
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<tr>
<td>Selective neck dissection</td>
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<tr>
<td>Extended radical neck dissection</td>
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The classical radical neck dissection (RND) has no role to play in elective treatment of the N0 neck. The choice lies between a modified RND or a selective neck dissection (SND). A recent prospective study has suggested that SND is just as effective as MRND type III31. As initially stated, most tumours will metastasise in a predictable manner and it is possible to tailor the dissection accordingly. Sentinel node biopsy has been shown to be feasible in head and neck tumours, and may become an alternative to elective neck dissection, at least for oral cavity tumours32,33. In oral cavity and oropharyngeal tumours a selective neck dissection of levels I to IV should be performed due to the possibility of skip lesions in level IV3,36, although level IV metastases are considered rare by some authors34,35. In laryngeal and hypopharyngeal tumours a selective neck dissection of levels II-IV should be performed. Adjuvant postoperative radiotherapy should certainly be used when nodes are found to be involved and bad prognostic factors are present (>1 positive node, presence of extracapsular spread, perivascular or perineural infiltration) and may be beneficial if any node involvement at all is found37.

**Elective Radiotherapy**

When the primary tumour is treated with radiotherapy, the lymph node regions at the greatest risk of harbouring occult disease are included in the treatment field. Elective irradiation is reported to eliminate 95% of nodal metastases with less morbidity than radical surgery17 and is, therefore, preferred when both sides of the neck are to be treated, such as in nasopharyngeal cancers18.

**Clinically Positive Neck (N1-3)**

Treatment of the positive neck will involve surgery, radiotherapy or a combination of the two.

Single modality treatment may be sufficient for N1 disease (especially where the primary tumour arises from the tonsil), but combined modality treatment (surgery plus post-operative radiotherapy) will generally be indicated for stage N2 and N3 necks38. The previously stated criteria for post-operative radiotherapy will apply. The dose should be varied according to the bulk of the disease.

The classical RND has traditionally been the gold standard for treating clinically node positive necks, but has a significant cosmetic and functional morbidity. Shoulder function is reportedly better following MRND than classical RND (and SND better than either)13. MRND appears to be as oncologically effective as RND, even for N2/N3 disease, when combined with postoperative radiotherapy38. MRND type I is now the standard technique for management of the node positive neck in many centres39. The need for comprehensive (i.e. levels I-V) neck dissection in the node positive neck has been questioned: for instance, it may not be necessary to dissect level I for laryngeal tumours40,41. Level V involvement is uncommon.
However, if a MRND is being performed and there is involvement of non-lymphatic structures (accessory nerve, internal jugular vein or sternomastoid muscle) then conversion to RND is generally advised. Where bad prognostic features are found by the pathologist, postoperative radiotherapy is indicated (as above).19

Management of the Neck in cases of an Occult Primary Tumour
The patient will present with palpable disease in the neck in the absence of a primary site. No more than 5% of patients with head and neck cancer genuinely fall into this category as the primary site can nearly always be identified. Any metastatic lymph node containing SCC in the neck apart from supraclavicular fossa nodes should be considered to be a metastasis from an upper aerodigestive tract primary. Supraclavicular fossa nodes are more likely to arise from outwith the head and neck.

Diagnosis
- Careful upper aerodigestive tract examination is mandatory and will identify primary tumours in 40-50% of patients presenting with a metastatic cervical lymph node and no other symptoms.21
- General Examination. Examination of the breasts, the chest, the abdomen and central nervous system should be performed.
- Fine needle aspiration cytology is mandatory, but this is only immediately useful if positive for carcinoma. If negative repeat attempts should be considered.
- Radiology – Chest X ray should be performed to look for chest metastases and an obvious bronchial primary. A CT or MRI scan of the head and neck should be performed prior to any biopsy in an attempt to identify a primary site. The consensus among head & neck oncologists is that a CT scan of the chest should be carried out in such circumstances. PET scanning may be useful if it is available.
- Endoscopy under general anaesthetic. Panendoscopy should include examination of the oral cavity, pharynx (naso/oro/ and hypo/) the larynx, and upper oesophagus. If there is still no obvious tumour present, biopsies should be taken of the nasopharynx, the ipsilateral tonsil removed and the tongue base biopsied. The primary will be found to be arising from the tonsil or tongue base in up to 33% of such cases. Bilateral tonsillectomy has been advocated as there is a 10% incidence of contralateral node metastasis from an occult tonsillar primary.45
- Where no primary is identified serology for EBV IgA/Early Antigen (and possibly Viral Capsid Antigen) should be taken. If positive it is highly predictive for NPC and multiple biopsies should be taken of the postnasal space.23

Management of the Neck in the Occult Primary
Following this protocol, it is reported that in only 5% will the primary site remain truly occult.24 The evidence supporting the management options is retrospective and of variable quality. However it is apparent that surgical salvage after failed RT is not effective in terms of survival.25 The consensus approach is as follows;

If the FNA is positive for SCC a neck dissection should be performed. If the tumour is staged N1, RT should be given for poor prognostic features, and the neck observed if there are good pathological prognostic features. For N2+ necks, combined modality treatment (surgery and RT) and possibly cisplatin based chemotherapy should be administered.

If the FNA is negative an excision biopsy is performed. A frozen section may be performed and, if positive, a neck dissection (MRND or RND) is carried out. If bad prognostic features are present on histology, postoperative radiotherapy (RT) should be given. When only excision biopsy has been performed for N1/NX disease this is always followed by RT.

For N2+ disease, a neck dissection is performed with adjuvant postoperative RT and possibly cisplatin based chemotherapy. Chemotherapy should be administered within a clinical trial.26
Treatment of the likely Primary Sites
The site of cervical lymph node should be considered when trying to identify the primary tumour. Squamous carcinomas of the head and neck region do have a typical cervical node region to which they tend to metastasise.
Evidence for elective mucosal irradiation (EMI) is divided into papers advocating EMI on the basis of reduced occurrence of primary site disease and those opposing EMI because observation demonstrated only a small percentage of patients developing primaries. Treatment should be individualised bearing in mind the potential for severe morbidity and the fact that many patients may be treated unnecessarily. EMI does not improve survival. Ipsilateral mucosal irradiation is advocated as an alternative with less morbidity.

Recurrence after combined treatment
Recurrence in the neck after surgery and radiotherapy carries a very poor prognosis and is often associated with distant metastases. However, the neck recurrence causes distressing symptoms, such as pain, bleeding, offensive fungation and contracture. In selected cases retreatment by means of excision of as much as possible of the tumour, brachytherapy and replacement of the overlying skin with a distant skin or myocutaneous flap is worth considering as a palliative procedure.

RADIOThERAPY TECHNIQUES AND DOSES

Radical radiotherapy
Radiotherapy should be delivered within an accredited department as described in section 3. Modern treatment will use of megavoltage photons from a linear accelerator (typical energies 4-6 MV).

Adequate irradiation of the neck often requires the use of multiple phases of treatment using photons and electrons of appropriate energy with patients immobilised during the treatment. In early cancer of the oral cavity, oropharynx, hypopharynx and larynx first station nodes are treated in continuity with the primary.

The same principles should be used for selecting the nodes for radiotherapy treatment as are described above for surgery. The probability of microscopic involvement of the other nodal groups rises with increasing T-stage and will require the use of large matched fields to encompass these areas. Commonly this may involve the use of asymmetric fields to ensure accurate junctioning between the anterior lower neck field and those treating the primary site.

The number of fields and energy of photons/electrons used will depend on the exact geometry of the individual patient. This information is best obtained by performing a CT scan in the treatment position. The morbidity of neck irradiation is higher in the patient who has undergone a radical neck dissection. In the future implementation of intensity modulated radiotherapy techniques may be of value in reducing these side effects in the unexplored neck. There is now good evidence that the use of concomitant chemo-radiotherapy may improve progression free survival in head neck cancer and this is being increasingly used although it is associated with increased acute morbidity.

The use of elective nodal irradiation achieves ninety to ninety five percent local control with doses of 50Gy in 25 fractions or equivalent.

The indications for postoperative radiotherapy in the neck are derived from careful pathological examination of the neck dissection specimen. Multiple nodal level involvement and extracapsular spread are definite indications for treatment. An increased risk of recurrence is associated with oral cavity primaries, presence of perineural invasion around the primary site. A randomised trial carried out in advanced head and neck cancer at the MD Anderson found no significant dose-response relationship for total doses ranging from 57.6Gy to 68.4Gy -given in 1.8Gy fractions.
A follow on trial demonstrated the importance of completing post-operative radiotherapy within 11 weeks of surgery in the high risk group. There may be a role for accelerated radiotherapy schedules in order to achieve this. This question is being addressed by the current CHARTWEL trial being run in the UK.

**Palliative treatment**

Incurable nodal disease may be treated with either palliative chemotherapy or radiotherapy. Available chemotherapy agents include cisplatin, 5FU and methotrexate. Palliative radiotherapy should be delivered using simple field arrangements, by lateral parallel pair or a single anterior field. Doses used will be lower than in radical treatment, but up to 45 Gy or occasionally more may be required for useful disease control.

**References:**

CONFIRMATION OF DIAGNOSIS

EUA/endoscopy and biopsy

- Careful physical examination is mandatory and has not been superceded by imaging. Examination should include either fiberoptic or direct examination of pharynx, larynx and postnasal space.
- EUA - direct palpation to adequately assess floor of mouth, tongue and neck.
- Panendoscopy for second primary, in those at high risk.
- Tumour sites and areas of field cancerisation should be carefully documented using standardized tumour maps
- Biopsy report should include degree of differentiation, tumour thickness, vascular and peri-neural invasion

Specific imaging

- Superficial low-volume lesions do not require radiological studies.
- Deep invasion, mandibular invasion and nodal status may be assessed by CT or, in selected cases, by MRI. No single imaging modality exists with adequate sensitivity and specificity to accurately assess mandibular invasion. A variety of techniques including MRI, CT, plain radiography and radioisotope scans may be necessary to accurately assess neoplastic disease in the mandible.
- All imaging methods, including specialist dental films, may not show early mandibular invasion. The patient’s dentition is evaluated by a combination of orthopantography and standard dental views.

CONSULTATIONS

All patients with a diagnosis of head and neck cancer must be seen in a multidisciplinary team meeting to allow adequate discussion of the case and appropriate decision-making.

Team members should include:

- Oncologist specializing in head & neck cancer
- Appropriately trained head & neck surgeons with skills encompassing ablation and reconstruction including free tissue transfer
- Specialist head and neck nurse
- Appropriately trained dental surgeon to assess the status of the dentition and make recommendations in relation to the dentition in respect of radiation therapy. The evaluating clinician should be aware of the treatment portals planned for radiotherapy.
Additionally prosthetic support may be required both perioperatively and in the post operative phase for example obturation of surgical defects. An appropriately equipped and staffed oral & maxillofacial laboratory is required to support this aspect of care

- Speech and language therapists for pre-operative counseling regarding possible post-operative speech and swallowing rehabilitation. To assess nutritional status and need for percutaneous gastrostomies.

- Internal medicine, cardiology, respiratory medicine, or anaesthetics as needed to evaluate co-morbidities that may preclude or increase the risk of general anaesthesia.

- Smoking and alcohol cessation counseling

Data regarding the cancer care spell and follow up should be collected prospectively in an appropriate electronic database. Pooling of data and analysis would provide a powerful tool to advance the care of patients with head & neck cancer.

**Early Oral Cancer**

- T1/smaller T2 - this may be treated effectively by single modality therapy, either surgery including ablative transoral laser treatment or radiotherapy (both external beam megavoltage and interstitial techniques). In contrast, larger T2 lesions (>3 cm) require **combination therapy**. Smaller lesions may be allowed to heal following excision by a primary repair or by secondary infection.

- For anterior tongue, buccal mucosa and floor of mouth surgery is preferred if tumour invades the periosteum.

- Radiation therapy is preferred if the oral commissure is involved and for lesions confined to the soft palate.

- Most gingival/palatal lesions are treated surgically.

- Brachytherapy close to the submandibular salivary gland duct can cause scarring requiring surgical intervention to relieve outflow obstruction. Surgical treatment with an ablative laser may reduce this type of problem.

- There is evidence supporting the use of photodynamic therapy as an alternative for low volume disease with complete remission rates and maintenance of response similar to other treatment modalities. It may have a further role in the management of field change cancerisation.

- Moh's micrographic surgery has also been used for low volume disease of the lips.

- Trans-oral laser resection of the primary disease with the use of an operating microscope is being increasingly advocated in some centres.

- Discontinuous neck dissection is often carried out at a separate operative procedure to manage the neck allowing the margins of the primary resection to be histologically assessed and further treatment to the primary site carried out if appropriate. Survival rates compare favourably with conventional surgery with improved functional results.

**The surgical treatment includes:**

- Frozen section evaluation of margins as needed to ensure adequate resection.

- Tracheostomy at the discretion of the surgeon.
• Dental extractions if necessary.
• Insertion of a feeding tube or gastrostomy (optional).
• Orientation of the primary and neck dissection specimen for the pathologist, by the surgeon.

**RADIOTHERAPY**

• Radiotherapy affects function in different ways to surgery. Radiotherapy may be an appropriate option, especially in the older age group, and in those where anaesthesia is a particular risk. Equivalent survival rates can be achieved compared to excision and repair in T1 and low volume T2 tumours of the dorsum of the tongue, the lateral border of the middle third of the tongue, and the floor of mouth.

• As at other head and neck sites, verrucous tumours are treated like any SCC. Tongue tip tumours should be excised. The above only applies if some or the entire radiation dose is given by brachytherapy. Surgery is preferable if the facilities for brachytherapy are not available, or the patient should be referred to a centre where brachytherapy can be offered.

• The only situation where external beam radiotherapy alone is a valid alternative to surgery is in early lesions of the retro-molar trigone.

**Disadvantages of external beam irradiation:**

• Cannot be used again at or near same site.
• Salvage surgery for radiation failure is associated with low survival and high morbidity.
• Side effects include xerostomia, mucositis and osteo-radionecrosis.
• Patients may require full dental clearance prior to treatment.

**Large volume T2, T3 and T4**

These tumours should be treated with combination surgery/post-operative radiotherapy if the patient is fit enough and there is confidence that the tumour can be completely excised macroscopically and repaired with satisfactory functional results.

**Special Surgical Considerations**

• **Mandible** A marginal/segmental mandibulectomy is carried out where invasion of the bone has occurred. The key to a successful outcome is adequate assessment and accurate resection bearing in mind both over and underestimates from existing imaging modalities.

• The use of smears from the resected mandible margins has a high predictive value in the assessment of tumour status.

• The surgeon must be aware of the patterns of tumour invasion, which differ between dentate and edentulous patients. Previous radiotherapy alters the pattern of invasion. It is difficult for good functional results to be achieved after excision of the largest tumours.

• Dental rehabilitation should be considered for patients with mandibular reconstruction. This may include the use of osseointegrated implants.

• **Reconstruction** Primary closure if possible is the method of choice but significant functional impairment may result from the over zealous use of this technique.
The complex three dimensional and composite nature of many oral defects means that a variety of reconstructive techniques may be indicated. These include:

- Local skin flaps, skin grafts, vascularised free tissue transfer (including composite flaps) with myocutaneous flaps less commonly used due to their higher morbidity.

- Reconstruction of the mandible can be carried out using a suitable plating system (for short lateral defects) or free tissue transfer for more extensive defects (including the anterior mandible). The most appropriate bone flaps for mandibular reconstruction include the fibula and deep circumflex iliac artery free flaps. Consideration should be given to the soft tissue defect when assessing the bone flap best suited to the defect. The composite radial free flap produces excellent soft tissue for mucosal reconstruction but has limitations in bone quantity and volume. There is an associated incidence of fracture of the osteotomised radius that may be reduced with the use of prophylactic plating. The best reconstruction may be achieved by the use of more than one flap but consideration must be given to the patients general condition and the potential greater morbidity associated with this approach.

- There are a number of plating systems available to aid reconstruction of the mandible and the surgeon should be familiar with the indications for their use.

- The selection of a flap to be used for oral reconstruction must consider the type of defect to be encountered, the impact upon function of the reconstruction and the donor site morbidity. For oro-pharyngeal soft tissue defects requiring free tissue transfer the fasciocutaneous radial forearm flap is a versatile, reliable and robust flap. It enables a large volume of pliable thin soft tissue to be harvested with a long pedicle and good-sized vessels for anastamosis. It is suited to a two-team approach to surgery thus minimizing operative time and has a low donor site morbidity. Where more bulk is required the rectus abdominus flap provides an abundant volume of skin and muscle or muscle only for reconstruction. Vessels are again of a good size with minimal donor site morbidity.

**TREATMENT OF THE NECK**

- **N0** - The incidence of occult metastasis is approximately 34% according to most series, therefore expectant management of the N0 neck is not recommended. Necks becoming positive during a “wait and see policy” often do so at high pathological stage with poor salvage rates.

- Elective radiotherapy may be used, but the advantage of surgery for the N0 neck is that the specimen is sent for histopathological examination, providing significant prognostic information and objective assessment of adequacy of resection (elective selective staging neck dissection). This surgical staging of the N0 neck is preferable in T2 and non-infiltrating T3 tumours, high grade T1 lesions and low-grade T2 and T3 lesions.

- Because of lymphatic crossover and retrograde flow, especially with anterior lesions and those that are located at or near the mid-line, consideration should be given to treatment of the neck bilaterally, whether radiotherapeutic or selective neck dissection.

- Primary oral tongue lesions may have a different biological behaviour than other oral cavity sub sites, metastasising more frequently to levels II and III than level I. In addition to levels I to III, level IV is also at significant risk of skip lesions from primary oral tongue tumours. Peppering of nodes at multiple levels may also occur in up to 10% of patients with primary lesions of the tongue. Therefore, a selective neck dissection, including levels I to IV, is the most appropriate type of neck dissection for oral tongue primary tumours.
• **The N+ Neck** With palpable neck node involvement or evidence following imaging of the neck, surgical treatment is required which may be in the form of a modified radical neck dissection, radical neck dissection or extended radical neck dissection. In most instances, post-operative irradiation is indicated.

**CRITERIA FOR POST-OPERATIVE RADIOThERAPY**

**Primary Site**

- Any stage with microscopically positive margins.
- Large T2, all T3 and T4, *irrespective of nodal status*
- Peri-neural or intra-vascular invasion
- Poorly differentiated tumours.

Radiation therapy should begin as soon as possible and not > 6 weeks post surgery. It may include a brachytherapy boost when indicated by pathological findings such as unsatisfactory margins.

**LIP 6-14**

**Upper lip & commissures** Rare, but less well-differentiated, lymphatic metastases earlier and more diffuse. Upper lip may spread to pre-auricular and parotid nodes.

**Lower lip** Common - > 90% of cases.

**Clinical evaluation.**

- Complete history, including history of sun exposure and tobacco usage
- Complete examination of the head and neck. Include examination of the lip and skin of the head and neck, entire oral cavity and oropharynx mucosa, indirect or direct fibreoptic laryngoscopy of the larynx and hypopharynx. Evidence of neural involvement of the lower lip musculature (marginal nerve weakness) or hyperaesthesia of the mental nerve should be documented.

**Biopsy** - presence of peri-neural invasion significantly decreases survival.

**Specific Imaging Studies**

Orthopantomogram - indicated if evidence of neural or bony involvement is present or when appropriate to evaluate the patient's dentition prior to radiation therapy.

**CONSULTATIONS**

- Dermatology - in consideration for dermatological treatment of malignant or pre-malignant lesions or Moh’s surgery for the primary lesion.
- Medicine or anaesthetics - as needed to evaluate existing conditions that may preclude or increase the risk of general anaesthesia.
- Dental assessment - in consideration for pre-radiation extractions, restorations or prophylactic treatment.
DEFINITIVE TREATMENT

Treatment of Primary Tumour

Surgery

Adequate surgical excision is generally the preferred initial treatment modality. Margins should be assessed with frozen section, as needed to ensure adequate resection. Most early lesions may be excised under local anaesthesia +/- intravenous sedation. Adjacent pre-malignant tissue should also be excised.

Superficial lesions of the vermillion may be treated with thermal laser ablation or PDT

Full thickness lip lesion defects can be repaired by:

- < 1/3 of lip - closed primarily with V-lip or W-lip technique.
- >1/3 – 2/3 of lower lip - local flap reconstruction with Abbe Estlander flap, Bernard flap or Karapandizic flap.
- >2/3 of lower lip - usually requires micro-vascular free tissue transfer or pedicled regional flap reconstruction.
- Approximation of the tumour to the mandible without evidence of erosion or invasion may require marginal mandibulectomy. Evidence of bone or neural foramina invasion should be removed with segmental resection. Free tissue transfer offers the best reconstruction of the anterior mandible defect.

Radiation and chemoradiation

T1, T2, N0

Fewer than 6% of T1 and T2 lesions are N+. Radiation therapy is a satisfactory treatment for cancer of the lower lip, particularly for patients where the anticipated functional and cosmetic outcome is unsatisfactory with surgery, as well as for those patients medically unfit to undergo surgery.

Treatment by external beam radiotherapy, brachytherapy or a combination of the two is employed as dictated by the size and location of the tumour. Gingival shielding will reduce mucositis. External beam may be electron or megavoltage (250-300 kV) photon fields.

T3, T4, N+

Generally only indicated as primary treatment modality in patients medically unfit to undergo surgical excision and necessary reconstruction.

In some advanced cases, radiation therapy may be utilised as primary therapy if the cosmetic and functional results from surgery are felt to be inferior to that anticipated with radiation alone. 60-70 Gy should be utilised to treat the primary, in 1.8-2 Gy fractions.

Treatment of the Neck Surgery

N0

- T1 - 5% risk of nodes - no treatment
• T2 - 15-35% chance of nodes - controversial - usual spectrum of opinion ranging from support of almost routine selective neck dissection to total lack of support for any form of elective management. No firm evidence.

• For poorly-differentiated high risk or T3/T4 lesions unilateral or bilateral selective neck dissections levels I – III.

N1
• Ipsilateral modified radical neck dissection encompassing levels I-V with or without contra-lateral supra-omohyoid neck dissection.

• N2, N3 Ipsilateral or bilateral radical or modified radical neck dissection. The spinal accessory nerve should be preserved if not involved or approximated in tumour.

The jugular vein should be preserved on at least one side.

• Radiation
  • May be used in N0 neck patient at high risk for metastases (poorly differentiated or advanced primary) or in patients unable to undergo general anaesthesia. Approximately 50 Gy in 1.8-2.0 Gy fractions should be utilised for N0 neck patients. For N1 patients, a radiation dose of 60-70 Gy should be utilised.

Post-operative radiation
  • Beneficial to patients with close or positive primary resection margins, extensive peri-neural invasion, extra-capsular spread or positive nodes.
  • Radiation is initiated after healing has occurred, preferably not > 6 weeks post-operatively.

Recurrence
  • Best managed with aggressive surgical resection with frozen section control. Elective neck dissection advocated as 25% have occult metastases. High success rate in management of local recurrence, 75-85%.

References

Assessment

Tongue

Lip

Neck

20. Haddadin KJ, Soutar DS, Oliver RJ, Webster MH, Robertson AG, MacDonald DG,.Improved survival for patients with clinically T1/T2 N0 tongue tumours undergoing a prophylactic neck dissection Head & Neck 1999 21: 517-5

Mandible


Radiotherapy and chemotherapy


Photodynamic therapy

Section 5  Chapter 3  Hypopharynx

This chapter has been updated and modified using the work carried out by the faculty of the 5th Evidence based Management Consensus Conference on Head & Neck Cancer in 2001 as well as the conclusions emanating from the discussion. The abstracts and full reference list supporting the meeting is available on the NOTO website (www.noto.org).

EVALUATION

Endoscopy. The tumour site and extent are recorded using staging diagrams and a biopsy taken for histological examination. Oesophagoscopy is currently the best way to eliminate synchronous primary tumours (1,2). Bronchoscopy may be useful to exclude tracheal invasion in those tumours extending to the upper oesophagus. Tumours should be staged according to the UICC TNM Classification of Malignant tumours (2002). It may be appropriate to perform a Percutaneous Endoscopic Gastrostomy (PEG) at this stage.

Chest x-ray is better than bronchoscopy in defining a second primary. Chest CT is preferable. Spiral CT and Magnetic resonance imaging (MRI) are complementary. Cross sectional imaging should be performed in all cases due to the upstaging that these investigations lead to in a significant proportion of cases (3). CT has the benefit of speed and may be the best technique to assess the presence of thyroid cartilage invasion. MRI may offer better soft tissue imaging and may be better at assessing the extent of cartilage invasion. CT including the chest is adequate for most tumours with the addition of MRI in difficult cases (3 - 5).

Pulmonary function testing is occasionally useful prior to partial resections involving the supraglottic larynx.

Thyroid function. This is useful as a baseline. If the thyroid is included in the radiation field there is a high incidence of late thyroid failure (6).

TREATMENT

Treatment planning takes the patient’s physical and mental state and their wishes into consideration and recommendations may be modified according to individual circumstances. There is a consensus that optimal treatment for all except the earliest stage tumours is combined surgery and radiotherapy, although there is little in the way of evidence. The validity of survival as a parameter to compare the effectiveness of primary treatment is hindered by the high incidence of regional and distant metastases. Local control appears to be improved by combined surgery and radiotherapy according to the little comparative data that are available (7-10). This may improve quality of survival but more data on functional outcomes of treatment is needed. The nature of the surgery principally depends on the site of the tumour. Conservation surgical techniques are preferable in early stage disease if feasible. Precise treatment depends on the site of the tumour. Some tumours may be suitable for endoscopic resection (11). The use of different modalities to treat the primary tumour and cervical nodes is appropriate in some circumstances. Resection should be wide enough to provide clear margins because positive margins have a poor prognostic factor. Submucosal spread of tumour is more extensive in piriform sinus carcinoma (c.10mm) than postcricoid carcinoma (c.5mm) (12)

Patients should be counselled to stop smoking because of the adverse effect on response to radiotherapy and survival (13).
The use of neoadjuvant chemotherapy to increase laryngeal preservation has been the subject of investigation (14) but at the present time there is insufficient evidence of improvement in either locoregional control or survival to recommend it outside the setting of a clinical trial (15-17). Evidence based recommendations for treatment are hindered by the lack of high quality evidence and the absence of any evidence on function and quality of life after any intervention. In view of the fact that the survival figures for each modality of treatment are broadly similar the treatment modality that is expected to produce the best quality of life should be used.

**Surgery**

**T1 and T2** tumours are very infrequent. Single modality treatment of the primary tumour by partial pharyngolaryngectomy, radiotherapy or endoscopic resection has been reported but there is insufficient evidence to distinguish between them on the basis of local control or survival (11, 18). There appears to be a worse survival if the piriform sinus apex is involved and if disease is bulky. Radiotherapy appears to be less effective in such circumstances (19).

**T3-4** tumours are likely to be optimally controlled by combining radical surgery with post-operative radiotherapy. The nature of the surgery depends on the site and extent of the tumour. Some advanced tumours may be suitable for more extensive conservation or endoscopic surgery (11) but most require total laryngectomy with partial pharyngectomy or total pharyngolaryngectomy. Studies concerning conservation surgery (near total laryngectomy and supracricoid hemilaryngopharyngectomy) have reported highly selected cases but good functional results in terms of voice function have been described (20, 21).

**Resection and reconstruction**

It is difficult to be dogmatic about individual techniques in the absence of good quality data on functional outcomes. Endoscopic resection is particularly suited to some early posterior wall and pyriform fossa tumours and is likely to offer the best functional results and lowest morbidity (11). Partial pharyngectomy with or without partial laryngectomy may be used for more advanced tumours. Reconstructive options range from none (small posterior wall defects) through primary closure to flaps. Radial forearm, myofascial, myocutaneous and jejunal patch flaps have all been successfully used to reconstruct partial pharyngectomy defects. Reconstruction of total pharyngolaryngectomy by single stage techniques is the accepted standard. Free jejunal transfer is the technique that is best supported by the literature (22,23) but tubed free radial forearm free flaps are an appropriate alternative with less donor site morbidity. More extensive defects involving the oesophagus may require gastric transposition.

**Radiotherapy**

Primary radiotherapy with salvage surgery, although only effective in a small number of patients, is a recognised treatment in the UK and is used in some centres. Primary radiotherapy is an appropriate treatment for small hypopharyngeal tumours (5). It is also indicated in those patients who are medically unfit for surgery and can obviate the need for pharyngo-laryngectomy in some circumstances. The dose given when using primary radical radiotherapy as initial treatment varies from 55 Gy in a short overall treatment time of four weeks, to 70 Gy in seven weeks depending on the site, volume irradiated and the treatment philosophy of different cancer centres. Whilst primary radiotherapy for advanced tumours does not produce as good survival figures as radical surgery, the quality of studies is impaired by selection bias in favour of surgery for fitter patients.

**Postoperative radiotherapy**

Postoperative radiotherapy is indicated for

- T3-T4, N0-N3 tumours
- T1-T2 N0 tumours if histology shows:
  - Microscopically positive margins
  - Vascular invasion
- Perineural invasion
- Extracapsular spread
- If neck dissection has not been performed (pNX)

Radiotherapy should be initiated within 6 weeks. The dose for adjuvant radiotherapy is in the range 45 Gy in 18 fractions to 50 Gy in 20 fractions, well below a radical dose. The philosophy in giving adjuvant radiotherapy does differ from that in giving primary radiotherapy where the dose has to be radical or cure will not be achieved. If there is macroscopic or extensive microscopic disease left in the neck then a radical course of radiotherapy should be given. An adjuvant course should be given if there is no indication that there is any gross disease left but that there may be some microscopic residuum.

**Lymph node metastases**
Approximately 2/3 of patients are N+ at presentation. Occult metastases are found in about 40% of neck dissections for N0 staged disease (24). Pathological studies show spread in the N0 neck occurs to levels II - IV and rarely to levels I or V (25-27). Spread is bilateral in midline or bilateral tumours. Level VI involvement may occur in apical pyriform fossa or post-cricoid tumours (28).

**N0 Neck**
There is very little good randomised control trial or even retrospective evidence on how to manage the node negative neck most effectively. If a patient is having primary radiotherapy then first echelon nodes can be covered with the radiotherapy field. If surgical treatment is used then selective neck dissection of levels II, III, IV as a minimum is recommended, with the inclusion of level VI in those tumours that extend to the postcricoid region or apex of the pyriform fossa. If there is oropharyngeal extension then level I should be included. Bilateral dissection is required for central tumours.

**N1-3 Neck**
Comprehensive neck dissection has been the accepted standard for the N+ staged neck. The pattern of invasion of the nodes will dictate whether radical or modified radical dissection is required. This would usually mean a modified radical (functional) dissection for N1-2 disease. Pathological studies suggest that selective neck dissection of levels II - IV may be adequate for N1 disease (23-27). Although its use is controversial, there is some comparative evidence of its clinical effectiveness (29). The available evidence on functional and quality of life outcomes suggests that selective neck dissection is superior to comprehensive dissection (30, 31). The use of supradose cis-platinum and radiotherapy is being trialled for advanced and inoperable neck disease.

**Recurrent or residual disease**
In general surgery should be used for recurrent disease if it is resectable. Post-operative radiotherapy is used if it has not been given before. Chemotherapy may have a palliative role.

**Rehabilitation**
The help of the speech therapist is invaluable for restoring normal swallowing and speech after partial surgery. Consideration should be given to surgical voice restoration as either a primary or secondary procedure. Low-pressure valves are necessary when free tissue transfer has been used for reconstruction. These patients can have quite marked nutritional problems and the input of a dietician is important. In some circumstances the long term use of gastrostomy feeding may be required. PEG feeding is usually required during radical or adjuvant radiotherapy.

**Palliation**
Approximately one third of patients are incurable at presentation. The specialist palliative care team should be involved as early as possible in the disease process. Pain can be controlled and local protocols should be followed. Percutaneous endoscopic gastrostomies (PEGs) placed early can help maintain nutrition in an acceptable manner.
Palliative radiotherapy can produce tumour shrinkage and hence symptom relief. Dysphagia and dyspnoea due to obstruction and pain due to infiltration may all help. Doses approaching radical doses are often required. However, 40Gy given in 10 fractions three times weekly is also helpful.

Re-treatment with radiotherapy and especially using superficial electrons to recurrent neck disease might be useful. Total cord doses have to be carefully assessed in this situation. Endoscopic tumour debulking with the laser may help relieve obstructive symptoms. In inoperable cervical node recurrence further surgery and brachytherapy are sometimes very effective. For bone metastases standard radiotherapy techniques are used. Palliative radiotherapy is not indicated for asymptomatic distant extension or spread since it will cause iatrogenic symptoms and will not produce cure. The role of chemotherapy as adjuvant or neo-adjuvant treatment is not supported by randomised controlled trials. Toxicity due to palliative chemotherapy is unacceptable. Carboplatin plus or minus 5 fluorouracil is a reasonable palliative regime. Taxanes may have a role to play in fitter patients. In the absence of a response, persistence is not rational.

References:
When considering tumours in this subsite it should be remembered that there are significant differences in management for each of the anatomical areas which may be involved (1).

The principle areas for registration of index tumours are tonsil, base of tongue, soft palate and pharyngeal wall. As with many areas of the head and neck it is frequently difficult to determine the index site of large indeterminate tumours.

The main effects of treatment are on the functions of speech and swallowing and are at their greatest when the soft palate and tongue are involved.

ASSESSMENT OF TUMOUR/CONFIRMATION OF DIAGNOSIS

Although full assessment in the outpatient setting is of value in giving an appreciation of the extent of the tumour it is mandatory to carry out an EUA and biopsy of the tumour. This will:

- determine the histological diagnosis
- enable staging
- assess extent of surgical resection; with respect to extension across the midline of the tongue base, involvement of the mandible, larynx, or prevertebral muscles.
- give indication of type and extent of reconstruction required
- allow assessment of potential airway compromise/requirement for tracheostomy
- exclude synchronous head and neck tumours
- assess/treat dentition

INVESTIGATIONS

These are principally imaging. Assessment of the primary is best achieved by either CT or MR scanning with the latter being the preferred option. The use of imaging allows further assessment of the neck and chest, which is of importance in this group as oropharynx has the highest incidence of distant metastases of all the head and neck tumours (2). If there is further doubt about the neck status ultrasound with FNAC can be performed, an orthopantomogram is frequently required when assessing the dentition.

CONSULTATIONS

- Dietary; most of the tumours involving the tongue base, tonsil, or pharyngeal wall will require dietary support to correct the frequently encountered cachexia and for continuing support during treatment. Although there should be a low threshold for gastrostomy insertion, either by endoscopic or radiological techniques, it carries the potential for additional morbidity (3).

- Speech and Language Therapy; Early referral is required to allow management of both communication and swallowing problems.

- Oral Surgery; Treatment of caries must be complete in view of potential mandibular surgery and/or radiotherapy.

In view of the significant differences between subsites, each will be considered in turn.
TONSIL

Tonsil carcinoma may be found as part of the diagnostic workup of a patient presenting with SCC lymphadenopathy. The tonsil dissection for such a T1 lesion is sufficient as the initial treatment but radiotherapy will be required post operatively.

T1/T2 lesions can be treated by transoral surgery or radical radiotherapy.

T3/T4 lesions require radical resection, frequently some form of clearance of the ipsilateral neck, reconstruction and radical radiotherapy.

To ensure adequate exposure of the tumour ipsilateral paramedian mandibulotomy is generally the most effective and when carried out in the most appropriate site results in little morbidity.

Irrespective of the neck status neck dissection is performed to facilitate the reconstruction of the defect, this will range from a selective neck dissection to a modified radical neck dissection.

Reconstruction is by either pectoralis major myocutaneous flap or free radial flap with little conclusive evidence of a better functional result with one than the other (4). If resection of the mandible is required, for example due to involvement of the retromolar trigone every effort should be made to preserve at least the rim of the mandible as the functional result is superior to that if a segmental resection is required. If it is necessary then a free fibula flap will be required for the defect.

BASE OF TONGUE

Although T1 lesions can be treated by surgery, for example via a transhyoid approach, there has been increasing interest in the use of endoscopic laser techniques. There is little evidence in the relative value of any of these techniques or methods of reconstruction. They can however be treated equally well by radical radiotherapy. T2, T3 and T4 lesions require radical treatment, the principal decision being between the use of brachytherapy in conjunction with bilateral neck dissections or external beam radiotherapy possibly in conjunction with chemotherapy (5-8). There are various chemotherapy regimes which can be used, but in general they include the platinum based drugs, carboplatin or cisplatin, and 5FU (9-13). If there is recurrence, or if the extent of the original tumour is deemed incurable by the above techniques, total glossectomy with laryngectomy may be considered. This has extremely high morbidity, but with careful counselling and extensive rehabilitation it can be performed in an appropriate group of patients.

SOFT PALATE

Tumours in this area tend to appear on the free edge of the soft palate or uvula, are predominantly T1 or T2 at presentation, and can be treated effectively by either endoscopic resection, with or without the CO2 laser, or by radical radiotherapy. If they are larger and radical resection is employed effective, functional reconstruction is a challenge and frequently results in significant morbidity.

POSTERIOR PHARYNGEAL WALL

T1/T2 lesions can be treated by either endoscopic resection or by radical radiotherapy.

If more extensive than this then they will tend to involve the lateral pharyngeal wall and so treatment will be very similar to that for tonsillar tumours. If more advanced then to effect a cure is difficult, however to attempt this or to maximise palliation radical radiotherapy +/- chemotherapy can be used.
Difficulty in swallowing and aspiration occur in most patients, with little evidence in support of any combination in treatments reducing this morbidity.

NECK

As base of tongue, posterior pharyngeal wall, and palatal lesions predominantly affect the midline then spread to lymph nodes on both sides of the neck must be considered when planning treatment. (14-17). In the N0 neck consideration should be given to elective treatment, either by radical radiotherapy or by selective neck dissection if open resection is being employed (18). In T1/T2 lesions levels I-III are cleared, whereas with more extensive lesions level IV is also included. In the last few years there has been increasing interest in the use of sentinel node biopsy. Although it shows promise, either on its own or in combination with imaging such as ultrasound, its use can be regarded as research based at present. (19, 20). The N1 neck is treated by neck clearance either by selective or type I modified radical neck dissection, depending on the treatment of the primary. In early lesions radical radiotherapy may be used. In N2/N3 necks a type I modified radical neck dissection is indicated but may not be possible depending on the site of the disease in N2 and the extent in N3. Post operative radical radiotherapy is indicated if more than one node is involved or if extracapsular spread is present.

RADIOTHERAPY

This may be indicated in the following circumstances:

Postoperatively to the index site following excision of large tumours and to the neck if more than one node is involved.

To the index tumour in the case of T1/T2 lesions with a curative aim or in T3/T4 lesions, with or without chemotherapy, as either with a curative or palliative aim.

The dose and fractionation is determined by local factors, from 50Gy being used electively for the N0 neck, 60/65 Gy for the index site to 68Gy for advanced base of tongue lesions in conjunction with chemotherapy.

References:


CONFIRMATION OF DIAGNOSIS

Clinical and radiological evaluation are necessary for accurate staging. Laryngoscopy or microlaryngoscopy and pharyngoscopy are supplemented with oesophagoscopy and bronchoscopy where necessary. Hopkin’s rods should be used in assessing the subglottis. If available, photo documentation is a valuable record of the lesion for the case record. PET (positron emission tomography) has shown promise in detecting primary and recurrent laryngeal disease (1).

TREATMENT

An accurate anatomical description of the tumour extent is essential to stage the tumour, and to allow the best treatment selection. All patients must be accurately staged clinically and radiologically. It is essential that this information is recorded. Glottic, supraglottic and subglottic tumours differ significantly in patterns of behaviour and are considered separately. Even tumours involving different anatomical sub-sites within the same region may behave differently. These tumour related factors, as well as the medical condition and the needs of the individual patient, all require consideration in formulating a treatment strategy. In general, radiotherapy and conservation surgery alone are options for T1-2, N0 lesions, and combined surgery and radiotherapy is used for advanced (high volume T3 and 4) and N+ disease (2). The UK standard practice for treating T1 and T2 laryngeal cancer is currently radiotherapy.

The sole use of radical radiotherapy, or sole use of surgery for all T stages of laryngeal cancer is not acceptable.

It is no longer acceptable for surgeons to manage patients laryngeal cancer on the basis of only one surgical option (total laryngectomy)

No patient should be subjected to laryngectomy in the absence of modern methods of voice restoration including valved speech.

The surgeon’s repertoire must include conservation techniques, including laser surgery, partial laryngectomy, selective neck dissection, and surgical voice restoration.

Early glottic cancer

The goal of treatment in glottic cancer is to effect a cure and preserve a normal voice without impairing breathing or swallowing. To select the appropriate treatment modality for early glottic cancer is difficult in the face of extensive conflicting literature supporting different approaches (3,4) Early glottic cancer is potentially curable with either surgery or radiotherapy (5) Single modality treatment is recommended. Endoscopic or open resection (partial laryngectomy), or radiotherapy are the treatment options. There is a need for a randomised control trial to evaluate (a) disease control, (b) functional outcomes, (c) treatment morbidity. Tumour distribution and the age and performance status of the patient determine the choice of treatment.

A comparison of outcomes in Canada and the US suggested better larynx preservation in treatment plans consisting of primary radiotherapy with salvage surgery. Survival was the same (6).

Tis: Evidence suggests carcinoma-in-situ can be reversible with cessation of smoking (7) Excision biopsy is probably the treatment of choice although excellent control rates can be achieved with both endolaryngeal microsurgery and radiotherapy.

Excision with preservation of the vocal ligament if possible is probably the best option, if not then an initial biopsy followed by definitive treatment is indicated. Close follow-up where possible by the same treatment team is important (8).

T1a – T1a: A large amount of literature, almost exclusively case series, reports that radiotherapy or endoscopic laser resection offer comparable local control rates for accessible lesions.
Similar local control rates can be achieved with partial laryngectomy (9) although voice results are radiotherapy and laser surgery seem better (10).

Cost analysis shows endolaryngeal laser surgery to be more cost-effective that radiotherapy.

T1b: Treatment options here are the same as T1a (Radiotherapy, Laser surgery and Partial laryngectomy) In the UK radiotherapy remains the overall treatment of choice. Involvement of the anterior commissure in some series correlates with poorer outcome for both radiotherapy and laser therapy. This may however relate to under-staging of the disease at diagnosis (11,12).

For partial laryngectomy satisfactory functional results can be achieved if more than half of the contralateral cord is preserved.

The literature supports the belief that continued smoking and alcohol consumption during and after radiotherapy worsens the outcome in terms of local control (13,14).

T2: Superficial tumours without restricted cord mobility (T2a) can be treated by radiotherapy or surgery. Radiotherapy may be preferable for extensive superficial tumours because of better functional results. Tumours impairing cord movement (T2b) can be treated with radiotherapy or partial laryngectomy (15). One recent paper reports high local control rates (93.8-95.7%) in patients undergoing induction chemotherapy and partial laryngectomy for T2 glottic carcinoma (16) In the absence of randomised controlled trial evidence however the impact of combined versus single modality therapy in early glottic cancer remains unclear.

Advanced glottic cancer

T3: Large variations in tumour volume affect treatment outcome in similarly staged tumours (17). Treatment needs to be individualised. It may be that some T3 tumours are under-staged and are really T4 due to unsuspected cartilage invasion. Treatment options are surgery or radiotherapy or combined therapy. Partial laryngectomy may be suitable for small volume tumours while radiotherapy may be the preferred option for medically unfit patients and can give better voice outcomes.

The role for combination therapy including chemotherapy is unclear at the present time. Overall survival is comparable with all treatment options although laryngeal preservation is obviously higher in those patients undergoing primary non-surgical treatment. Loco-regional control may however be better in the surgically treated patient. One non-randomised study from India reported higher 4 year survival rates in the Surgery and radiotherapy group compared with the Radiotherapy and salvage surgery group (18). A review of prognostic factors in T3 laryngeal cancers concluded that glottic lesions, female patients and N0 disease did better with primary radiotherapy and salvage surgery while patients with pre-operative tracheostomy or N+ disease did better with primary surgery (19).

Salvage surgery generally implies total laryngectomy however some authors now report good results with salvage partial laryngectomy (20,21).

As the role of chemotherapy remains unclear in advanced disease its use should be confined to the setting of a clinical trial.

T4: Primary surgery with post-operative radiotherapy may be the treatment of choice since authors report higher disease-free survival compared with primary radiotherapy (22). Patients who are not fit for or refuse surgery may be candidates for radiotherapy and/or chemotherapy.

A randomised controlled trial of alternating chemo-radiotherapy compared with radiotherapy alone in patients with advanced inoperable head and neck cancer (including laryngeal) reported higher disease-free and overall survival in the combined treatment group (23).

Supraglottic Tumours

Treatment should take into account the high incidence of overt and occult metastatic disease. T staging does not correlate well with tumour volume, suitability for partial laryngectomy, or prognosis. In general early stage tumours are treated with a single modality, advanced tumours with combined surgery and radiotherapy. Surgery may offer better local control for early stage supraglottic cancer (24,25) but not all patients are medically suitable, with the risk of post-operative swallowing problems and aspiration.
**Early Supraglottic Tumours – T1-2**
Conservation surgery (including endolaryngeal resection) or radiotherapy are alternatives. The choice depends on the tumour distribution and performance status. Consideration should be given to bilateral elective neck treatment.

**Advanced Supraglottic Tumours – T3-4**
The only prospective randomised trial for advanced laryngeal cancer suggests that total laryngectomy with post operative radiotherapy confers no survival advantage when compared to induction chemotherapy followed by radiotherapy, and if necessary salvage laryngectomy (26). Other studies have suggested there is no survival advantage for T3 laryngeal cancers in performing laryngectomy with or without follow up radiotherapy, compared to radical radiotherapy with salvage surgery (27,28), but that primary laryngectomy with follow up radiotherapy does confer a significant survival advantage in T4 laryngeal disease compared to radical radiotherapy, with if necessary, salvage surgery (22). This may suggest chemoradiation to be the treatment of choice in low volume T3 laryngeal disease, but that primary surgery with follow up radiotherapy should be considered for high volume T3 supraglottic disease. Primary surgery does have the advantage of allowing access to the neck nodes. Midline cancers must have both sides of the neck treated (29). Patients with supraglottic tumours involving the pre-epiglottic space or upper piriform fossae may be suitable for an extended supraglottic or subtotal laryngectomy rather than total laryngectomy (30). To perform conservation laryngeal surgery the patient must have good health and pulmonary function otherwise total laryngectomy is the surgical option of choice.

**Subglottic Tumours**
Most tumours are indistinguishable from glottic tumours with subglottic extension. Radiotherapy may be suitable for early tumours, but most present at a late stage often with stridor, and treatment with total laryngectomy and post-operative radiotherapy is indicated.

**MANAGEMENT OF THE NECK IN LARYNGEAL CANCER**

**N0**
In early glottic cancer (T1-T2), there should be no need for elective neck treatment as the risk for occult neck metastases is low.

There is no Level I or II evidence that elective treatment of the N0 neck in laryngeal cancer improves survival. Elective treatment of the neck inevitably carries some morbidity. Elective neck irradiation is as effective as elective neck dissection. Some groups adopt an aggressive surgical policy for the N0 neck (31).

However, with advanced glottic cancer (T3-T4), transglottic, all T stages of supraglottic cancer, and subglottic cancer there is at the present time a consensus that the neck needs to be treated electively. Treatment of the primary site will determine how the neck is treated. If the primary site is treated with radiotherapy then elective neck radiation should be performed. If the primary site is treated with surgery then an appropriate elective neck dissection should be performed. Glottic cancer would be treated with an ipsilateral selective neck dissection, and a bilateral selective neck dissection for supraglottic cancer. Levels II, III, IV are at greatest risk. Subglottic extension of glottic cancers and subglottic cancer tend to spread to the paratracheal nodes (level VI) and these should be included in the bilateral neck dissection (32) If the paratracheal nodes are positive then the mediastinum should be included in the post-operative radiotherapy fields. If histopathology demonstrates multiple nodal metastases, or extra capsular spread, post-operative radiotherapy of the neck is indicated.

In Salvage surgery treatment of the N0 neck should always be considered (33).

**N+**
If radiotherapy (with or without chemotherapy) is used to treat the primary tumour both sides of the neck should be included in the irradiation fields.
If post radiotherapy assessment at 6 weeks demonstrates, on clinical examination or CT scan, residual neck disease, then this should be treated with a modified or radical neck dissection.

If the primary tumour is treated with surgery then a modified radical neck dissection(s) is performed with post-operative radiotherapy for multiple positive nodes, or nodes demonstrating extra capsular spread. Histological positive paratracheal nodes would indicate post-operative radiotherapy to the mediastinum.

Bilateral dissection is arguably required for all supraglottic disease, but is essential when the disease crosses the midline.

Special Circumstances
The patient presenting with stridor presents a difficult problem. Most have advanced disease that dictates combined modality treatment. Endoscopic debulking is carried out when possible. This can be with the microdebrider or laser (34) Tracheostomy, although not desirable, may be necessary. Emergency laryngectomy should be used only in exceptional circumstances. The diagnosis being established with frozen section.

Recurrent or Residual Disease
The treatment will depend on whether the patient has been treated with initial irradiation or surgery. Recurrence after previous irradiation is managed with salvage surgery. The surgical approach for recurrent disease is planned according to the site and extent of the original lesion. Total laryngectomy is the most commonly performed salvage surgery, although conservation laryngeal procedures are still appropriate in carefully selected cases (20). Unresectable post-surgical recurrences are treated with radiation, with or without chemotherapy. Stomal recurrence, particularly if arising superiorly may be resectable, requiring mediastinal resection and possible pharyngeal replacement.

CHEMOTHERAPY IN LARYNGEAL CANCER

The role of chemotherapy in the management of laryngeal cancer is evolving through mounting evidence from randomised trials and non-randomised studies. The contribution of chemotherapy must be judged by its effect on laryngeal preservation, disease free and overall survival, and on levels of treatment related morbidity. Studies of chemotherapy on laryngeal preservation have shown that the larynx could be preserved in a quarter to a third of cases without detriment to overall survival. The American VA trial, which has the longest follow-up (median follow-up over 8 years) confirms the larynx can be preserved in up to 2/3 of patients achieving complete or partial response to induction chemotherapy without jeopardising survival (26).

The EORTC trial (of larynx preservation in patients with hypopharyngeal cancers) showed a similar effect (35). An interim report on a French randomised trial (GETTEC) concerning just 68 patients of a planned 300 accrual showed a worse survival outcome for those receiving induction chemotherapy (36). Meta-analysis of all three trials demonstrated there was no differences in overall or disease free survival, but a functional larynx was preserved in two thirds of surviving patients (37). A more recent authoritative meta-analysis (Meta-Analysis of Chemotherapy on Head and Neck Cancer Collaborative Group, MACH-NC) confirms a significance benefit from cisplatin and fluorouracil given concurrently with radiotherapy – absolute 2 year survival benefit=7% (38). Such benefits however must be balanced against the side-effects of the chemotherapy.

Combining chemotherapy concurrently with radiotherapy may be superior to induction or neo-adjuvant chemotherapy in advanced head and neck cancers, and may improve overall survival. Two randomised trials have shown improved local control and survival in over 5 years of follow-up in patients receiving an alternating chemo-radiotherapy regime (23).
The role of chemotherapy in laryngeal cancer continues to evolve. Carefully controlled trials of chemotherapy with other treatment modalities should be supported. Patients treated with chemotherapy in conjunction with other modalities outside of trials should be managed in strictly controlled situations.

- High response rates to chemotherapy are achievable in laryngeal cancers.
- Chemotherapy with radiotherapy may improve larynx preservation rates but remains under investigation.
- The effect of concurrent chemo-radiotherapy may be to increase toxicity but the impact on survival remains unclear.
- The optimal combinations of chemotherapy with differing schedules of radiotherapy have yet to be determined.
- Further studies and trials are required and should be supported.

References:


Additional References:


XVI. Mendenhall WM, Tannehill SP, Hotz MA, Kasler M, Remenar E. Should chemotherapy alone be the initial treatment for glottic squamous cell carcinoma? Eur J Cancer 1999;35:1309-1313
This section has been updated and modified with reference to the ‘Evidence Based Conference of the Management of Salivary Gland Neoplasms’ held at the Freeman Hospital, Newcastle upon Tyne on the 19th November 1998. A full reference list from the meeting is available on the NOTO website – www.noto.org (1)

Salivary gland malignancies are rare and present a diverse range of histology and clinical behaviour. The original 1972 WHO histological classification of salivary gland tumours consisted of four subgroups: Epithelial tumours, Non-epithelial tumours, unclassified tumours and allied conditions of which the epithelial group was the larger in number. This was modified in 1991 (2) such that tumours of similar pathology are now grouped together even though they may have differing prognoses. Under the new system there are seven categories:

- a. Adenomas
- b. Carcinomas
- c. Non Epithelial tumours
- d. Malignant Lymphomas
- e. Secondary tumours
- f. Unclassified tumours
- g. Tumour -like disorders

Carcinomas are often further classified as high grade, low grade or mixed, the latter inferring a variable behaviour depending on the histological picture. Except in the case of mucoepidermoid tumours however, clinicopathological correlation has proved to be unreliable and often the clinical behaviour rather than the histology of a tumour provides a better guide for treatment. Although, overall, tumours are more common in the parotid, the incidence of malignancy is higher in the submandibular, sublingual and minor salivary glands (3). The more common adenomas and carcinomas are shown below:

<table>
<thead>
<tr>
<th>Adenomas</th>
<th>Carcinomas</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Pleomorphic adenoma</td>
<td>• Acinic cell carcinoma</td>
</tr>
<tr>
<td>• Myoepithelial (myoepithelial adenoma)</td>
<td>• Mucoepidermoid carcinoma</td>
</tr>
<tr>
<td>• Basal cell adenoma</td>
<td>• Adenoid cystic carcinoma</td>
</tr>
<tr>
<td>• Warthin’s tumour (adenolymphoma)</td>
<td>• Polymorphous low-grade (terminal duct)</td>
</tr>
<tr>
<td>• Ductal Papilloma</td>
<td>• Papillary cystadenocarcinoma</td>
</tr>
<tr>
<td>• Cystadenoma</td>
<td>• Mucinous adenocarcinoma</td>
</tr>
<tr>
<td></td>
<td>• Adenocarcinoma</td>
</tr>
<tr>
<td></td>
<td>• Carcinoma in pleomorphic adenoma</td>
</tr>
<tr>
<td></td>
<td>• (malignant mixed tumour)</td>
</tr>
<tr>
<td></td>
<td>• Squamous cell carcinoma</td>
</tr>
<tr>
<td></td>
<td>• Undifferentiated carcinoma</td>
</tr>
</tbody>
</table>

CONFIRMATION OF DIAGNOSIS

Malignant tumours may be clinically indistinguishable from benign lesions and definitive histology is usually unavailable until after surgical resection. Diagnosis is therefore based on:

Clinical presentation - pain, rapid growth, fixation, nerve involvement or neck metastasis all suggestive of malignancy.
FNAC - useful for major salivary gland lesions - must be examined by cytopathologist experienced in the diagnosis of salivary gland disease. This should distinguish malignant from benign disease in 90% of cases (4,5).

MRI scanning - non-homogeneity, muscle infiltration or suspicious regional lymph node appearance suggests malignancy.

Open biopsy - this should not be done for major salivary gland lesions because of tumour spillage although it may be indicated for minor salivary gland lesions where the overlying epithelium will be excised at the time of surgery.

Frozen section - accurate diagnosis often difficult and false negative rates significant and therefore should not be relied on. (6)

SUBMANDIBULAR GLAND

TREATMENT POLICY

SURGERY

Primary Tumour
- A total excision is appropriate for most tumours confined to the gland. Some argue in favour of wider resection for adenoid cystic tumours including the lingual, hypoglossal and marginal mandibular nerve if there is any suspicion of involvement by tumour (7), but evidence is not strong, and the advice is counter to recommendations for the uninvolved facial nerve in parotid disease. Clinically high-grade tumours in young individuals should be treated more aggressively with excision of the gland plus a 2cm margin of apparently healthy tissue (8).
- Large Tumours with bone involvement - composite resection of tumour, adjacent soft tissue cuff and segmental mandibulectomy.

Neck Disease
- All Patients - clearance of nodes in suprahyoid region. Clinically high-grade tumours or suspicious MRI appearance should have an elective supra-omohyoid dissection.
- Patients with clinically confirmed neck metastasis should have a radical or modified radical neck dissection (8,9).

RADIOThERAPY

- Inoperable tumours where palliation can be achieved.

Post-operative (7,10)
Indications:
- High grade or advanced stage tumours with a high risk of local recurrence
- Residual neck disease or microscopic extra-capsular spread from lymph nodes
- Surgery for recurrent disease
- Adenoid cystic tumours
- Radiotherapy should be commenced within six weeks of surgery.
PAROTID GLAND

TREATMENT POLICY

SURGERY

Primary Tumour

• Apart from lymphomas, surgery is the treatment of choice. A conservative parotidectomy should be performed with preservation of the facial nerve providing there is no tumour invasion. For deep lobe tumours this will involve a total parotidectomy plus resection of adjacent structures if necessary to achieve an en-bloc resection. Any part of the facial nerve not infiltrated by tumour should be preserved (11,12). Primary nerve grafting should be considered if microscopic clearance of the main trunk and peripheral branches can be achieved.

• Adenoid cystic carcinoma requires a total parotidectomy with sacrifice of any part of the nerve involved by tumour (7).

• Resection of an intact nerve has been shown to improve local disease control but will not improve survival (13). It is unlikely that nerve grafting will compromise outcome and therefore could be considered particularly if combined with post-operative radiotherapy.

Neck

• Neck dissection should be performed in patients with evidence of nodal disease either clinically or on MRI scanning.

• A prophylactic neck dissection should be considered for patients with clinically high grade tumours, i.e. adenocarcinoma, squamous and undifferentiated (14).

RADIOThERAPY (7,12,15)

Post-operative

• As for the submandibular gland plus any patient in whom the facial nerve has been preserved despite close approximation of the tumour.

Recurrent Disease

• Requires careful evaluation of the patient with repeat imaging and a review of the histology from the initial excision. This will usually require more radical surgery with sacrifice of the nerve and overlying skin if any suspicion of involvement by tumour. Super-radical resections of skull base have not to date shown convincing evidence of improved survival. Consider chemotherapy and/or radiotherapy for palliation.

MINOR SALIVARY GLANDS

Confirmation of diagnosis

Clinical history and examination with particular reference to palate and nasal cavity. Open biopsy permissible to confirm diagnosis. The prognosis for these patients is more closely related to stage of disease rather than histology (16).

Treatment

En-bloc resection with depth of excision equal to width to ensure adequate resection margins. Significant defects are repaired as appropriate if necessary.

Neck

Therapeutic neck dissection indicated for lymph node involvement. Elective neck dissection indicated for adenocarcinoma, carcinoma in pleomorphic adenoma and undifferentiated carcinoma.
Radiotherapy Indications: (16)
- Post-operative
- Microscopic residual disease
- Adenoid cystic tumours
- Aggressive undifferentiated tumours

NATURAL HISTORY OF COMMON TUMOURS

Acinic cell carcinoma
- Accounts for about 3% of parotid tumours where it occurs most commonly. Peak incidence 5th decade
- Demonstrates a variable histological pattern and can be multifocal in origin and occasionally bilateral.
- Determinate survival rates of 90% at five years and 55% at 20 years
- Lymph node metastases occur in approximately 10% of cases (11)
- Total parotidectomy with preservation of uninvolved nerves recommended. Elective neck dissection not indicated.

Mucoepidermoid Tumour (11)
- A tumour of variable malignancy with low-grade lesions showing a ‘benign’ nature
- Most common major salivary gland tumour (4%-9%) >90% in the parotid but overall more frequent in minor salivary glands
- Commonest malignant salivary gland tumour in children
- Highest incidence third to fifth decade M=F
- Almost always in the superficial lobe of the parotid
- Histological division into low, intermediate and high-grade shows correlation with prognosis; although so called ‘low-grade’ tumours can on occasion be aggressive. Five year survival varies between 86% for low-grade to 22% for high-grade tumours
- 40% incidence of lymph node metastases in intermediate and high-grade tumours
- Low-grade tumours require local resection with adjuvant radiotherapy indicated for high-grade tumours

Adenoid Cystic Carcinoma (17)
- Most common salivary gland malignancy - mucosal sites more frequent than major salivary gland
- 2-6% of parotid and approximately 15% of submandibular tumours
- Slow, pervasive growth and a high incidence of perineural infiltration
- Variable histologic appearance but difficult to correlate with clinical behaviour
- High rate of morbidity due to recurrence both locally and at distant sites particularly lung. Note 20% with pulmonary metastases survive more than 5 years
- Slow growth rate make five year survival unreliable. Spiro reports 5-year survival of 60% and 20-year survival of 20%
- Treat by widest local excision with preservation of uninvolved major nerves. Adjuvant post-operative radiotherapy indicated.

Adenocarcinoma
- Uncommon tumour most frequently found in the parotid gland
- M=F affects any age and is one of the commoner tumours seen in children
- Histologic appearance varies between low-grade well-differentiated papillary or mucinous patterns to high-grade, undifferentiated lesions
- The incidence of distant metastases is about 40% for high-grade tumours and is directly related to survival rates - 75% 5-year survival for low-grade tumours and 19% 5-year survival for high-grade tumours
- Treatment is by wide local resection with elective neck dissection and adjuvant radiotherapy for clinically high-grade tumours.
Malignant Mixed Tumour

- 99% arise from a pleomorphic adenoma usually after a period of 10-15 years. Frequency varies between 2% and 5%
- The most aggressive of all malignant neoplasms with a high incidence of haematogenous metastases and 5-, 10- and 15-year cure rates of 40%, 24% and 19% respectively
- Radical resection with adjuvant radiotherapy offer the best form of management

Squamous cell carcinoma

- M:F = 2:1
- A rare tumour often mistaken for either a high-grade mucoepidermoid lesion or metastasis from another primary site
- Tends to occur in the elderly - 7th decade
- Very bad prognosis - Should be treated as high-grade mucoepidermoid lesions.

References
TUMOURS INCLUDED
Tumours in the sinonasal region are rare, affecting <1/100,000 people per year but it represents the area of greatest histological diversity in the body. Although squamous cell carcinoma remains the commonest tumour, every tissue type may occur. The commoner epithelial tumours include adenocarcinoma, olfactory neuroblastoma, adenoid cystic carcinoma and malignant melanoma. Sarcomas include chondrosarcoma and rhabdomyosarcoma. All areas of the nasal cavity and paranasal sinuses can be affected but the lateral wall, ethmoids and maxillary sinus are the most common primary sites. The sphenoid and frontal sinuses are rarely primarily affected for reasons that are unknown.

CONFIRMATION OF DIAGNOSIS
Biopsy (optimally performed under general anaesthesia to obtain representative tissue. An experienced histopathologist is essential as up to 1 in 5 cases may have their diagnosis modified after expert review). An endonasal endoscopic approach should be used to avoid transgression of normal tissue planes.

Imaging (experienced radiologist) CT - direct coronal and axial cuts with intravenous contrast enhancement
MRI - three planar T1 pre- and post-gadolinium DTPA, +/- T2, fat suppression & MRA as appropriate

CONSULTATION
• Oral and orbital prosthetic rehabilitation
• Neurosurgical expertise
• Medical oncology

TREATMENT OPTIONS
Can only be determined following biopsy and imaging.
• Surgery alone or in combination with radiotherapy is required in the majority of cases.
• Radiotherapy may be given before or after surgery. The patient should be immobilised in a beam directing shell. The target volume is determined on the basis of CT and MR images and clinical and/or surgical assessment. Planning may be done with orthogonal simulator pictures using bony landmarks, or the patient may be CT planned. Care should be taken to shield the brain stem, optic pathways, eyeball, lacrimal gland and orbit wherever possible. Megavoltage photons should be used. The usual dose will be 60-66Gy in 30-33 fractions over 6-6.5 weeks, treating five days a week. Alternative established fractionation schedules are acceptable. The neck nodes do not require prophylactic treatment.
• The use of concomitant chemotherapy with radiotherapy is increasingly indicated in both the preoperative and postoperative situation for patients with squamous cell carcinoma, other tumours such as embryonal rhabdomyosarcoma, and advanced lymphoma.
• Topical chemotherapy (5-FU) combined with repeated meticulous surgical debulking has been advocated for squamous cell carcinoma and adenocarcinoma in two centres but their results have not been replicated elsewhere. Success for the technique requires careful adherence to the established protocol and is best reserved for those centres who have carefully studied it. It is, however, not without its problems.
• Brachytherapy may be used for squamous cell carcinoma of the columella and anterior nasal septum.
• Radiotherapy alone is required for tumours such as lymphoma or for palliation.
Primary Tumour

Surgery

(a) Craniofacial Resection

- Tumours commonly treated by craniofacial resection include adenocarcinoma, olfactory neuroblastoma, chondrosarcoma and squamous cell carcinoma.
- Tumours involving the skull base requiring surgery may be performed via an extended lateral rhinotomy or a combined midfacial degloving and bifrontal craniotomy through a bicoronal flap or spectacle incision.\(^1,3,21,31\)
- May be extended to include a maxillectomy and resection of orbital structures.
- Neurosurgical expertise may be required.

(b) Maxillectomy

- Squamous cell carcinoma is commonest indication for this procedure.
- Tumours involving the palate and/or extending through the posterior wall to involve the pterygoid region and infratemporal fossa.
- Via a midfacial degloving, lateral rhinotomy or Weber-Fergusson/Diffenbach incisions.
- Medial, subtotal, total, extended.
- May be combined with clearance of the orbit or extended to craniofacial resection.
- Immediate prosthetic rehabilitation with modified denture and gutta percha is optimal. The postoperative care may be reduced where an obturator cannot be fitted, by insertion of a free flap e.g. rectus abdominis.

(c) Lateral rhinotomy (partial or medial maxillectomy)

- Commoner indications include malignant melanoma of the nasal mucosa.
- Tumours involving the nasal septum and lateral wall (i.e. medial maxilla, ethmoids and frontal sinus) of the nose, up to but not including the anterior skull base.
- Offers rapid access with reasonable cosmesis e.g. elderly patients.

(d) Midfacial degloving\(^11\)

- Procedure used.
  - Alone to access both maxilla, ethmoids and sphenoid and nasal cavity.
  - Combined with a bicoronal incision for access to the skull base for craniofacial resection.

(e) Orbit (see attached algorithm) : resection with (exenteration) or without (clearance) of the eyelids

(f) Total rhinectomy and tumours of the cartilaginous nasal skeleton

- Required for extensive tumours of the anterior cartilaginous septum and nasal dorsum.
- Commonest indication is for squamous cell carcinoma.
- Prosthetic or local flap reconstruction.

(g) Endoscopic endonasal approaches

Suitable for selected cases of benign neoplasia e.g. inverted papilloma.\(^{23,36,33}\) and a small number of malignant tumours e.g. malignant melanoma.\(^{6,34}\) There are insufficient numbers and/or follow-up to justify an exclusive endoscopic approach in the majority of malignant tumours at present though it may be combined with other approaches e.g. bifrontal craniotomy.\(^{35}\)

(h) Neck dissection is only indicated in the presence of cervical metastases.\(^{24}\)
HISTOLOGY

Expert histology with frozen section service is required. When in doubt, services of a superspecialist laboratory eg Royal National Throat, Nose & Ear Hospital may be useful.

Individual histologies

Squamous cell carcinoma
Maxilla or antro-ethmoid, rarely nasal vestibule, columella & septum
Combined surgery and radiotherapy (may be given pre or post-operatively)
Five year actuarial survival 32%, ten years 32% , 7,13,21

Adenocarcinoma associated with hard wood exposure though not exclusive29
Ethmoid or antro-ethmoid
Surgery (craniofacial +/- maxillectomy) +/- DXT
Five year actuarial survival 43%, ten year actuarial survival 38% 21
87% five year survival claimed for 5FU/debulking 15

Adenoid cystic carcinoma
Widespread local dissemination by perineural lymphatic and embolic dissemination.
Blood borne metastasis to lungs common.
Locally extensive surgery + post-operative radiotherapy. The addition of radiotherapy delays local recurrence but does not affect overall prognosis.
Five year actuarial survival 53-73%, ten year survival 7-41%, twenty year survival < 8%.10,21

Olfactory neuroblastoma
Arises olfactory epithelium e.g. superior nasal cavity with frequent involvement of anterior cranial fossa via olfactory tract 8
Craniofacial resection to stage disease, followed by radiotherapy (or chemotherapy) if anterior cranial fossa involvement present
Five year actuarial survival 62-78%, ten year survival 47-71% 5,21,28
In patients with cervical metastases (5%) survival was 29% compared with 64% for patients with N0 disease (odds ration 5.1)

Chondrosarcoma
Involves perpendicular plate of ethmoid and skull base
Surgery (repeated as required) eg craniofacial resection, midfacial resection, lateral rhinotomy
Five year actuarial survival 62%, ten year survival 47% 21

Malignant melanoma
Arises from melanocytes, most frequently within nasal cavity
Behaviour - capricious and unpredictable with disseminated metastatic and local recurrence possible at any time. Outcome not determined by depth of invasion as in cutaneous lesions
Wide surgical excision primarily
Post-operative radiotherapy shows tendency to delay recurrence but no statistical benefit demonstrated 2,22
Chemotherapy & immunotherapy not shown to be effective25

Chordoma
Embrological remnant of notochord
Surgery + post-operative radiotherapy.
Inverted papilloma
Typically arises in middle meatus, extending into maxillary sinus, ethmoid and frontal sinuses; may invade adjacent bone leading to frequent ‘recurrence’
Potential for malignant transformation in 1-2% of cases
Surgical excision, approach determined by extent eg endoscopic, midfacial degloving, lateral rhinotomy. 16,23,36,32

Angiofibroma
Arises sphenopalatine region with erosion of medial pterygoid plate and extension into nasopharynx, sphenoid, pterygo-palatine canal and infratemporal fossa. 17
Surgery eg mid-facial degloving 11 +/- radiotherapy for recurrence. Recurrence significantly reduced by drilling out of basisphenoid. 12 Endoscopic excision combined with embolisation also possible. 14,26

FOLLOW UP
• Imaging: (ideally MRI; T1 pre- and post-gadolinium DTPA) baseline at three months, then at four to six monthly as appropriate to histology. 20
• EUA and decrusting of cavity initially three to four monthly, then six monthly as appropriate to individual patient and histology.

SINONASAL MALIGNANCY AND THE ORBIT

Biopsy/Imaging

Craniofacial Resection

Lamina papyracea Eroded

Tumour through periosteum

Orbital Clearance

Lamina papyracea Intact

Tumour adjacent to periosteum

Resect periosteum Frozen section

· Recurrence

Orbital preservation

Periosteum clear
References:


17. Lloyd G., Howard DJ, Phelps P, Cheesman A. Juvenile angiofibroma the lessions of 20 years of modern imaging. J of Laryngol and Otology,1999 113:127-134 (Level 3)


DIAGNOSIS AND STAGING

Common presenting symptoms include:
- Nasal blockage
- Conductive deafness due to an obstructing primary tumour
- Cranial nerve palsies due to skull base invasion
- Cervical lymphadenopathy.

Tumour within the nasopharynx may be either obvious or undetectable on examination in the outpatient clinic. Either way, examination under anaesthesia and biopsy is essential to confirm the diagnosis and to stage the disease. Carcinoma needs to be distinguished from lymphoma and other rarer tumours, which can also occur at this site but are managed differently. In the patient who presents with cervical lymphadenopathy only, the finding of Epstein-Barr virus (EBV) titres in serum, e.g. IgA antibodies to early antigen - diffuse (Ead) and viral capsid antigen (VCA) and anti Epstein-Barr nuclear antigen (EBNA) may help in confirming the diagnosis. If available, biopsy material may be tested for genomic material by amplification of DNA with a polymerase chain reaction thereby providing strong evidence to support a diagnosis of a nasopharyngeal primary tumour.

Even if the nasopharynx appears normal at EUA in the investigation of cervical lymphadenopathy, blind biopsies should be taken from the nasopharynx (as well as from the tongue base and an ipsilateral tonsillectomy should also be performed) to detect an occult primary tumour. CT scanning and MRI provide complementary information in staging the local extent of disease. Because there is a significantly greater incidence of distant metastases at presentation in patients with nasopharyngeal carcinoma compared with those with tumours at other head and neck primary sites, all patients should be fully staged to exclude the presence of bone, liver and lung metastases. Patients with suspected or confirmed nasopharyngeal carcinoma, should be jointly assessed by a head and neck surgeon and an oncologist in a combined clinic.

TREATMENT OPTIONS IN LOCALISED DISEASE

Radiotherapy
High dose radiation therapy with or without chemotherapy is the primary treatment of nasopharyngeal carcinoma both for the primary tumour site and both sides of the neck even in patients without palpable neck disease and regardless of T stage. Radiation therapy dose and field margins are individually tailored to the location and the size of the primary tumour and lymph nodes. Although most tumours are treated with external beam irradiation exclusively, in some tumours radiation therapy may be boosted by intracavitary or interstitial implants when clinical expertise is available and the anatomy is suitable.

Chemotherapy
There is now level I evidence, in addition to a substantial body of less strong evidence, to show that platinum based chemoradiotherapy produces better results than radiotherapy alone, albeit at a cost in terms of toxicity. Not all randomised controlled trials comparing radiotherapy and chemotherapy with radiotherapy alone for nasopharyngeal carcinoma have shown improved outcome, however.

Nonetheless, most oncologists will choose to use chemotherapy as well, especially in more advanced disease.

Surgery
Surgery has no place in the initial management of nasopharyngeal carcinoma, other than to obtain tissue for diagnosis.
It is reserved for the salvage, when feasible, of patients with nodes that fail to regress after radiotherapy, or whose nodes reappear following complete clinical response, and in highly selected cases of localised recurrence at the primary site.

**METASTATIC DISEASE**

Patients with distant metastases at presentation are probably incurable by any means, nonetheless relatively aggressive systemic treatment with chemotherapy is often beneficial in fit patients and can result in prolonged remissions. Consolidation of an early response with high dose radiotherapy to the primary site and neck may then be indicated to provide durable symptom control.

**RECURRENT NASOPHARYNGEAL CANCER TREATMENT OPTIONS**

Selected patients who either have failed primary treatment with radiotherapy of postnasal space carcinoma or have a recurrence of disease may be treated with further external beam or interstitial radiotherapy, or be considered for surgical resection. Most experience has been with the surgical implantation of gold grains into the nasopharynx either by a palatal split or under direct vision. Five-year survival rates have been reported as 79.1% for residual disease and 53.6% for recurrent disease. Patients however who had evidence of disease extension outside the nasopharynx had lower control rates. Surgical resection can also be considered for persistent or recurrent disease in the nasopharynx, and this is particularly applicable to disease that has spread into the paranasopharyngeal space but not yet involved the internal carotid artery. Five-year survival rates have been reported as high as 73% in selected cases with a transpalatal, transmaxillary and/or transcervical approach. Most experience is with the transmaxillary approach. Neck dissection is also indicated for nodal recurrence. If a patients has metastatic disease or local recurrence no longer amenable to surgical or radiation treatment palliative chemotherapy may be an option.

**MORBIDITY**

Radical treatment is associated with substantial mucositis, which can compromise nutrition, and PEG feeding may be beneficial. Chemoradiotherapy with cisplatin and inevitable irradiation of the petrous temporal bone makes sensorineural deafness common. Carboplatin may be less ototoxic but there are fewer data about its efficacy. The parotid salivary glands are inevitably irradiated and xerostomia is universal. Oral pilocarpine benefits a few patients. Dentition is at risk in the absence of saliva. Meticulous attention to oral hygiene is therefore necessary to prevent accelerated dental decay. If extractions of carious teeth become necessary there is the added risk of osteoradionecrosis. Thyroid function testing should be performed prior to therapy and as part of follow up as about one third of patients may develop primary hypothyroidism following neck irradiation. Reirradiation may lead to temporal lobe and temporal bone necrosis.

**OUTCOME**

Small localised cancers of the nasopharynx, although rare, are highly curable by radiation treatment with 5 year survival rates approaching 80 to 90 percent. Moderately advanced lesions without clinical evidence of spread to lymph nodes are often curable with survival rates of 50 to 70 percent.

Patients with advanced lesions especially those associated with clinically positive cervical lymph nodes, cranial nerve involvement and bone destruction are poorly controlled locally by radiation treatment, with or without surgery and often develop distant metastases despite local control. Most recurrences occur within 5 years of diagnosis.

**References:**

Section 5  Chapter 9  Ear and Temporal Bone

In reviewing the current literature on this subject, both Medline & PubMed were used to search the literature, and papers reviewed using a 15 year limit (although some earlier papers were sourced by reviewing references noted in recent papers). In the available literature there are no prospective randomised or nonrandomised trials evaluating the treatment options, and no uniformity in terms of treatment exists.

Introduction.

Cancers arising in the structures of the temporal bone are extremely rare accounting for only 0.05% of head and neck cancers (1). In a study from England & Wales, the age-adjusted incidence remained stable at about 1 / 1000,000 per year for women and 0.8 / 1,000,000 for men from 1968 to 1977 (2).

In several large series approximately 70% of malignancies arose from the auricle, 20 % from the external auditory canal, and 10% from the middle ear and mastoid (3).

Tumours may involve the ear in the following ways:

- Primary cancers involving the ear can arise from the auricle, the external auditory canal 2w(EAC), or the middle ear and temporal bone. More than 70% of ear cancers originate on the pinna (4).
- Tumours from adjacent sites can extend into the structures of the temporal bone. These include malignancies arising from the parotid gland, temporomandibular joint (TMJ), and skin of the preauricular area or postauricular sulcus.
- Metastases from tumours arising in the breast, kidney, lung, prostate, and other sites have been documented (5).

Optimal setting & team membership.

A multidisciplinary approach is required for all types of cancer involving the ear, and management may involve one or more of the following:

- Surgical oncologist (usually an otolaryngologist re EAC/Temporal bone)
- Neurosurgeon (for those more extensive tumours with dural involvement)
- Plastic & reconstructive surgeon (for more extensive tumours of the auricle and reconstruction of larger resection defects)
- Clinical oncologist
- Neuroradiologist
- Cytopathologist with an interest in Head & Neck pathology
- Macmillan specialist nurse
- Speech therapist
- Dietitian

Confirmation of diagnosis.

Histologic confirmation of the diagnosis is usually required before the planning of definitive treatment and this may be obtained under local or general anaesthetic. Some lesions on the pinna may be suitable for excisional biopsy, which will prove to be the definitive treatment.

In the case of parotid tumours invading the ear (and secondarily involved lymph nodes), fine needle aspiration cytology should distinguish benign from malignant disease in 90% of cases (6).
Imaging

Tumours of the pinna may require no imaging unless there is associated cervical lymphadenopathy. However, for those tumours involving the EAC high-resolution computed tomography (CT) is the investigation of choice for assessing the bony anatomy of the temporal bone. Arriaga et al (7) investigated the utility of preoperative CT and concluded that the pathologic extent of tumour growth can be accurately assessed with this modality. Magnetic resonance imaging (MRI) is more useful to define the tumour from brain and from the reactive / inflammatory changes that may occur, and gives information regarding internal carotid artery (ICA) and sigmoid sinus patency, by the presence or absence of flow void signals. Carotid angiography may however be necessary to establish unequivocally involvement of the carotid artery. If involvement of the ICA is suspected on angiography, then its sacrifice (or reconstruction) may be considered by some surgeons as part of the resection. Under these circumstances some assessment of the effect of ICA occlusion on intracranial circulation is required – in the UK this would usually involve a test balloon occlusion of the ICA under local anaesthetic in order to assess any neurologic sequelae.

Staging

The American Joint Committee on Cancer (AJCC) states that the goal of a staging system is to evaluate the efficacy of treatment and to provide a sound basis for therapeutic planning in cancer patients, by describing in comparable form the survival and results of treatment of different patient groups. Currently no staging system for malignancies involving the ear is accepted by either the International Union Against Cancer (UICC) or the AJCC.

Treatment Guidelines

1. **Cutaneous carcinoma of the pinna.**
The most common method of treatment for auricular skin cancer is surgical resection. Resection can be accomplished in a traditional fashion or with Mohs micrographic technique (8). If there are involved nodes then this surgery may need to be performed together with a parotidectomy and neck dissection. Surgical treatment of auricular skin cancer poses several anatomic challenges that are unique to the ear. The greatest challenge comes in dealing with the underlying cartilaginous skeleton of the external ear. A judgment must be made whether auricular cartilage must be sacrificed. Reconstruction of the pinna would normally require the input of a plastic and reconstructive surgeon, and there are occasions when the use of a prosthetic ear is the most suitable option. Radiotherapy can offer an excellent cure rate for smaller carcinomas of the ear. Concerns about radiotherapy centre on the uncertainty of tumour margin control, the possibility of radiation necrosis, and questionable efficacy if cartilaginous and bony invasion are present (9).

2. **Carcinoma involving the EAC / Temporal bone.**
The lack of an accepted staging system, the rarity of malignancy involving the EAC and temporal bone and the wide variety of treatment plans that have been employed make it difficult to extract definitive information from the literature regarding the optimal treatment and prognosis. Clinical experience therefore dictates management decisions and complete surgical resection with a clear microscopic margin must be the preferred initial primary treatment goal in a patient with a resectable cancer.

The steps in the decision making process involve the following:
- Histopathologic confirmation of malignancy.
- Radiologic determination of disease extent and metastatic spread.
- A realistic assessment of whether surgery is aimed at palliation or cure.
- The need to include the parotid, TMJ, infratemporal fossa, neck exploration, carotid artery, and dural or cerebral resection or reconstruction as part of the planned resection.
• The necessity or type of reconstructive options.
• An opinion from a clinical oncologist regarding adjuvant radiotherapy.
• A neurosurgical opinion if a more formal skull base resection is required

A widely accepted operative concept is to free the involved temporal bone from its surrounding venous sinuses, protect the internal carotid artery and brainstem, and avoid injury to cranial nerves if they are still functioning.

A number of surgical approaches (that largely depend on the extent of the tumour) are available that satisfy these criteria and they include:

• Mastoidectomy – includes all types of modified radical and radical mastoidectomy.
• Lateral temporal bone resection (TBR) – removal of the osseous and cartilaginous external auditory canal, tympanic membrane, malleus and incus.
• Subtotal TBR – includes the additional removal of the otic capsule.
• Total TBR – involves the additional removal of the petrous apex

The above procedures may be combined with superficial parotidectomy and neck dissection dependent on the extent of the local disease and the presence of associated lymphadenopathy. Involvement of dura or brain, have led some workers to perform extended temporal bone resections. No groups have shown any impact on survival with this approach except Moffat et al (10). Moffat performed salvage surgery in 15 cases with a 47% 5 year survival.

Surgical resection and reconstruction of carcinomatous involvement of the ICA is also controversial, with no studies able to show an improved survival with this aggressive approach. Radiotherapy is seldom advocated as the sole treatment modality. The precise role of preoperative or postoperative radiotherapy is unclear. A review by Prasad & Janecka (11) has indicated that the addition of radiotherapy to patients with disease confined to the EAC and treated with lateral TBR confers no survival advantage. The role of radiotherapy in patients treated with subtotal TBR was unclear.

Guidelines from the American Society for Head & Neck Surgery and The Society of Head & Neck Surgeons (12) for postoperative radiotherapy include:

• Close resection margins (less than 5 mm), when proximity of tumour to important structures such as ICA or facial nerve preclude wide margins.
• Positive resection margins.
• Perineural invasion.

These conditions apply to the majority of resections for temporal bone carcinoma thus postoperative radiotherapy is indicated in most cases.

Prognosis.

Cutaneous carcinoma of the pinna has been described by several authors as having a higher rate of recurrence and a worse prognosis than with other skin cancers. Byers & colleagues (13) in a review of 486 patients with squamous cell carcinoma of the ear reported a local recurrence rate of 14% with death in 2.5% from local failure.

The prognosis in carcinoma involving the EAC / Temporal bone where the disease is confined to the EAC is approximately 50% 5 year survival but falls to approximately 29% with middle ear involvement (11).
Summary

Carcinoma involving the temporal bone is a rare disease. Lack of prospective randomised or non-randomised trials evaluating the treatment options, means that clinical experience currently dictates management decisions. Complete surgical resection with a clear microscopic margin must be the preferred initial primary treatment goal in a patient with a resectable cancer. In patients with disease confined to the EAC this would involve lateral TBR.

The precise role of preoperative or postoperative radiotherapy is unclear. Postoperative radiotherapy is indicated in the most cases.

References:
Introduction

Approximately 8% of the UK population have nodular thyroid disease. A significant proportion are clinically solitary. Thyroid nodules are more common in women with increasing age. In contrast, thyroid cancer is rare (2 to 3 cases per 100,000 per annum in the UK). The commonest presentation is as a solitary thyroid nodule in a euthyroid patient and in that group, the incidence of malignancy is between 10 and 20%. The majority are differentiated thyroid cancer, and most have an excellent prognosis. A minority present with local and distant metastases e.g. with metastatic neck disease, airway and pharyngoesophageal obstruction, and bony or pulmonary secondaries. Differentiated thyroid cancer (90% of all thyroid cancer) consists of papillary or mixed papillary and follicular (80%) and follicular cancer (10%). There have been no prospective randomised studies investigating treatment outcomes for this condition, so that the consensus view expressed in these guidelines is formed predominantly from retrospective analyses (level II-III evidence).

Optimal Setting and Team Membership
Multidisciplinary thyroid clinic to include:  Endocrinologist (ideally 2 for leave cover)
Specialist Surgeon (ideally 2 for leave cover)
Specialist in Thyroid Cytology and Pathology
Specialist Radiologist
Clinical Oncologist
MacMillan Specialist Nurse

Diagnostic evaluation

History should include past history of:-
- Ionizing radiation to the head or neck
- Family history of thyroid cancer
- Personal history of previous thyroid disease
In addition, patients should be asked whether they have developed a hoarse voice, difficulty in swallowing or breathing.

Examination includes not only neck evaluation but also vocal cord function

Initial investigations
- All patients should have baseline thyroid function tests (TSH, and free T4 with free T3 as appropriate). Suppression of TSH is a common and non-specific finding in subjects with goitre. Overt thyroid dysfunction, indicated by raised serum concentrations of free T4 and T3 or biochemical hypothyroidism, is frequently associated with a goitre and such patients do not generally require further investigation. Assessment of thyroid antibody status may assist in the diagnosis of chronic lymphocyte thyroiditis, predict postoperative hypothyroidism and aid the interpretation of both thyroid function and serum thyroglobulin measurements.
- All euthyroid patients presenting with thyroid enlargement should have fine needle aspiration cytology (FNAC) as a mandatory initial investigation. It is important to document which side and what part of the goitre has been aspirated. Fine needle aspiration cytology results are reported and classified as inadequate, benign, suspicious, or frankly malignant. A strategy for the evaluation of the solitary or dominant thyroid nodule is shown in the appendix.

The following are not routine, but may be of value following initial evaluation if the diagnosis is not clear, or when the diagnosis of cancer has been established.
• **Radiological Imaging**
  Chest x-ray may demonstrate tracheal deviation, mediastinal extension of disease or lymphadenopathy, and pulmonary metastases or associated co-morbidity. Soft tissue x-rays of the neck and thoracic inlet may show tracheal deviation and compression but are less accurate than respiratory flow loop studies. Anatomical imaging with CT or MRI can be used to evaluate the thyroid, neck, midline visceral compartment, mediastinum, chest and abdomen.

• **Ultrasound of the neck**
  This can be helpful in differentiating a solitary from a multinodular swelling, in measuring tumour size and assessing the contralateral lobe. It may be used to evaluate complex cysts and distinguish purely cystic nodules from complex cysts with a solid component, but it is unable to differentiate reliably between benign and malignant disease.

• **Thyroid scintigraphy**
  The commonest investigation currently used is scanning with Technetium (Tc$^{99m}$) pertechnetate. This test is poorly specific and sensitive in the diagnosis of thyroid cancer. This is, in part, because technetium is trapped and not organified by the thyroid and since some cancers retain the ability to trap and not organify, (and also have a high blood flow), “cold” nodules may appear “hot” using this technique. However, scintigraphy can be useful to assess the state of the thyroid bed following previous surgery, to measure tumour size and assess the contralateral lobe. The best way to evaluate whether or not a thyroid nodule is truly functioning is to use $^{123}$I scintigraphy. Truly functioning nodules, (ie “hot”) are unlikely to be malignant (less than 1%) and in those nodules with a suspicious FNA (hyperplasia versus low grade neoplasia), $^{123}$I scintigraphy may in the future be an alternative in some cases to a diagnostic lobectomy.

**Treatment Guidelines**

- The following treatment modalities are available for thyroid cancer.
  - Surgery
  - Radioactive iodine
  - External beam radiotherapy
  - Thyroxine therapy
  - Chemotherapy

- The main stay of treatment for differentiated and medullary thyroid cancer is surgery
- All differentiated and medullary thyroid cancers should be TNM staged
- There are a number of prognostic factors associated with differentiated thyroid cancer:
  - **Patient factors** Age, gender
  - **Tumour factors** Stage and histology
  - **Management factors** Delay in therapy, extent of surgery, experience of the surgeon, thyroid hormone therapy and treatment with radioiodine post-operatively.
  - Patients should be given proper informed consent and the opportunity to contribute openly and freely in any management discussions. Patient information leaflets should be readily available.

**Papillary Thyroid Cancer**

**Primary Tumour**
- The aim of treatment is to remove all macroscopic and microscopic disease with minimum morbidity. The minimum operation that should be performed on a solitary thyroid nodule is a diagnostic lobectomy (or isthmusectomy for a midline lesion).
- Although there is no prospective evidence, a consensus view based on retrospective data suggests that risk stratification treatment for differentiated thyroid cancer can be based on the TNM (tumour, nodes, distant metastases) classification combined with patient gender. This is shown below.
Risk stratification treatment based on TNM classification including gender, age, stage and histology (GASH)

- Age (less or more than 45 years)
- Gender (males carry a worse prognosis than females)
- Tumour size
- Tumour histology
- Nodal and distant metastases

Selection of a treatment policy for differentiated thyroid cancer by allocation into risk groups depending on gender, age, stage and histology facilitates surgical treatment which is shown in Figure 1.

Table 1 - Surgical treatment of differentiated thyroid cancer

<table>
<thead>
<tr>
<th>Risk Group</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Low risk patient with low risk tumour</td>
<td>lobectomy</td>
</tr>
<tr>
<td>Low risk patient with high risk tumour</td>
<td>lobectomy or TT</td>
</tr>
<tr>
<td>High risk patient with low risk tumour</td>
<td>lobectomy or TT</td>
</tr>
<tr>
<td>High risk patient with high risk tumour</td>
<td>total thyroidectomy</td>
</tr>
</tbody>
</table>

Intermediate group

TT = Total thyroidectomy

Low risk patients include females under the age of 45
High risk patients include all males, and females over 45
Low risk tumours include papillary carcinomas less than 1cm in size, and minimally invasive follicular carcinomas less than 1cm in size.
High risk tumours include papillary and follicular carcinomas greater than 1cm in size. Also included is any tumour associated with significant multifocality, or local or distant spread.

Patients under 16 with a diagnosis of cancer should be regarded as high risk and, are usually best treated aggressively.

Papillary thyroid carcinoma (PT1)
Solitary papillary thyroid cancers which measure less than 1cm in size (PT1) and which are surrounded by an adequate margin of normal thyroid tissue, in the absence of cervical or distant metastases, can be treated by unilateral lobectomy, subsequent TSH suppression with thyroxine and thyroglobulin surveillance. Tumours in the isthmus can similarly be treated by an isthmusectomy and a 1cm margin of resection of normal tissue.

Papillary thyroid carcinoma (PT2)
The treatment of this lesion (1-4cm in size) is controversial. Low risk patients, that is females under 45 years of age with solitary, non metastatic tumours greater than 1cm but less than 3cm in size (small PT2 N0 M0 tumours) can also probably be safely treated with lobectomy, TSH suppression with thyroxine and subsequent monitoring of the serum thyroglobulin. However, in men, and women with larger tumours, the risk of recurrence is higher. Such patients should have total thyroidectomy, postoperative radio-iodine ablation of residual thyroid tissue, TSH suppression with thyroxine and monitoring of the serum thyroglobulin.

Papillary thyroid carcinoma (tumours greater than 4cm in size, PT3)
All patients with tumours greater than 4cm in size and any tumour where there is significant multi-focal disease, extracapsular spread, lymph node or distant metastases should have a total thyroidectomy, with preservation of both recurrent laryngeal nerves (and external laryngeal nerves where possible) selective neck dissection as required to clear disease, followed by radio-iodine ablation of residual tissue. Long-term TSH suppression with thyroxine and serial measurements of serum thyroglobulin are important.
For many patients with thyroid cancer, total thyroidectomy with parathyroid preservation will be the operation of choice. If there is any concern regarding the morbidity resulting from a truly total thyroidectomy, a near-total thyroidectomy can be performed in the absence of contralateral disease. In this procedure, a sliver of the contralateral lobe including the parathyroids is preserved. The remnant of normal thyroid tissue can then be ablated with radioiodine.

During thyroidectomy, the use of the nerve stimulator to identify the recurrent laryngeal nerve is not routinely recommended but may be of value in re-do surgery, and also when attempts to identify the external laryngeal nerve prove difficult.

Postoperative hypocalcaemia should be managed in line with local protocols.

Papillary thyroid cancer arising in a thyroglossal duct cyst can be managed conservatively as long as the patient has undergone Sistrunk’s operation, that the resection has been microscopically complete and that the remaining thyroid gland is normal both on palpation and on investigation by ultrasound or radioscintigraphy. Postoperatively, treatment is with TSH suppression with thyroxine and sequential measurement of the serum thyroglobulin.

When an unsuspected papillary thyroid carcinoma is discovered in a multinodular goitre resected by either lobectomy, near- total or total thyroidectomy, if excision margins are free of disease, then treatment is TSH suppression with thyroxine and monitoring of the serum thyroglobulin. If clearance is questionable, or a follicular or multifocal component present, then completion thyroidectomy and post operative radioiodine ablation are necessary.

The Neck
Node negative disease (N0)
The extent of nodal surgery is determined by the degree of lymphatic involvement. In the clinically N0 neck, level 6 should be routinely dissected and levels 2 to 5 and 7 palpated at the time of surgery. Suspicious nodes can be subjected to frozen section and if involved, a selective neck dissection (at least levels 2 to 4) should be performed.

Node positive disease (N+)
Clinically palpable disease in levels 2 to 5 indicates the need for at least a selective neck dissection involving these levels and depending on the size and extent of disease, a modified radical, radical or extended radical neck dissection may be required.

The aim of combined total thyroidectomy and limited lymphadenectomy is to remove all microscopic malignant cervical lymphadenopathy with minimum associated co-morbidity ie, the preservation of non-lymphatic structures such as the internal jugular vein, the sternomastoid muscle and the accessory nerve.

Follicular thyroid cancer
The management of follicular adenocarcinoma is similar to that of papillary tumours. The mainstay of treatment is surgery. All follicular adenomas and small follicular cell carcinomas < 1cm with minimal vascular invasion (PT1) can be managed safely by lobectomy alone. Surgery should be followed by TSH suppression with thyroxine and sequential measurement of the serum thyroglobulin.

For the more advanced tumours, surgery usually entails a near-total or total thyroidectomy although some patients in the intermediate group with tumours less than 3cm in size with minimal invasion (as for papillary cancer) may be treated conservatively with hemithyroidectomy. The neck and the mediastinum are managed as described for papillary carcinoma.

In advanced cases of differentiated thyroid cancer, invasion of local visceral structures by continuity and contiguity may indicate that, after appropriate imaging, extensive extirpative surgery is appropriate to
include total laryngectomy, tracheal resection, partial or complete pharyngectomy and neck dissection. It is important to exclude any major vessel involvement prior to surgery and the help of a cardiothoracic surgeon may be required for mediastinal access.

Hürthle cell adenomas may be treated by lobectomy alone. Hürthle cell carcinomas represent variants of follicular cell carcinoma but are more aggressive and are less likely to concentrate radioiodine and therefore, should be managed more aggressively with total thyroidectomy.

Radiotherapy for differentiated thyroid cancer.

Radioiodine ablation and therapy

- $^{131}I$ (Radio-iodine) is used following surgery for a differentiated thyroid cancer to ablate residual normal thyroid tissue and to search for and treat any metastatic disease. The protocol to be followed for both ablation of residual normal thyroid tissue and treatment of any functioning local residual disease or distant metastases is shown in Figure 2.
- The therapeutic isotope used is $^{131}I$
- Patients should be admitted to hospital and accommodated in a single room equipped for radiation protection.
- As many patients with thyroid cancer are women of child bearing age, the possibility of pregnancy must be excluded before radioiodine therapy is given.
- The patient must be off thyroxine for 4 weeks, Liothyronine (T3) for 10 days to allow exogenous TSH levels to rise and stimulate uptake of iodine. In the future, administration of human recombinant TSH may be useful in preventing iatrogenic hypothyroidism.
- The TSH levels should be measured before radioiodine therapy.
- The protocol for therapy should be followed and serum thyroglobulin sequentially measured as indicated.

External beam radiotherapy

This may be appropriate in differentiated thyroid cancer when macroscopic disease has been left behind following surgery, when ablative radioiodine therapy has been ineffective, or when tumours are inoperable.

Chemotherapy

There is no role for chemotherapy in the management of differentiated thyroid cancer.

Management of lung and bone metastases.

Long term survival may be achieved after radioiodine treatment of lung metastases. Caution must be taken in the treatment of miliary nodules because of the risk of radiation pneumonitis and subsequent fibrosis. Fractionated radioiodine dosage is recommended. Soft tissue extradural spinal metastases, considered unsuitable for surgical management, should be treated similarly in order to minimise the risk of spinal cord compression.

Bone metastases are associated with a poor prognosis because of their relatively low level of radioiodine uptake. Some are suitable for resection. Pre-surgical embolisation may be required, particularly in the axial skeleton, in order to reduce vascularity.

Following surgery patients should be considered for radioiodine therapy and external beam radiotherapy to the resection bed. This approach may offer long term disease free survival.

Monitoring of serum thyroglobulin

Thyroglobulin (Tg) is a serum protein which is the key substrate for the biosynthesis and storage of thyroid hormones. In differentiated thyroid cancer, thyroglobulin is a specific, sensitive and therefore extremely useful tumour marker. It is secreted by both normal and cancerous thyroid cells. Following near-total or total thyroidectomy and $^{131}I$ ablation, the measurement of serum thyroglobulin when TSH has been suppressed by T4 therapy is helpful since Tg secretion from normal thyroid tissue is
more TSH-dependent than Tg secretion from neoplastic tissue. Thus, a detectable serum thyroglobulin is highly indicative of residual or recurrent disease. In such patients, the diagnostic sensitivity of serum Tg measurement is enhanced by the elevation of TSH so measurement of Tg after thyroid hormone withdrawal prior to $^{131}$I scanning and ablation may be useful. Thyroglobulin is measured by a specific radioimmunoassay; results should be interpreted with caution if endogenous Tg antibodies are present.

**Follow up**
After initial successful treatment, patients should be followed up by clinical evaluation and serial thyroglobulin measurements (Figure 1). They should be seen 6 monthly for 18 months and then yearly for life.
Figure 1 - Treatment of differentiated thyroid carcinoma Birmingham oncology centre protocol

“TOTAL” THYROIDECTOMY

- Liothyronine (T3) 20 Micrograms tds stop 2 weeks before I-131
- Avoid iodine containing medicines, fish, added salt & x-ray contrast examinations

I-131 Ablation 3GBq
day 3 check Thyroglobulin (Tg) & TSH

- Thyroxine (T4) for 6 weeks only

6 Weeks out-patient appt.
check Tg, TSH, T4

- Change to T3 20 micrograms tds for 2 weeks (stop 2 weeks before I-131)

10 Weeks
I-131 Diagnostic scan 200 MBq check Tg and TSH

Scan positive
Thyroglobulin raised
Metastases

- Recomence T3 20 micrograms tds for 2 weeks (stop 2 weeks before I-131)

14 weeks
I-131 therapy dose 5.5GBq day 3 scan

Scan negative
Thyroglobulin < 10 micrograms/L
No metastases

- Commence T4

Follow-up check
Tg, TSH, T4
(6-12 monthly appt.)
**Medullary thyroid carcinoma (MTC)**

This cancer is rare with 75-80% being sporadic tumours, the remainder a familial syndrome. Multiple endocrine neoplasia (MEN) syndrome 2A consists of MTC in association with phaeochromocytomas and occasionally primary hyperparathyroidism. MEN2B consists of MTC together with neuromas of the lips, tongue and conjunctivae, ganglioneuromatoses of the bowel, a Marfanoid appearance and occasionally phaeochromocytomas. It is usually a new mutation with no obvious previous family history. The third condition is familial MTC in isolation. All 3 familial disorders have been linked to abnormalities of the RET proto-oncogene. MEN2B is a much more aggressive disorder usually presenting in childhood. In MEN2A and familial MTC the clinical disorder usually presents in adult life. Surgery for this disease should be performed in a unit with access to facilities for appropriate biochemical investigations and to clinical and molecular genetics.

**Making the diagnosis**

**Clinical evaluation**
- A family history of thyroid, parathyroid or adrenal tumours or renal stones
- Any history or presence of any evidence of hyperparathyroidism or phaeochromocytoma
- Any history of neuromas of the lips, tongue or conjunctivae or gastrointestinal tract.

**Head and Neck Examination**
- Evaluation of the thyroid, and its relation to visceral structures.
- Any evidence of extrathyroid extension
- The presence or absence of any lymph nodes including their number, size and level
- Examination of vocal cord function.

**General examination**

This should include assessment of the chest and abdomen (including the liver). Look for evidence of a Marfanoid appearance or any evidence of neuromas of the lips, tongue or conjunctivae.

**Genetic studies**

In excess of 90% of patients with any form of familial MTC have been shown to have a mutation of the RET proto-oncogene. Moreover, 98% of cases of MEN2A have mutations in one of just 6 codons. These plus mutations in an additional two codons have been found in isolated familial MTC while just one single different mutation has been found in 95% of all cases of MEN2B. Such a restricted range of mutations means that selective analysis of the RET gene is a very feasible and useful technique.

**Screening for familial MTC**

It is important to identify the apparently sporadic case which in fact is an index case of a familial form of the disease. The finding of multiple foci of tumours together with definite ‘C’ cell hyperplasia virtually guarantees the diagnosis of familial disease although their absence does not exclude it. A family history of any of the tumours associated with MEN2 should raise suspicion.

In likely sporadic cases, family screening should not be undertaken initially and the emphasis should be on increasing the certainty that the case is sporadic. If the clinical information is inconclusive, a limited analysis of the RET proto-oncogene should be undertaken. If this is normal, the likelihood of familial disease is < 0.5%. If a genetic abnormality is found, then this should be used to screen other family members. Patients with likely MEN2B should also have their RET proto-oncogene analysed expecting to find an abnormality which can be used to screen offspring.

Stimulated calcitonin measurements which previously were the mainstay of diagnosis. The test has been replaced by genetic testing. They should only be considered where there is very strong suspicion of familial disease and a normal genetic analysis in the affected case.
It is strongly advised that family screening and genetic counselling is done by, or in close association with, a clinical genetics department so that everyone is aware of the implications of the tests before they are undertaken.

**Imaging studies**
- Chest radiograph (If CT not performed)
- CT to evaluate the thyroid bed, any extension into the midline visceral structures, metastatic neck disease, evidence of disease in the mediastinum as well as the chest. Abdominal CT to assess both the liver and adrenals to exclude phaeochromocytoma (if MEN 2A is suspected). The adrenals are probably best evaluated with MRI.
- Whole body imaging with Te$^{99m}$ (v) dimercaptosuccinic acid (Pentavalent DMSA) together with conventional Te$^{99m}$ MDP bone scanning may be necessary to evaluate, localise and stage systemic spread.
- Imaging with I$^{123}$ - MIBG (Metiodobenzylguanidine) may be useful to evaluate and stage systemic spread, and to confirm or refute the diagnosis of a phaeochromocytoma.

**Laboratory tests**
These should include:
- Thyroid function tests
- basal serum calcitonin levels.
- Serum calcium (if this is elevated, then a parathyroid hormone level should be obtained)
- Serum albumin
- 24 hour urine catecholamine excretion (If MEN 2A is suspected).

**Prognostic factors**
- TNM stage
- Pre- and postoperative calcitonin levels
- Previous surgery

**Treatment**

**Surgery**
- The primary tumour
All patients should undergo a total thyroidectomy, with preservation of both recurrent laryngeal nerves (and external laryngeal nerves where possible). An attempt should be made to preserve all the parathyroids but if there is any evidence of tumour involvement, these should be removed.

- The Neck

**Node negative disease (N0).**
In combination with a total thyroidectomy, the medial compartment of the neck (levels 6) should be dissected from the level of the hyoid down to the brachiocephalic vein, and from one carotid sheath to another. In addition, nodes in levels 2, 3 and 4 should be palpated and any suspicious ones sent for frozen section. The presence of positive disease at the time of surgery indicates the need for at least a selective neck dissection (levels 2-5).

**Node positive disease (N+)**
In those instances where there is either radiological or clinical evidence of cervical nodal spread, then the appropriate neck dissection should be performed.

This should be as conservative as possible and at least levels 2 to 5 should be dissected with preservation of the accessory nerve, the internal jugular vein and the sternomastoid muscle wherever possible. Occasionally, either a modified radical or radical neck dissection may be required.
Radiotherapy
Since MTC does not concentrate iodine, there is no role in this disease for post-operative radio-iodine ablation and therapy. In those cases where tumour excision has been complete, no further treatment is indicated. If there is microscopic evidence of residual disease, then external beam radiation therapy may be beneficial in the following instances
- Multiple large nodal metastases with extracapsular extension.
- Residual macroscopic disease
As an alternative to radiotherapy (or in conjunction with it), some patients who concentrate I$^{123}$ MIBG should be considered for therapeutic treatment with I$^{131}$ MIBG.

Associated phaeochromocytoma
Any evidence of a phaeochromocytoma which is discovered during the preoperative work up of a patient with MTC should be treated and resected first, by a specialist endocrine surgeon before definitive treatment of the thyroid tumour.

Chemotherapy
There is no established role for chemotherapy in the management of patients with MTC

The role of parathyroidectomy
In patients who have the MEN 2A syndrome, parathyroid hyperplasia is common and may occur in 30 - 40% of patients. In this situation, a subtotal or total parathyroidectomy with auto transplantation may be necessary at the time of initial surgery. This avoids the need of having to re-operate later with potential risk for damage to both recurrent laryngeal nerves. Those parathyroids that are removed can be cryopreserved so that if hypoparathyroidism subsequently develops, these glands can be reimplanted.

Follow up
Patients should be followed up for life. The particular schedule for each patient should be tailored to their initial clinical course and subsequent risk of recurrent disease but the following protocol is recommended.
- Six monthly for 5 years
- Then yearly for life.
- Patients should have a head and neck examination and a once yearly measurement of thyroid function
- Yearly serum calcium levels and urine catecholamines should be done in patients with familial MTC
- The role of routine postoperative measurements for serum calcitonin is controversial. If the preoperative calcitonin drops post-treatment to an undetectable level, then this is a good prognostic sign. The presence of an elevated serum calcitonin postoperatively indicates residual or recurrent disease which is highly likely to be incurable. In this situation, there is little evidence that subsequent regular routine measurements of calcitonin and the search for residual and recurrent disease makes any difference to long term prognosis.

Patients should be followed up clinically, and any significant symptoms or signs investigated appropriately. Recurrent disease should be then treated on its merits.

Thyroid Lymphoma
This rare condition presents with a rapidly expanding thyroid mass and often coexistent dysphagia and stridor. There may be a preceeding Hashimoto’s thyroiditis. FNAC may suggest lymphoma. A tru-cut or open biopsy is then required for histological evaluation. This condition needs to be differentiated from anaplastic carcinoma. Formal lymphoma staging is required by a designated clinical oncologist and the subsequent treatment protocol will include chemotherapy plus or minus radiotherapy. The outlook is generally good.
Investigations required.
- Thyroid function tests
• Thyroid antibodies
• FNAC and open biopsy (or tru-cut)
• Whole body CT
• Bone marrow and trephine
• ESR, LDH.

**Anaplastic thyroid cancer**
This condition presents in a similar way to thyroid lymphoma with a rapidly expanding mass, dysphagia and airway obstruction in an elderly patient. Clinical examination usually reveals a firm, fixed, infiltrating mass. There is often a past history of a goitre. An open biopsy is usually required to make the diagnosis and differentiate this condition from lymphoma. Further surgery is not usually indicated other than an isthmusectomy to relieve airway obstruction when a tracheostomy may also be necessary. Subsequent treatment is with external beam radiotherapy. Chemotherapy has no role. The outlook is generally very poor and most patients survive less than 12 months.

**Investigations required**
• Thyroid function tests
• Thyroid antibodies
• FNAC and open biopsy (or tru-cut)
• Neck and chest CT (or chest x-ray).

**Data Collection**
Surgeons carrying out thyroid surgery should collect their data using standardised information packages in line with local guidelines and also audit their results.

**Surgical workload**
Surgeons carrying out this sort of work should be performing at least 30 cases per year (which includes benign and malignant disease). It is recommended that the surgical management of advanced disease, (total thyroidectomy for cancer, neck dissection etc) is performed in centres by recognised experts.

**Outcome measurements**
There are a number of performance indicators which can be used to monitor effective management of thyroid cancer and they are listed below.
• Adequate primary surgery
• Incidence of vocal cord paralysis
• Incidence of hypoparathyroidism (temporary and permanent)
• Achievement of TSH suppression
• Provision and treatment with radiiodine postoperatively when indicated.
• Regular monitoring of serum thyroglobulin
• Abnormal thyroglobulin acted upon
• Quality of life
• Disease free interval
• Survival
A DIAGNOSTIC STRATEGY TO EVALUATE THE "SOLITARY" OR "DOMINANT" THYROID NODULE

1. **Malignant**
   - Surgery
   - Consider Scintigraphy
   - I\(^{123}\) Scan
     - "Hot"
       - Observe
     - "Warm" or "Cold"
       - Surgery

2. **Suspicious (Hyperplasia or low grade neoplasia)**
   - Consider Scintigraphy
   - I\(^{123}\) Scan
     - "Hot"
       - Observe
     - "Warm" or "Cold"
       - Surgery

3. **Benign**
   - Watch.
   - Discharge after 2 negative FNAC's

4. **Inadequate sample**
   - Repeat FNAC

**References:**

9. Mazzaferri, EL. An overview of the management of papillary and follicular thyroid cancer. Thyroid. 1999. 9: 421-427


Section 5 Chapter 11 Non Melanotic Skin Tumours

Introduction

Non-melanotic skin tumours (NMST) encompass an array of malignancies including epithelial, vascular, adventitial, neuroendocrine, haematologic and adnexal. They account for more newly diagnosed cancer cases annually than any other tumour group with the rate rising at between 4% and 8% a year. The vast majority of these tumours are represented by two epithelial malignancies, namely basal cell carcinoma (BCC) and squamous cell carcinomas (SCC)\(^1\)

In the USA, the incidence of basal-cell carcinoma is 300 per 100000\(^2\) and 38.8 per 100000 for squamous carcinoma\(^3\). Of these, up to 90% present within the head and neck region\(^4\)

The histologic morphology of BCC is divided into different patterns with the nodular BCC accounting for the majority of lesions (75%) followed by superficial BCC (10%)\(^5\). There are many histological patterns of SCC lesions as\(^1\)

Presentation and diagnosis

Most ENT centres within the UK rely heavily on a good working relationship with their respective dermatology departments. With this in mind, however, it should be stated that the diagnosis of skin tumours still remains very much in the domain of the dermatologist.

The major predisposing factors for the development of BCC are UV radiation and severe sunburn during childhood. Other factors include Fitzpatrick skin types 1-11 red hair, freckling in childhood, ionising radiation, immunosuppression and a family history of skin cancer\(^6\). The Fitzpatrick Skin Type Scale is based on a combination of genetic disposition, reaction to such exposure and tanning habits (see appendix). There are also some dermatological conditions that predispose individuals to BCC formation including albinism, linear epidermal naevii and xeroderma pigmentosum. The above factors all predispose to the development of SCC as well. In addition, the following factors also predispose to the formation of SCC, namely osteomyelitis, HPV, arsenic exposure and chronic skin conditions (discoid lupus, chronic ulcers, dyskeratosis)\(^7\). Diagnosis of the vast majority of these lesions is by clinical evaluation taking into account the risk factors mentioned above. Where there is doubt, an intra-lesional biopsy may be required to gain a diagnosis. To date there is no system available for NMST staging.

Treatment

Treatment options depend on several criteria, namely tumour type, patient profile, size and location of tumour, recurrence, physician experience and patient preference\(^8\). The management of both SCC and BCC can be divided into surgical and non-surgical treatments with a combination of the two approaches needed in certain cases.

1. Surgical
   This modality can be subdivided into destruction or excision of the lesion:
   • Destruction

   BCC
   
   Curettage and cautery is used for small (< 4mm), well-defined BCC lesions with non-aggressive histology in non-critical sites with a 5-year cure rate of up to 97%. This decreases dramatically to 60% for treatment of recurrent BCC\(^9\).
**Cryosurgery** can be used for single or multiple BCC lesions and the same principles for selection apply as shown above. Again, it is less effective for recurrent lesions but, on the whole, it is well tolerated on an outpatient basis producing good cosmetic results.

**Carbon dioxide laser** is not widely used but can be useful for large or multiple superficial lesions.

**SCC**

Non-excisional modalities can be used in small SCC lesions less than 1cm.

- **Excision**
  Surgical excision is either radical excision with predetermined surgical margins or excision under total microscopic control – Mohs micrographic surgery (MMS)

**BCC**

In the excision of small, well-defined BCC lesions, 2-3mm margins will clear 85% of cases but in the larger or morpheaform BCC's wider margins of 5mm enable an 82% clearance rate. According to Thissen et al, the 5-year recurrence rates of BCC after MMS and surgical excision are 1% and 5.3% respectively. The indications for MMS is for tumours with indistinct margins, in areas where maximum tissue preservation is important (nose, lips) and in cases there is a high risk of recurrence (> 2cm, midface/ears, aggressive histology, long duration).

**SCC**

Treatment is similar to BCC treatment with a few differences. Metastasis to lymphatics and other organs must be evaluated and excluded. Tumours at high risk for local or distant spread are those larger than 2cm or deeper than 6mm, recurrences, poor differentiation, immunosuppressive host and perineural invasion. Excision is the commonest form of treatment with a 4mm margin for low risk lesions and 6mm for high risk ones. MMS is advised in the high-risk group.

**Incomplete Margins of Excision**

The treatment of incompletely excised lesions is controversial. An expectant policy is appropriate where the residual tumour is on the lateral margin(s) only, sparing the deep margin. Added to this, non-aggressive histology, a non-critical anatomical site and a previously non-recurrent tumour are also considered to be suitable for this approach. The study by Sarma et al revealed that only 7% of incompletely excised lesions contained residual tumour upon re-excision, thus justifying a "watch and wait" policy.

Re-treatment would be the most appropriate course of action if the lesion were present on the deep margins, aggressive histologically, in a critical anatomical site or previously a recurrence.

Bieley et al in their study demonstrated residual tumour in 55% of cases where BCC was incompletely excised.

Liu et al used radiotherapy to treat lesions with residual tumour demonstrating a 91% 5-year survival rate.

2. **Non-Surgical**

- **Radiotherapy**

**BCC**

This is a useful mode of treatment for BCC barring those that are so large that the radiation dose approaches the maximum tissue tolerance. A review of studies since 1947 showed a 5-year survival rate of 91.3% for primary lesions and 90.2% for recurrences.
SCC
In the treatment of SCC it may be appropriate for lesions in the elderly and/or those with contraindications to anaesthesia or surgery. The dosage is 4000Gy given in 5-16 fractions.

There are certain cases where radiotherapy is not appropriate:
- Lesions previously treated with radiotherapy
- Young patients: increased incidence of late onset cutaneous atrophy and telangiectasia.
- Lesions close to the eye: increased incidence of epiphora and ectropion plus cataract formation\textsuperscript{20}.
- Verrucous carcinoma

Some suggest that recurrent BCC previously radiated behave more aggressively but this may simply reflect that the lesion was highly infiltrative and aggressive from the start\textsuperscript{15}.

Treatments under Investigation
There are some treatment modalities that are still under investigation but are mentioned along with references to preliminary work that has been done.
These include:
- Topical therapy with 5-fluorocytosine. This is used primarily in the treatment of the pre-malignant condition, Bowens disease.
- Intralosional interferon\textsuperscript{22}.
- Photodynamic therapy\textsuperscript{23}.
- Retinoids\textsuperscript{24}.
- Chemotherapy\textsuperscript{25}.

Metastases
Cervical metastatic disease from these tumours is rare but is a much commoner occurrence in SCC (0.3 - 13.7%) than in BCC (0.0028 - 0.4%)\textsuperscript{4}.

Certain high-risk lesions predisposing to metastatic spread include those > 2cm in diameter and / or > 6mm thickness, perineural/vascular invasion, local recurrence, poor tumour differentiation, invasion of auricular cartilage and invasive lip disease plus involvement of the commissure or upper lip\textsuperscript{26}. Parotid nodal metastases arise from cutaneous malignancies more than from any other cancer and, when present, demonstrate a poor 5-year survival rate of 27.8% if single modality treatment is used\textsuperscript{26}. Cassisi et al\textsuperscript{27} showed that the majority of parotid metastases occurred within 1 year of treatment of the primary tumour and it was controlled in 89% of cases with a combination of surgery and radiotherapy to the parotid region plus neck\textsuperscript{27}.

Follow-up
Metastases to the neck in facial SCC are relatively low as shown previously in this paper. Tavin et al\textsuperscript{28} and Netterville et al\textsuperscript{29} showed that primary cutaneous tumours in the preauricular region and cheek tended to metastasise most frequently of all the facial SCC.

The average time to development of the delayed metastases was 19 months\textsuperscript{28} and 11 months\textsuperscript{29}. It would thus seem reasonable to follow these cases for 3 years post treatment.
The follow-up regime for BCC is different to that for SCC, as recurrences are not as common. The majority of single, nodular or superficial lesions are followed up once post treatment to ensure that the site of excision has healed with a cosmetically acceptable result.
However, patients considered high risk (as previously discussed) plus patients presenting with multiple BCC are followed routinely for 5 years as studies show that over 80% of recurrences will present within 5 years of treatment.30

References:
Fitzpatrick Skin Type Quiz

This information will help our office to better evaluate your skin type so the laser treatment will be more effective. Skin type is often categorised according to the Fitzpatrick skin type scale which ranges from very fair (skin type I) to very dark (skin type VI). The two main factors that influence skin type and the treatment program devised by your practitioner are:

- Genetic disposition
- Reaction to sun exposure and tanning habits

Skin type is determined genetically and is one of the many aspects of your overall appearance, which also includes the colour of your eyes, hair, etc. The way your skin responds to sun exposure is another way of correctly assessing your skin type. Recent tanning, whether by the sun or an artificial tanning booth, even tanning creams, can have a major impact on your skin colour evaluation. By using the information you provide on this form, we can be better prepared to provide you with the best care. Please take a few minutes to fill out this questionnaire.

Mark 0 through 4 for each question.

Genetic Disposition

<table>
<thead>
<tr>
<th>Score</th>
<th>0</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
</tr>
</thead>
<tbody>
<tr>
<td>Your eye colour?</td>
<td>Light blue, Grey, green</td>
<td>Blue, grey or green</td>
<td>Blue</td>
<td>Dark brown</td>
<td>Brownish black</td>
</tr>
<tr>
<td>Natural colour of your hair?</td>
<td>Sandy, red</td>
<td>Blond</td>
<td>Chestnut, dark blond</td>
<td>Dark brown</td>
<td>Brownish black</td>
</tr>
<tr>
<td>Colour of your skin non-exposed?</td>
<td>Reddish</td>
<td>Very pale</td>
<td>Pale with beige tint</td>
<td>Light brown</td>
<td>Dark brown</td>
</tr>
<tr>
<td>Do you have freckles on unexposed areas?</td>
<td>Many</td>
<td>Several</td>
<td>Few</td>
<td>Incidental</td>
<td>None</td>
</tr>
</tbody>
</table>

Total score for genetic disposition: _____
### Reaction to Sun Exposure

<table>
<thead>
<tr>
<th>Score</th>
<th>0</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
</tr>
</thead>
<tbody>
<tr>
<td>What happens when you stay too long in the sun?</td>
<td>Painful redness, blistering, peeling</td>
<td>Blistering, followed by peeling</td>
<td>Burns sometimes, followed by peeling</td>
<td>Rarely burns</td>
<td>Never burns</td>
</tr>
<tr>
<td>To what degree do you turn brown?</td>
<td>Hardly or not at all</td>
<td>Light colour tan</td>
<td>Reasonable tan</td>
<td>Tan very easily</td>
<td>Turn dark brown quickly</td>
</tr>
<tr>
<td>Do you turn brown within several hours after sun exposure?</td>
<td>Never</td>
<td>Seldom</td>
<td>Sometimes</td>
<td>Often</td>
<td>Always</td>
</tr>
<tr>
<td>How does your face react to the sun?</td>
<td>Very sensitive</td>
<td>Sensitive</td>
<td>Normal</td>
<td>Very resistant</td>
<td>Never had a problem</td>
</tr>
</tbody>
</table>

**Total score for reaction to sun exposure:**

### Tanning Habits

<table>
<thead>
<tr>
<th>Score</th>
<th>0</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
</tr>
</thead>
<tbody>
<tr>
<td>When did you last expose your body to sun or tanning booth/cream?</td>
<td>More than 3 months ago</td>
<td>2-3 months ago</td>
<td>1-2 months ago</td>
<td>Less than one month ago</td>
<td>Less than 2 weeks ago</td>
</tr>
<tr>
<td>Did you expose the area to be treated to the sun?</td>
<td>Never</td>
<td>Hardly ever</td>
<td>Sometimes</td>
<td>Often</td>
<td>Always</td>
</tr>
</tbody>
</table>

**Total score for tanning habits:**

### Summary

Add up the total scores for each section for your Skin Type Score to give you a better evaluation of your skin type.

**Total score for Genetic Disposition**

**Total score for Reaction to Sun Exposure**

**Total score for Tanning Habits**

**SKIN TYPE SCORE**
Your Fitzpatrick Skin Type:

<table>
<thead>
<tr>
<th>Skin Type Score</th>
<th>Fitzpatrick Skin Type</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-7</td>
<td>I</td>
</tr>
<tr>
<td>8-16</td>
<td>II</td>
</tr>
<tr>
<td>17-25</td>
<td>III</td>
</tr>
<tr>
<td>25-30</td>
<td>IV</td>
</tr>
<tr>
<td>Over 30</td>
<td>V-VI</td>
</tr>
</tbody>
</table>

Name: __________________________________________________ Date: _____________________

Which of the following best describes your skin type?  Please circle one:

1- always burn, never tan
2- always burn, sometimes tan
3- sometimes burn, tan somewhat
3- rarely burn, tan with ease
4- moderately pigmented, tans very easily
5- deeply pigmented, never burn

Ethnic background is of importance when considering skin colour and laser hair removal. If known what is your ethnic background:

________________________________________________________