Ear and Temporal Bone Tumours Histopathology Reporting Guide

ICCR

Family/Last name	
Given name(s)	

Date of birth: DD – MM – YYYY

Patient identifiers:

Date of request: DD – MM – YYYY

Accession/Laboratory number:

Elements in **black text** are CORE. Elements in grey text are NON-CORE.
☐ indicates multi-select values   ◯ indicates single select values

SCOPE OF THIS DATASET

CLINICAL INFORMATION (Note 1)

◯ Information not provided
◯ Information provided (select all that apply)
 ▼ Previous therapy
  ☐ Surgery
  ☐ Chemotherapy
  ☐ Radiotherapy
  ▼ Targeted therapy, *specify if available*

  ▼ Immunotherapy, *specify if available*

 ☐ Other clinical information, *specify*

OPERATIVE PROCEDURE (select all that apply) (Note 2)

◯ Not specified
◯ Biopsy (excisional, incisional, core needle (diagnostic sampling)), *specify*

 ☐ Resection
  ☐ Sleeve resection: cartilaginous portion of ear canal, and skin of the external auditory canal (EAC), and ear drum
  ☐ Limited temporal bone resection: removal of mastoid bone, preserving bony EAC
  ☐ Lateral temporal bone (LTB): removal of the temporal bone including ear canal, middle ear, mastoid
  ☐ Subtotal temporal bone resection (STBR): removal of the temporal bone as stated in LTB, and inner ear structure
  ☐ Total temporal bone resection (TTBR): removal of the temporal bone as stated in STBR, and petrous apex
 ☐ Parotidectomy
 ☐ Neck (lymph node) dissection,[a] *specify*

 ☐ Other, *specify including laterality*

SPECIMEN(S) SUBMITTED (select all that apply) (Note 3)

◯ Not specified
◯ Biopsy only
☐ Sleeve resection of temporal bone
☐ Lateral temporal bone
☐ Middle ear contents
☐ Subtotal temporal bone resection
☐ Partial mastoidectomy with middle ear contents
☐ Radical mastoidectomy
☐ Pinna
☐ Parotidectomy
☐ Temporomandibular joint (TMJ)
☐ Dura
 ◯ Temporal fossa
 ◯ Posterior fossa
☐ Neck (lymph node) dissection,[a] *specify*

☐ Other, *specify including laterality*

TUMOUR SITE (select all that apply) (Note 4)

◯ Not specified
☐ External auditory canal (EAC)
☐ Middle ear
☐ Temporal bone (including mastoid, petrous)
☐ Inner ear
☐ Other, *specify*

TUMOUR LATERALITY

◯ Not specified
◯ Left
◯ Right

[a] *If a neck (lymph node) dissection is submitted, then a separate dataset is used to record the information.*

TUMOUR FOCALITY (Note 5)

- ◯ Unifocal
- ◯ Bilateral
- ◯ Multifocal
  - Specify number of tumours [   ]

TUMOUR DIMENSIONS (Note 6)

Maximum tumour dimension (largest tumour)
(pathology and/or imaging determination)

[   ] mm

Additional dimensions (largest tumour)

[   ] mm x [   ] mm

BLOCK IDENTIFICATION KEY (Note 7)
(List overleaf or separately with an indication of the nature and origin of all tissue blocks)

HISTOLOGICAL TUMOUR TYPE (select all that apply) (Note 8)
(Value list based on the World Health Organization Classification of Head and Neck Tumours (2024))

External auditory canal

- ☐ Ceruminous adenoma
- ☐ Ceruminous adenocarcinoma
  - ☐ Adenoid cystic
  - ☐ Mucoepidermoid
  - ☐ NOS
- ☐ Squamous cell carcinoma

Middle and inner ear

- ☐ Middle ear papilloma
- ☐ Vestibular schwannoma
- ☐ Middle ear neuroendocrine tumour
- ☐ Endolymphatic sac tumour
- ☐ Squamous cell carcinoma of the middle ear
- ☐ Middle ear adenocarcinoma

Other, *specify*

[   ]

HISTOLOGICAL TUMOUR GRADE[b] (Note 9)
(Applicable to squamous cell carcinoma and neuroendocrine tumours only)

- ◯ Grade 1, well differentiated, low grade
- ◯ Grade 2, moderately differentiated, intermediate grade
- ◯ Grade 3, poorly differentiated, high grade
- ◯ Undifferentiated
- ◯ High grade transformation

Grading system used, *specify*

[   ]

- ◯ Cannot be assessed, *specify*

[   ]

[b] *Grading of neuroendocrine tumours is non-core. Use only Grade 1, 2 and 3 for neuroendocrine tumours; neuroendocrine carcinomas are considered high grade by definition and are therefore not graded.*

EXTENT OF INVASION (Note 10)

- ◯ Not identified
- ◯ Present (select all that apply)
  - ☐ Clinical observation and/or imaging   ☐ Histologic

  - ☐ Skin involvement
  - ☐ Bone/cartilage (mastoid bone)
    - ◯ Superficial cortical involvement
    - ◯ Full thickness bone involvement
  - ☐ Dura involvement
  - ☐ Brain involvement
  - ☐ Temporomandibular joint (TMJ) involvement
  - ☐ Parotid gland involvement
  - ☐ Soft tissue involvement
    - ◯ ≤5 mm
    - ◯ >5 mm
    - ◯ Indeterminate, *specify reason*

    [   ]

  - ☐ Other, *specify*

    [   ]

  - ◯ Cannot be assessed, *specify*

    [   ]

LYMPHOVASCULAR INVASION (Note 11)

- ◯ Not identified
- ◯ Present, *specify if named* (select all that apply)
  - ☐ Internal jugular vein
  - ☐ Carotid artery
- ◯ Indeterminate, *specify reason*

[   ]

PERINEURAL INVASION (Note 12)

- ◯ Not identified
- ◯ Present, *specify nerve if possible (e.g., facial nerve, tympanic nerve, glossopharyngeal nerve, lesser petrosal nerve, greater petrosal nerve)*

[   ]

- ◯ Indeterminate, *specify reason*

[   ]

MARGIN STATUS (Note 13)

(Applicable for squamous cell carcinoma and adenocarcinoma only)

○ Not involved by invasive carcinoma

▼

　Distance of tumour from closest margin ☐ mm

　○ Distance not assessable

　Specify closest margin(s), if possible (select all that apply)

　☐ Skin
　☐ Bone
　☐ Soft tissue
　☐ Parotid gland

○ Involved by invasive carcinoma

▼ Specify margin(s), if possible

　☐

○ Cannot be assessed, *specify*

▼

　☐

COEXISTENT PATHOLOGY (select all that apply) (Note 14)

○ None identified
☐ Cholesterol granuloma
☐ Cholesteatoma
☐ Osteomyelitis (acute, chronic)
☐ Papilloma
☐ Other, *specify*

☐

ANCILLARY STUDIES (Note 15)

Neuroendocrine neoplasms (select all that apply)

○ Not applicable
☐ Neuroendocrine markers, *specify*

▼ ☐

☐ Cytokeratin(s), *specify*

▼ ☐

☐ Ki-67 proliferation index ☐ %

☐ Rb

▼
　☐ Retained
　☐ Deficient

☐ p53

▼
　○ Abnormal, *specify*

　☐

☐ Other, *record test(s), methodology and results*

☐

Other tumours

○ Not performed
○ Performed, *record test(s), methodology and results*

▼

☐

Representative blocks for ancillary studies, *specify those blocks best representing tumour and/or normal tissue for further study*

☐

PATHOLOGICAL STAGING (Note 16)

TNM Descriptors (only if applicable) (select all that apply)

☐ m - multiple primary tumours
☐ r - recurrent
☐ y - during or following multimodality therapy

Primary tumour (pT)[c]

○ Not applicable

○ T1　Tumour limited to the external auditory canal (EAC) without bony erosion or evidence of soft tissue involvement

○ T2　Tumour with limited EAC bone erosion (not full thickness) or limited (≤5 mm) soft tissue involvement

○ T3　Tumour eroding the osseous EAC (full thickness) with limited (≤5 mm) soft tissue involvement, or tumour involving the middle ear and/or mastoid

○ T4　Tumour eroding the cochlea, petrous apex, medial wall of the middle ear, carotid canal, jugular foramen, or dura, or with extensive soft tissue involvement (>5 mm), such as involvement of temporomandibular joint (TMJ) or styloid process, or evidence of facial paresis

[c] *Note that the results of neck (lymph node) dissection are derived from a separate dataset.*

Definitions

CORE elements

CORE elements are those which are essential for the clinical management, staging or prognosis of the cancer. These elements will either have evidentiary support at Level III-2 or above (based on prognostic factors in the National Health and Medical Research Council (NHMRC) levels of evidence[1]). In rare circumstances, where level III-2 evidence is not available an element may be made a CORE element where there is unanimous agreement by the Dataset Authoring Committee (DAC). An appropriate staging system e.g., Pathological TNM staging would normally be included as a CORE element.

Molecular and immunohistochemical testing is a growing feature of cancer reporting. However, in many parts of the world this type of testing is limited by the available resources. In order to encourage the global adoption of ancillary tests for patient benefit, International Collaboration on Cancer Reporting (ICCR) includes the most relevant ancillary testing in ICCR Datasets as CORE elements, especially when they are necessary for the diagnosis. Where the technical capability does not yet exist, laboratories may consider temporarily using these data elements as NON-CORE items.

The summation of all CORE elements is considered to be the minimum reporting standard for a specific cancer.

NON-CORE elements

NON-CORE elements are those which are unanimously agreed should be included in the dataset but are not supported by level III-2 evidence. These elements may be clinically important and recommended as good practice but are not yet validated or regularly used in patient management.

Key information other than that which is essential for clinical management, staging or prognosis of the cancer such as macroscopic observations and interpretation, which are fundamental to the histological diagnosis and conclusion e.g., macroscopic tumour details, may be included as either CORE or NON-CORE elements by consensus of the DAC.

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Scope

The dataset has been developed for the reporting of resection and biopsy specimens of the ear and temporal bone. It includes only primary tumours of the external auditory canal, middle and inner ear, including both benign and malignant entities (specifically due to anatomic confines and management alternatives which may require significant, destructive or disfiguring surgery).

By definition, all malignancies of the external ear (pinna, concha, scaphoid, lobe, etc., such as squamous cell carcinoma (SCC), basal cell carcinoma, pleomorphic dermal sarcoma, Merkel cell carcinoma and melanoma) are separately covered by the ICCR Skin datasets.[2] Primary parotid gland malignancies with direct extension in to the ear canal are excluded; the ICCR Carcinomas of the major salivary glands dataset should be used.[3] Haematolymphoid neoplasms are also excluded from this dataset.

Neck dissections and nodal excisions are dealt with in a separate dataset, and this dataset should be used in conjunction, where applicable.[4]

For bilateral tumours, a separate dataset should be completed for each tumour.

This dataset is intended for use for primary tumour resections. For resections of recurrent disease, the reporting guide may be used pragmatically although some data elements may be not applicable nor assessable.

The second edition of this dataset includes changes to align the dataset with the World Health Organization (WHO) Classification of Head and Neck Tumours, 5th edition, 2023.[5]

There is very limited evidence supporting the various core and non-core elements in this dataset. All elements have thus been included by unanimous consensus of the DAC.

A list of changes in this dataset edition can be accessed here.

The authors of this dataset can be accessed here.

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Note 1 – Clinical information (Core and Non-core)

In the context of a recurrent tumour, patients may have had previous surgery, radiotherapy, chemotherapy or immunotherapy. Diagnostic procedures may be performed, followed by various cytoreduction therapies. As such, a core needle biopsy, incisional biopsy or even debulking surgery may have been performed, and would be documented as 'surgery' in this context. Information regarding previous treatment should be recorded.

Currently, neoadjuvant therapy including radiotherapy, chemotherapy or immunotherapy is not deployed in the tumours of the ear and temporal bone. However, it will be wise to include this field for future utilisation and harmonisation across the suite of structured reporting protocols as the treatment paradigms evolve. Of note, criteria for assessment of pathological response will also need to be developed.

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Note 2 – Operative procedure (Core)

The anatomy and surgical interventions of the ear and temporal bone are complex,[6,7] with unfamiliar terminology frequently used (see Figure 1). Thus, it is absolutely critical to maintain open communication with the treating surgeon, oncologist, dermatologist and radiologist with respect to exact anatomic site of involvement, tumour laterality, and specific operative procedures or landmarks identified to yield the most accurate information.[8-11]

There is no internationally standardised terminology to describe the type and extent of temporal bone resection. For this version of the ICCR Ear and temporal bone dataset, the dataset authors have adopted widely used terminologies to describe the various types of temporal bone resection, in progressively more radical procedures. These include:

1. Sleeve resection: The tumour is completely resected with or without the cartilaginous part of the external ear canal (EAC). The bony EAC and the tympanic membrane (TM) are preserved. This type of resection is suitable for very superficial malignancy (or in situ disease) limited to the skin of the EAC.
2. Limited (or cortical) temporal bone resection: In this procedure the mastoid bone is removed while the EAC structure and the TM are preserved. This surgery is generally used for tumours involving

only the temporal bone, sparing the EAC and middle ear structures. It can also be used for tumours involving the skin posterior to the auricle, where a concurrent resection of the skin is performed.

3. Lateral temporal bone resection: In this procedure, the EAC, the TM, the middle ear structures and the mastoid are removed. The inner ear is preserved. This procedure is generally performed for tumour that involve the entire EAC where removal of the TM is necessary.

4. Subtotal temporal bone resection: In this procedure, the temporal bone, the EAC, TM, the middle ear structure and the inner ear are removed. Often, the facial nerve will need to be either re-routed or sacrificed. Depending on the extent of tumour, a concurrent resection of the posterior fossa dura or the temporal lobe dura might be required for disease clearance.

5. Total temporal bone resection: In this procedure, the temporal bone, EAC, TM/middle ear, the inner ear, as well as the petrous apex are removed. Often, ligation of the carotid artery will be required.

Any of these procedures may be accompanied by a parotidectomy and/or neck dissection.

Occasionally, the subtotal and total temporal bone resection may be accompanied by dura and brain parenchyma. Often, the specimens may be submitted for histopathological assessment in multiple parts and/or fragments. Communication with the surgeon and correlation with radiology is important for assessing the extent of invasion and particularly for margin assessment.
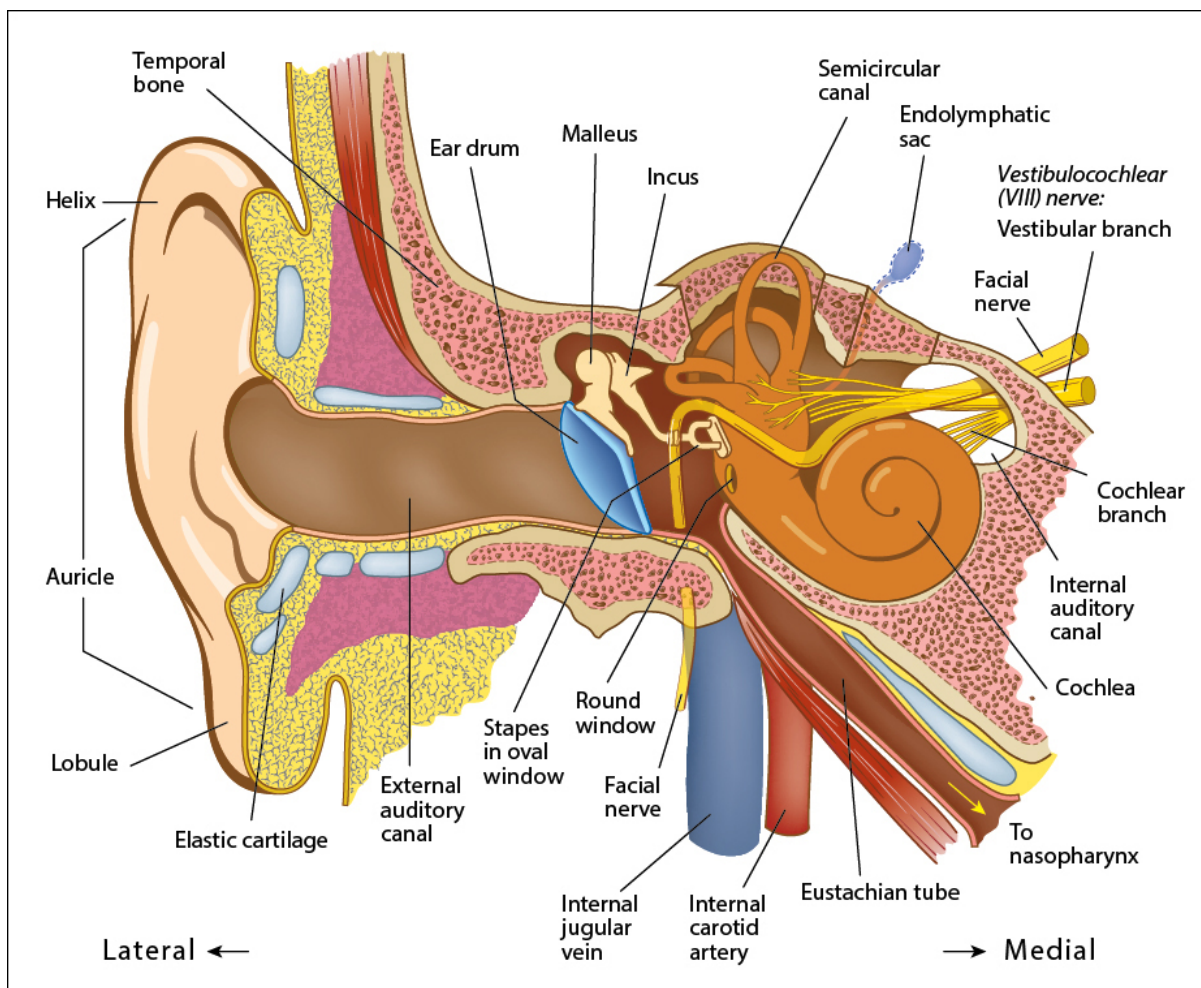

Figure 1: Diagram of ear and temporal bone anatomic landmarks.
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Note 3 – Specimen(s) submitted (Core)

In light of the complex anatomy and often unfamiliar surgical interventions of the ear and temporal bone, it is imperative to obtain information about the exact anatomic site of involvement, tumour laterality, and specific operative procedures or landmarks identified to yield the most accurate information.[12,13]

'Not specified' should be used rarely and only after good faith effort has been employed to obtain the requisite information.

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Note 4 – Tumour site (Core)

It is important to document the exact site of the tumour, as tumour location is correlated with patient outcome. As an example, patients with middle ear SCCs have a worse outcome than patients with SCC of the external auditory canal.[6,8,10,14-18]

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Note 5 – Tumour focality (Non-core)

The identification of bilateral tumours, especially in the setting of endolymphatic sac tumours,[19,20] paraganglioma,[21,22] acoustic/vestibular Schwannoma,[23] and meningioma,[23] increases the potential discovery of genetic tumour syndrome associated disease.

For bilateral or multifocal tumours, a separate dataset should be completed for each tumour.

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Note 6 – Tumour dimensions (Non-core)

The single greatest tumour dimension, using macroscopic and/or microscopic measurements, should be used to determine the most accurate extent of tumour. In biopsy samples, it may be underestimated. Thus, to be as thorough as possible, the documentation of the tumour dimension may require additional clinical/operative or imaging information to yield this value. However, as tumour size is not used in staging, this element is non-core. While tumour size is a non-core element, the extent of invasion contributes to the proposed staging system with 5 millimetres (mm) as a threshold with upstaging of tumours invading the soft tissues >5 mm (see **Note 10 – EXTENT OF INVASION**).

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Note 7 – Block identification key (Non-core)

The origin/designation of all tissue blocks should be recorded. This information should ideally be documented in the final pathology report and is particularly important should the need for internal or external review arise. The reviewer needs to be clear about the origin of each block in order to provide an informed specialist opinion. If this information is not included in the final pathology report, it should be

available on the laboratory computer system and relayed to the reviewing pathologist. It may be useful to have a digital image of the specimen and record of the origin of the tumour blocks in some cases.

Recording the origin/designation of tissue blocks also facilitates retrieval of blocks for further immunohistochemical or molecular analysis, research studies or clinical trials.

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Note 8 – Histological tumour type (Core)

Histologic diagnosis of ear and temporal bone tumours is based on the 2023 WHO Classification of Head and Neck Tumours, 5th edition (Table 1).[5,24] Neuroendocrine tumours (NET) (formerly middle ear adenoma) are also included.[25-27] While neuroendocrine carcinomas (NEC) are recognised, they are not documented yet as primary tumours in this anatomic site.

The types of ear and temporal bone primary tumours are limited. Few cases have been reported for several specific tumour categories, and thus prognostication about each specific tumour type is limited, at best. Overall, the most common tumour type is SCC, and it is known to have the worst patient outcome.[5,28-33] When adenoid cystic carcinoma and mucoepidermoid carcinoma are the ceruminous adenocarcinoma type, parotid gland evaluation is recommended to exclude origin from the parotid gland with secondary invasion into the external canal.[34,35]

Table 1: World Health Organization classification of tumours of the ear and temporal bone.[5]

Descriptor	ICD-O codes[a]
External auditory canal	
Ceruminous adenoma	8420/0
Squamous cell carcinoma of the external auditory canal	8070/3
Ceruminous adenocarcinoma	8420/3
Ceruminous adenoid cystic carcinoma	8420/3
Ceruminous mucoepidermoid carcinoma	8420/3
Middle ear	
Meningioma	9530/0
Middle ear papilloma	8121/0
Middle ear neuroendocrine tumour	8240/3
Squamous cell carcinoma of the middle ear	8070/3
Middle ear adenocarcinoma	8260/3
Inner ear	
Endolymphatic sac tumour	8140/3
Vestibular schwannoma	9560/0

[a] These morphology codes are from the International Classification of Diseases for Oncology, third edition, second revision (ICD-O-3.2).[36] Behaviour is coded /0 for benign tumours; /1 for unspecified, borderline, or uncertain behaviour; /2 for carcinoma in situ and grade Ill intraepithelial neoplasia; /3 for malignant tumours, primary site; and /6 for malignant tumours, metastatic site. Behaviour code /6 is not generally used by cancer registries.

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Note 9 – Histological tumour grade (Core)

Generally, histologic tumour grades are applied to SCC or NETs only (core). Ceruminous carcinomas (adenoid cystic and mucoepidermoid carcinoma exclusively) can be graded based on salivary gland primaries, but no grading is accepted for ceruminous adenocarcinoma, not otherwise specified (NOS). Poorly differentiated tumours indicate a poor patient survival.[37]

The same grading of central nervous system meningiomas is applied to ear and temporal bone, realising that >95% are WHO grade 1 tumours.[38,39]

A three tiered grading system has been proposed for NETs (non-core),[5] utilising proliferation index/mitoses to aid separation, similar to NETs of the larynx, lung and gastrointestinal tract. In general, a Ki-67 proliferation index <2% is grade I, 2-20%, grade 2, and >20% grade 3, although case numbers are small.[40] NECs are considered high grade by definition and are therefore not graded, although not yet documented as a primary tumour in this site.

Other tumour types for the most part do not have tiered grading systems (such as ceruminous adenocarcinoma, NOS, endolymphatic sac tumours, schwannoma or middle ear adenocarcinoma).

Still, several grading systems are available, with differing merits, and as such, recording which system has been applied is more clinically meaningfully (use 'specify' to state the system used), with the ICCR deferring to the WHO Classification current edition for grading guidance and preference.[5]

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Note 10 – Extent of invasion (Core)

The extent of invasion contributes to the proposed staging system and all attempts should be made to measure this accurately. Infiltration of the soft tissues by the tumour >5 mm indicates high stage (defined by measuring tumour involving submucosal stroma/soft tissues below the inter-rete basement membrane).

If there is involvement of any of these recognised structures, documentation will provide prognosis and management information.[38,41] For example, patients with primary ear and temporal bone carcinoma with parotid gland involvement have a worse prognosis than patients without parotid gland involvement.[34] If there is advanced disease clinically, then parotid gland resection is generally recommended.[34] The macroscopic and microscopic extent of tumour frequently overlap. Thus, invasion 'microscopically' into any of these structures is for the most part not recognised, unless the part is specifically stated to be from the site. Thus, on histologic examination, you may not recognise the specific structure. Therefore, correlation between macroscopic and microscopic findings is encouraged to yield the most meaningful findings.[5,11,30,35,42] As an example, patients who exhibit dura involvement, will have a worse patient outcome.[5,43] The extent of invasion may need to be evaluated by imaging or during intraoperative assessment, as histologic identification of these structures may not be feasible.

Similarly, when there is destructive cartilage and/or bone invasion, the patients tend to have a worse prognosis.[28,29,38,42-45] Bone and/or cartilage invasion may be a macroscopic feature, sometimes not seen on histology sections due to the nature of the clinical sampling performed, and so clinical/imaging observation and/or histologic evidence may be used to support the interpretation. However, it is recommended that a histologic section through the involved bone should be performed to obtain histologic evidence of the extent of bone and/or cartilage involvement (partial versus full thickness involvement). In general, stage correlates with bone and/or cartilage invasion, with high stage patients more frequently showing bone invasion than

low stage patients.[8] Further, patients with bone and/or cartilage invasion will usually have a worse prognosis and require more extensive treatment than patients without bone invasion.[42,46]

Due to the type of samples, tumour budding or tentacular pattern of invasion may not be histologically identified. However, if this type of growth is seen in SCC, patients tend to have a shorter survival.[37,46,47]

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Note 11 – Lymphovascular invasion (Core)

By inference and consensus of the DAC, lymphovascular invasion is considered to be associated with a worse clinical outcome. However, in ear and temporal bone tumours, this finding has not been independently evaluated in prospective or prognostic studies due to low cases numbers at any given referral centre.

Cases that are still equivocal after taking additional steps may be reported as 'indeterminate' for lymphovascular invasion, but this designation should be sparingly used and it is useful to provide the reason in a comment in the report.

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Note 12 – Perineural invasion (Core)

Patients who manifest perineural/intraneural invasion, especially if it is identified in large or named nerves (such as facial nerve and chorda tympani nerve), have a worse clinical outcome, irrespective of the tumour type or tumour grade.[47,48] If the biopsy is very small with only tumour included, it may be prudent to use 'cannot be assessed' in order to alert the clinician that perineural invasion cannot be reliably excluded in the sampled material.

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Note 13 – Margin status (Core and Non-core)

Margin status is reported for SCC and adenocarcinoma specifically, rather than for the benign or intermediate neoplasms (i.e., ceruminous adenoma, endolymphatic sac tumour, middle ear papilloma) or NETs. The best overall outcomes for tumours of ear and temporal bone are achieved when margins are negative. In general, mucosal/epithelial margins are reported, but bone and soft tissue margins carry similar prognostic value, and thus should also be reported, especially as the deep margins (bone and soft tissue) are often more clinically significant than superficial margins (skin). Tumours which are meticulously debulked have the best long term outcome.[11,14,29,31,43,49-54]

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Note 14 – Coexistent pathology (Non-core)

Management may be complicated by coexistent pathology. Patient with otitis media generally show a poor survival,[11] but if there is acute or chronic osteomyelitis, options for radiation and chemotherapy may be limited.[55,56]

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Note 15 – Ancillary studies (Core and Non-core)

For neuroendocrine neoplasms core elements are neuroendocrine markers, epithelial markers, and Ki-67 proliferation index. The diagnosis of neuroendocrine neoplasms (specifically NETs and NECs) must be confirmed immunohistochemically, with positive reaction for neuroendocrine markers (synaptophysin, chromogranin, INSM1), and for epithelial markers (pancytokeratin, cytokeratin). Furthermore, a proliferation index as determined by Ki-67 immunohistochemical analysis is recommended for grading all NETs, helping to confirm NECs, and p53 and Rb1 may be helpful in the distinction between NET and NEC, especially G3 NET from NEC.[25,57,58]

In most patients, further studies are not required for the diagnosis. However, additional molecular testing may be of benefit, especially in syndrome associated, bilateral, or uncommon tumour presentations. Translocation associated malignancies (*BRD4::NUTM1, DEK::AFF2, EWSR1::FLI1, EWSR1::ERG*) should also be considered in the morphologic context of poorly differentiated non keratinising SCC.[59-62]

Immunohistochemistry may also be considered to differentiate between tumour types especially in limited sampling or tumours affected by distortional changes. Ancillary tests rarely may be required to identify the primary site of metastatic disease.

Programmed cell death-ligand 1 (PD-L1) expression has been used as predictive biomarker for checkpoint inhibitor therapy since the anti-programmed cell death-1 receptor (PD-1) antibodies, nivolumab and pembrolizumab, have been approved for the treatment of patients with recurrent and/or unresectable metastatic head and neck SCC,[63-66] with various cutoffs of expression associated with betters responses, although not in all patients.[67]

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Note 16 – Pathological staging (Core)

There is no standardised staging system for this anatomic site, although it has been suggested by several groups.[13,68-70] However, staging is still of value in standardising therapy for these various unusual tumours. The pathological staging presented on the reporting guide is adapted from the Pittsburgh system,[69] and based on a consensus of the DAC. The T staging is most significant for SCCs and for salivary gland-type tumours, particularly of the external auditory canal and middle ear.[14,15,29,43,71-75]

Pathological staging has not been well developed for middle ear and inner ear tumours (i.e., endolymphatic sac tumour), where clinical staging may be more appropriate.[69] In inner ear cases, it is probably more important to make certain that a clinical stage is accurately determined, than necessarily being definitive about a pathological stage. As such, benign tumours and NETs do not require staging, although the reporting guide can be used to aid treatment and outcome prognostication. The studies used as a guide are

retrospective where the patient outcomes were not available, primarily used as a guide for therapy rather than prognosis.

Overall, there is a poor prognosis when lymph node metastases are identified, correlating to advanced stage, whether in the cervical lymph nodes or those of the parotid gland parenchyma.[11,15,35,75-77]

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