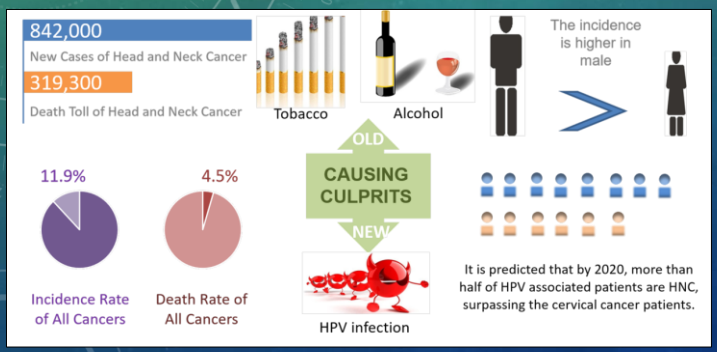
# 2020 UPDATES TO NEOPLASMS OF THE ORAL CAVITY, OROPHARYNX, AND THE MAJOR AND MINOR SALIVARY GLANDS




FCDS 2020-2021 WEBINAR SERIES

STEVEN PEACE, CTR

DECEMBER 17, 2020

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## CDC & FLORIDA DOH ATTRIBUTION

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FLccSC
Fundamental Learning Collaborative for the Cancer Surveillance Community

# FLCCSC LMS – CEU QUIZ –FCDS IDEA

- Attendees must take and pass a 3-5 question CEU Quiz to get CEUs
- CEU Awards are Restricted to Attendees with a FLccSC LMS Account
- The CEU Quiz will be posted to FLccSC 1-2 hours after the webcast ends
- Only registered FLccSC Users will be given access to the CEU Quiz
- Florida attendees must have a Florida FLccSC Account to take the Quiz
- South Carolina attendees must have a South Carolina FLccSC Account
- New FLccSC States will follow similar instructions for the CEU Quiz
- Attendees can attend any of the live webcasts without receiving CEUs
- Recorded Sessions are also available for non-FLccSC Users – No CEUs

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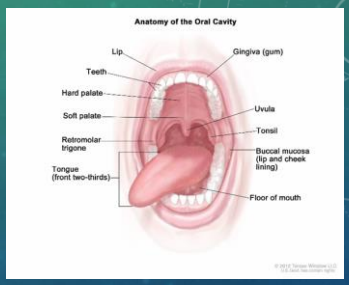
# PRESENTATION OUTLINE




- Welcome
- Introduction to Oral Cavity and Oropharynx
- Introduction to Salivary Glands (Major and Minor)
- Introduction to Cervical Lymph Node Regions
- Risk Factors for Oral Cavity and Oropharynx Cancers
- Risk Factors for Salivary Gland Cancers & Assessing Risk for All
- Types of Cancers in the Oral Cavity, Oropharynx and Salivary Glands
- Highlights from the 2021 Head and Neck Solid Tumor Rules – MP/H Rules
- Locations and Indications for when a Head and Neck Cancer is Likely a Metastatic Skin Cancer
- Coding and Documenting HPV and p16 // Using Histology Code 8085 and 8086
- Oral Cavity and Oropharynx Self-Examination and Dental or Oropharyngeal MD Screening
- Unknown Head and Neck Primary – Understanding the Rationale & Criteria to Abstract these Cases
- Staging Head and Neck Cancers – Tips for AJCC TNM, SEER Summary Stage 2000 and SSDIs
- New and Emerging Tumor Markers and Molecular Genetic Testing for Targeting Treatments
- Other Emerging Treatments and Techniques
- Questions

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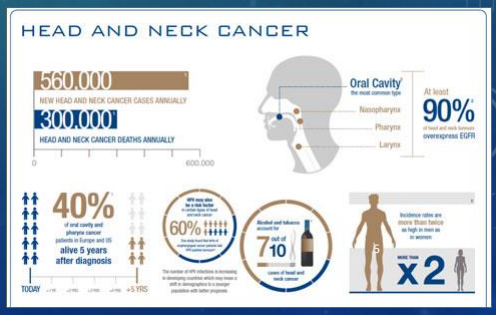
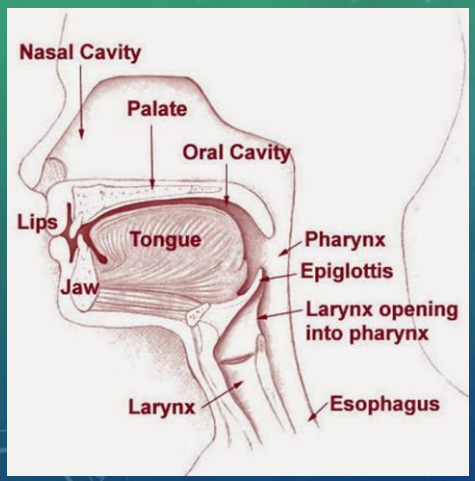
# LIMITATIONS OF THIS PRESENTATION

- We will discuss in detail the anatomy of the oral cavity, oropharynx and salivary glands.
- We will discuss cancers of oral cavity, oropharynx, and salivary glands.
- We will discuss the anatomy and function of cervical lymph node chains.
- We will discuss risk factors and screening for oral cavity and oropharyngeal cancers.
- We will show lots of pictures to help describe 'extension' from one site to another.

- We will not discuss ALL head and neck anatomic locations.
- We will not discuss anatomy of nasopharynx or hypopharynx.
- We will not discuss cancers of the larynx.
- We will not discuss cancers of the thyroid.
- We will not discuss cancers of the Brain or CNS.

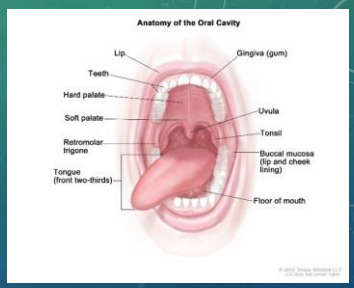



# INTRODUCTION TO ORAL CAVITY AND OROPHARYNX



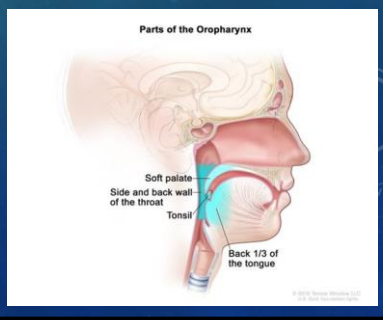

**ORAL CAVITY**

- Mouth, Lips, Inside the Cheeks and Lips, front 2/3 of tongue, upper gums, lower gums, floor of mouth under the tongue, roof of mouth, soft palate, behind wisdom




**OROPHARYNX**

- Base of the tongue, pharyngoepiglottic folds and the glossoepiglottic folds.
- Vallecula.
- Tonsillar region, which includes the fossa and the anterior and posterior pillars.
- Soft palate, which includes the uvula.
- Posterior and lateral pharyngeal walls



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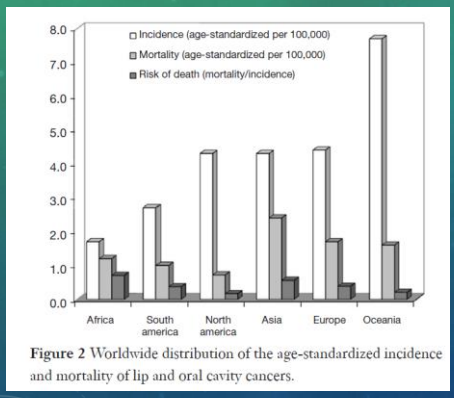

Figure 2 Worldwide distribution of the age-standardized incidence and mortality of lip and oral cavity cancers.

**ORAL CAVITY**

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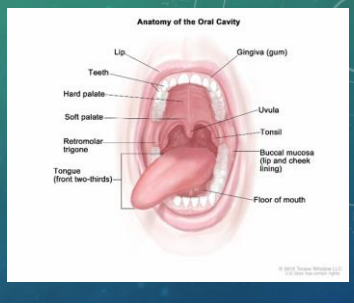


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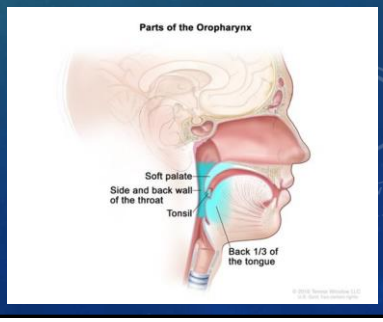
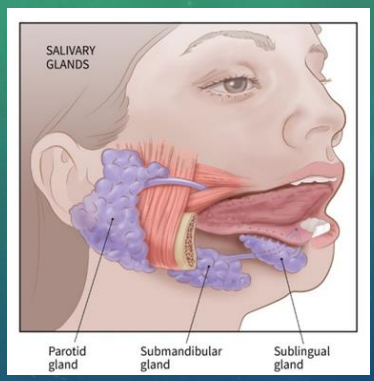



# LEUKOPLAKIA AND ERYTHROPLAKIA (POSSIBLE PRE-CANCEROUS CONDITIONS)

- Leukoplakia and erythroplakia are terms used to describe tissue changes seen in the mouth or throat:
  - Leukoplakia is a white or gray patch.
  - Erythroplakia is a flat or slightly raised, red area that often bleeds easily if it's scraped.
  - Erythroleukoplakia is a patch with both red and white areas.
- Your dentist or dental hygienist may be the first person to find these white or red patches.
- They may be cancer, they may be a pre-cancerous condition called dysplasia, or they could be a relatively harmless change.
- Dysplasia is graded as mild, moderate, or severe, based on how abnormal the tissue looks under the microscope. Knowing the degree of dysplasia helps predict how likely it is to progress to cancer or go away on its own or after treatment.
  - For example, severe dysplasia is more likely to become a cancer, while mild dysplasia is more likely to go away completely.
- The most common causes of leukoplakia and erythroplakia are smoking and chewing tobacco. Poorly fitting dentures that rub against the tongue or the inside of the cheek can also cause these changes. But sometimes, there's no clear cause. Dysplasia will often go away if the cause is removed.
- A biopsy is the only way to know for certain if an area of leukoplakia or erythroplakia contains dysplastic (pre-cancerous) cells or cancer cells.

# INTRODUCTION TO SALIVARY GLANDS (MAJOR/ MINOR)




**There are 3 sets of major salivary glands on each side of the face:**

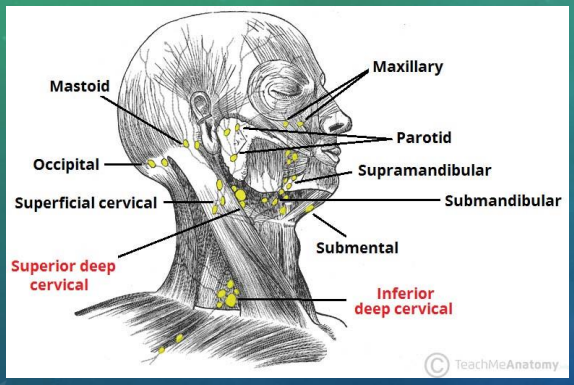
1. **The parotid glands**, the largest salivary glands, are just in front of the ears. About 7 out of 10 salivary gland tumors start here. Most of these tumors are benign (not cancer), but the parotid glands still are where most malignant (cancerous) salivary gland tumors start.

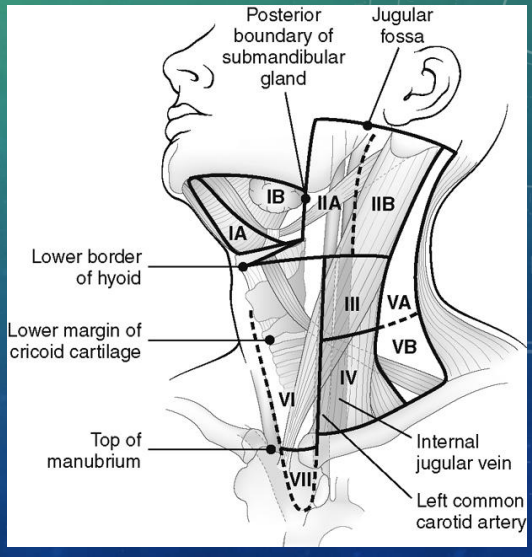
2. **The submandibular glands** are smaller and are below the jaw. They secrete saliva under the tongue. About 1 to 2 out of 10 tumors start in these glands, and about half of these tumors are cancer.

3. **The sublingual glands**, which are the smallest, are under the floor of the mouth and below either side of the tongue. Tumors starting in these glands are rare.

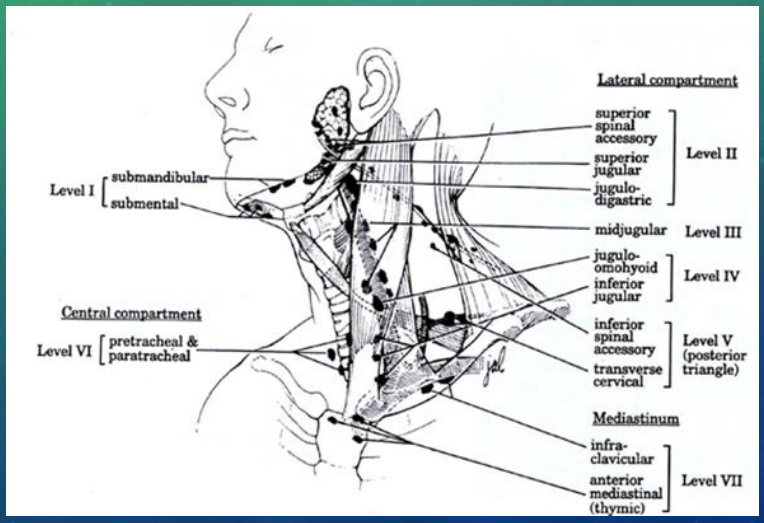
4. **There are also several hundred minor salivary glands that are too small to see without a microscope. These glands are under the lining of the lips and tongue; in the roof of the mouth; and inside the cheeks, nose, sinuses, and larynx (voice box).** Tumors in these glands are uncommon, but they are more often cancerous than benign. Cancers of the minor salivary glands most often start in the roof of the mouth.

# INTRODUCTION TO CERVICAL LYMPH NODE REGIONS



# INTRODUCTION TO CERVICAL LYMPH NODE REGIONS

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# INTRODUCTION TO CERVICAL LYMPH NODE REGIONS EXTRACAPSULAR/EXTRA-NODAL EXTENSION (ENE)

Extranodal extension (ENE) is defined as “the extension of a nodal metastasis through the lymph node capsule into adjacent tissue” and is a prognostic factor for most head and neck tumors.

| Code | Description |
|---|---|
| 0.0 | Lymph nodes positive for cancer but ENE not identified or negative |
| 0.1-9.9 | ENE 0.1 to 9.9 mm |
| X.1 | ENE 10 mm or greater |
| X.2 | ENE microscopic, size unknown<br>Stated as ENE (mi) |
| X.3 | ENE major, size unknown<br>Stated as ENE (ma) |
| X.4 | ENE present, microscopic or major unknown, size unknown |
| X.7 | Surgically resected regional lymph nodes negative for cancer (pN0) |
| X.8 | Not applicable: Information not collected for this case<br>(If this information is required by your standard setter, use of code X.8 may result in an edit error) |
| X.9 | Not documented in medical record<br>No surgical resection of regional lymph nodes<br>ENE not assessed pathologically, or unknown if assessed |

Microscopic ENE [ENE (mi)] is defined as less than or equal to 2 mm.
Major ENE [ENE (ma)] is defined as greater than 2 mm.
Both ENE (mi) and ENE (ma) qualify as ENE (+) for definition of pN.

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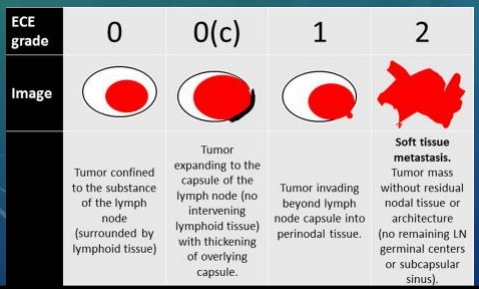
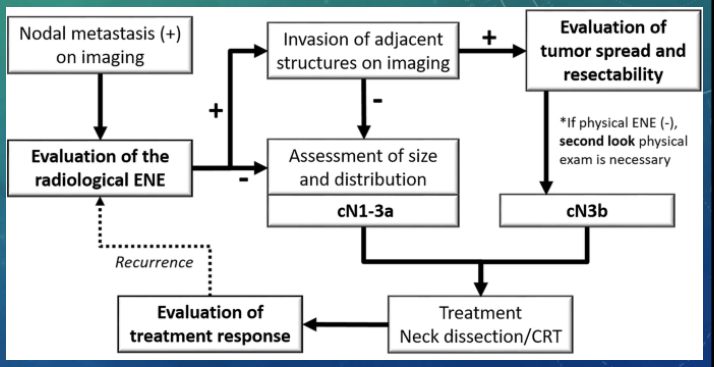
# WHY IS EXTRA-NODAL EXTENSION SO IMPORTANT? WHY ISN'T IT DOCUMENTED VERY WELL?

**Relationship of Node Location to Likely Disease**

- Nodes at certain levels more likely certain primaries
- Upper neck nodes are the most likely to be head and neck cancer
  - Subdigastric node may be virtually any head and neck primary, or a non-Hodgkin's lymphoma
  - Submandibular node suggests oral cavity, lip, nasal vestibule or salivary gland primary
  - Submental nodes are uncommon

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# RISK FACTORS FOR ORAL CAVITY/OROPHARYNX CANCERS

- Tobacco (including chewing tobacco and snuf)
- Alcohol (beer, wine, hard liquor, moonshine)
- Poor Dental Health & Malnutrition
- Chewing Betel Quid– a stimulant used Asia containing areca/betal leaf, tobacco – you chew it
- Viral: EBV, HHV-8, HPV Type 16
- Chronic Irritation from Dentures
- Radiation Exposure (total body or oropharyngeal)
- Age Over 50
- Environmental/Occupational exposure (rubber industry, solvents, pesticides, asbestos, silica dust)
- Genetic: KIT, MYB-NFIB, PLAG1, ECT1-MAML2, MAD1L1, HMGIC, HER2/neu, RAS, c-fos, Sox, BCL2, TrkC/NTRK316

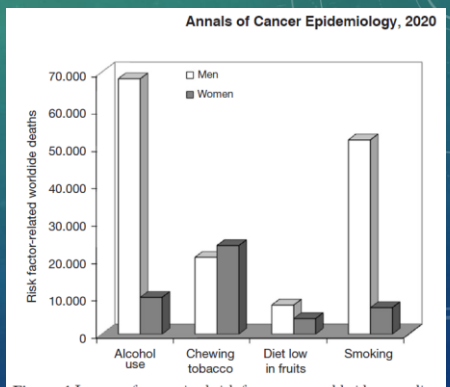


**Figure 4** Impact of recognized risk factors on worldwide mortality for lip and oral cavity cancers.

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# P-16, HPV, HISTOLOGY CODES 8585 AND 8586 VS 8070 CONFUSING ALONG WITH POOR INSTRUCTIONS

- What is p-16 testing? – NOT EXCLUSIVE TO H&N CANCER
- What is HPV testing? – NOT EXCLUSIVE TO H&N CANCER
- What is EBV testing? – NOT EXCLUSIVE TO H&N CANCER
- What is p16/HPV-mediated – OMG!! What's the Difference?
- What is histology code 8085 designed for?
- What is histology code 8086 designed for:
- Now make sense out of it all, please – in English.

**8/22/18 Summary of Changes**

8085/3- Squamous cell carcinoma, HPV-positive (C01.9, 09.9, C10.2, C10.3, C10.8, C10.9, C31.0–C31.3, C31.9). 09.9 added to the topography codes eligible for this histology.

8086/3- Squamous cell carcinoma, HPV-negative (C01.9, 09.9, C10.2, C10.3, C10.8, C10.9, C31.0–C31.3, C31.9). 09.9 added to the topography codes eligible for this histology.

8085 and 8086 can **ONLY** be used for Squamous Cell Carcinoma in a few H&N sites or they fail edits

**Oral Cavity**
C01.9 - Base of Tongue
C09.0 - Tonsillar Fossa
C09.1 - Tonsillar Pillar
C09.8 - Overlapping Lesion of Tonsil
C09.9 - Tonsil, NOS

**Oropharynx (C10)**
C10.0 – Vallecula
C10.1 – Anterior Surface of Epiglottis
C10.2 – Lateral Wall of Epiglottis
C10.3 – Posterior Wall of Oropharynx
C10.4 – Branchial Cleft
C10.8 – Overlapping Lesion of Oropharynx
C10.9 – Oropharynx, NOS

**Accessory Sinuses (C31)**
C31.0 – Maxillary Sinus
C31.1 – Ethmoid Sinus
C31.2 – Frontal Sinus
C31.3 – Sphenoid Sinus
C31.8 – Overlapping Lesion of Accessory Sinuses
C31.9 – Accessory Sinus, NOS

| Site remode | Site Description | Histology/Behavior | Histology/Behavior Description |
|---|---|---|---|
| C019 | BASE OF TONGUE | 8085/3 | Squamous cell carcinoma, HPV– positive |
| C019 | BASE OF TONGUE | 8086/3 | Suamous cell carcinoma, HPV– positive |
| C090-C091,C098-C104,C108-C109 | OROPHARNYX | 8085/3 | Squamous cell carcinoma, HPV– positive |
| C090-C091,C098-C104,C108-C109 | OROPHARNYX | 8086/3 | Squamous cell carcinoma, HPV– positive |
| C111 | POSTERIOR WALL OF NASOPH | 8085/3 | Squamous cell carcinoma, HPV– positive |
| C111 | POSTERIOR WALL OF NASOPH | 8086/3 | Squamous cell carcinoma, HPV– positive |
| C310-C313,C318 | SINUSES | 8085/3 | Squamous cell carcinoma, HPV– positive |
| C310-C313,C318 | SINUSES | 8086/3 | Squamous cell carcinoma, HPV– positive |
| C319 | ACCESSORY SINUS,NOS | 8085/3 | Squamous cell carcinoma, HPV– positive |
| C319 | ACCESSORY SINUS,NOS | 8086/3 | Squamous cell carcinoma, HPV– positive |

# WHAT IS P-16 TEST AND DIFFERENCE FROM HPV TEST WHAT IS DIFFERENCE BETWEEN 8070, 8071, 8085, 8086

- There are more than 100 types of Human papillomavirus or HPV that are extremely common and worldwide.
- There are at least 14 known high-risk cancer-causing types of HPV that are monitored worldwide.
- HPV infections usually clear up within a few months with no intervention, about 90% clear within 2 years.
- HPV-related cancers can be prevented or at least the risk of exposure reduced with protective measures.
- HPV exposure by any person can increase risk of HPV-related cancers. These include specific Head and Neck Cancers, Cervical Cancer, Anal and Rectal Cancers, Other External Genitalia Cancers in Women and Men,

There are now vaccines that help prevent exposure to and risking cancer for specific types of HPV strains.

HPV types 16 and 18 are common HPV strains associated with increased cancer risk.

HPV has even been found in lung, liver, and other unusual cancer sites.

Cervical Cancer remains the #1 preventable virus-associated cancer – many tests can now check for it – and vaccine.

Testing: 3 types currently;

- A Pap test only. If your result is normal, your doctor may tell you that you can wait three years until your next Pap test.
- An HPV test only. This is called primary HPV testing. If your result is normal, your doctor may tell you that you can wait five years until your next screening test.
- An HPV test along with the Pap test. This is called co-testing. If both of your results are normal, your doctor may tell you that you can wait five years until your next screening test.

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# CHARACTERISTICS OF CANCER AND DEGREE OF RISK

Table 1. Characteristics Associated With the Risk of Oropharyngeal Cancer[a]

| Degree of Risk | Characteristics | 3-y OS Rate |
|---|---|---|
| Low | HPV+, smoking history of ≤10 pack-years, and N0–N2a nodal history | 93% (95% CI, 88.3–97.7) |
| Intermediate | HPV+, smoking history of >10 pack-years, and N2b–N3 nodal disease; or | 70.8% (95% CI, 60.7–80.8) |
| | HPV-, smoking history of ≤10 pack-years, and N2b–N3 nodal disease or T2–T3 tumors | |
| High | HPV- and smoking history >10 pack-years; or | 46.2% (95% CI, 34.7–57.7) |
| | HPV-, smoking history ≤10 pack-years, and T4 disease | |

CI = confidence interval; HPV = human papillomavirus; OS = overall survival; + = positive; - = negative. (Refer to the AJCC Staging Groupings and TNM Definitions section of this summary for more information.)

[a]Ang KK, Harris J, Wheeler R, et al.: Human papillomavirus and survival of patients with oropharyngeal cancer. N Engl J Med 363 (1): 24-35, 2010.

Note 1: There is evidence that human papilloma virus (HPV) plays a role in the pathogenesis of some cancers. HPV testing may be performed for prognostic purposes; testing may also be performed on metastatic sites to aid in determination of the primary site.

Note 2: Record the results of any HPV testing performed on pathological specimens including surgical and cytological (from cell blocks) tissue from the primary tumor or a metastatic site, including lymph nodes. Do not record the results of blood tests or serology.

Note 3: There are several methods for determination of HPV status. The most frequently used test is IHC for p16 expression which is surrogate marker for HPV infection. Do not record the results of IHC p16 expression in this field. The rest of the tests (based on ISH, PCR, RT-PCR technologies) detect the viral DNA or RNA. This data item is only for HPV status determined by tests designed to detect viral DNA or RNA.

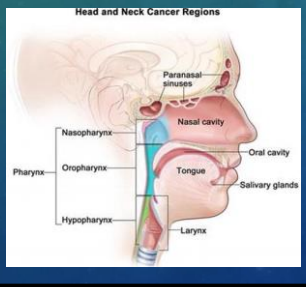
Note 4: HPV-type 16 refers to virus type and is different[17] from p16 overexpression (p16+)

# WHAT IS P-16 TEST AND DIFFERENCE FROM HPV TEST WHAT IS DIFFERENCE BETWEEN 8070, 8071, 8085, 8086

- Currently there is no consensus on the best detection method(s) that should be used to identify HPV-related oropharyngeal squamous cell carcinomas and serve as a standard test (or tests) for routine diagnostic use.
- Approximately 60–70% of newly diagnosed Oropharyngeal Squamous Cell Carcinomas are associated with HPV in the United States and some parts of Europe – usually HPV Type 16 – multiple methods of detection.
- Quantitative PCR-based HPV-16 DNA (E6 and E7 genes) in the saliva and plasma of Oropharyngeal Primary Squamous Cell Cancer samples before and after treatment has been explored as a biomarker of recurrence.
- p-16 Test - The test that is most widely available in clinical laboratories and most widely used in clinical practice is p16 (INK4A) detection by IHC. This protein is a surrogate marker of transcriptionally active high-risk HPV infection. p16 IHC is inexpensive, is performed on FFPE samples which are routinely generated in pathology laboratories as part of standard clinical surgical pathology practice, and has a sensitivity for transcriptionally active high-risk HPV that is almost 100%.
- p16 IHC has demonstrated good agreement with HPV E6/E7 mRNA expression detected by RT-PCR 22 and RNA ISH. Extensive literature shows that p16 expression in OPSCC is associated with improved overall and disease specific survival independent of all other known prognostic factors with two to five times lower risk of adverse outcomes. A significant issue with p16 IHC is that, until recently, there has been no consensus on the definition of a positive p16 IHC result.
- BUT - up to 26% of nonsquamous, basaloid carcinomas such as solid type adenoid cystic carcinoma, which are not associated with HPV, can be diffusely positive for p16. While some of these sinonasal carcinomas have been more recently shown to be the unique tumor 'HPV-related multiphenotypic carcinoma'. So, we must cautiously use this marker with its known specificities and limitations as to not jump to any conclusions regarding primary tumor sites.[18]

# ORAL CAVITY CANCER POST-SCREENING HPV TESTS

- **RULES FOR EBV, HPV and p16 Coding**
- You can ONLY use EBV testing for the following:
  - C11.9 (nasopharynx) with a T0 and not for C76.0
- You can ONLY use p16 as a surrogate marker for HPV for:
  - NOTE: They really want the p16 test not HPV surrogate
  - If another HPV test is positive – the p16 is still negative
  - C10.9 (oropharynx) with a T0 and not C76.0
  - C14.8 (pharynx other) – do not use C76.0 in this case
- AJCC and SEER have not agreed on addition of sites affected by EBV/HPV
- YOU CAN ONLY USE p16 for
- YOU CAN ONLY CODE HISTOLOGY 8585/8586 for C100-C109, C090-C099
- You may see EBV or p16 + tumors elsewhere – do not code EBV or p16
- DO INCLUDE THIS IN YOUR TEXT – HPV Affects Many Other H&N Sites




***Note 2***: If there is no evidence of the primary tumor, yet the physician "suspects" a specific head and neck subsite, do not assign that primary site, but code C760 (see exceptions for EBV positive or p16 positive cancers.)

| Code | Description | Disease |
|---|---|---|
| 0 | Not Occult | EOD/SS schema (Ill-Defined, Other; Soft Tissue Other for 8941) |
| 1 | Occult, Negative cervical nodes (regional head and neck nodes) | EOD/SS schema (Ill-Defined, Other; Soft Tissue Other for 8941) |
| 2 | Not tested for EBV or p16 in head and neck regional nodes (EBV and p16 both unknown) | 6: Cervical Lymph Nodes and Unknown Primary Tumors of the Head and Neck |
| 3 | Unknown EBV, p16 negative in head and neck regional nodes | 6: Cervical Lymph Nodes and Unknown Primary Tumors of the Head and Neck |
| 4 | Unknown p16, EBV negative in head and neck regional nodes | 6: Cervical Lymph Nodes and Unknown Primary Tumors of the Head and Neck |
| 5 | Negative for both EBV and p16 in head and neck regional nodes | 6: Cervical Lymph Nodes and Unknown Primary Tumors of the Head and Neck |
| <BLANK> | Not C760, discriminator does not apply<br><br>Positive p16 in head and neck regional nodes, EBV unknown or negative<br>Assign primary site C109<br><br>Positive EBV in head and neck regional nodes, p16 positive, negative, or unknown<br>Assign primary site C119 | Various<br><br>10: HPV-Mediated (p16+) Oropharyngeal Cancer (C109) **(Schema ID 00100: Oropharynx HPV-Mediated (p16+))**<br><br>9: Nasopharynx (C119) **(Schema ID 00090: Nasopharynx)** |

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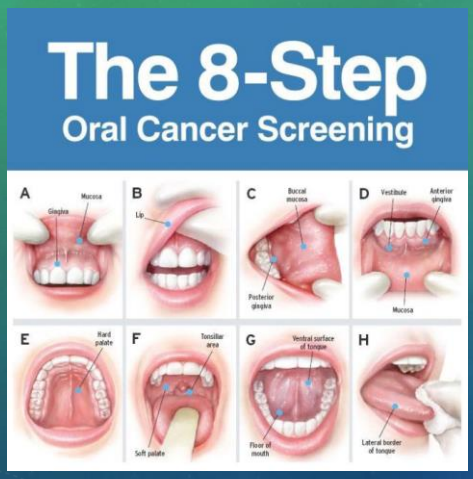
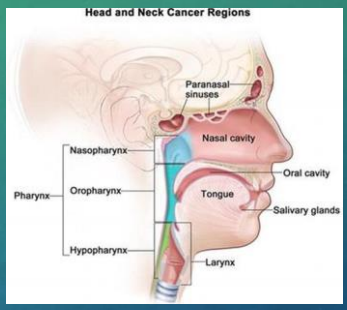
# ORAL CAVITY CANCER SCREENING - DENTISTS




EBV infections (a herpes virus) increases risk of nasopharyngeal cancer, lymphoma like Burkitt, DLBCL or Hodgkin Lymphoma as well as in stomach cancer.




REALLY IMPORTANT NOTE: HPV infections are very common and increase the risk of dozens of anatomic sites in the body including; cervix, vagina, vulva, oral cavity, oropharynx, base of tongue, tonsils and select other sites of the head and neck, anus, rectum, and penis.

THIS IS MISLEADING and INCORRECT IN BOTH THE SSDI & AJCC Manuals and Definitions

- Epstein-Barr Virus (EBV): EBV positive cancers are associated with nasopharyngeal cancer.
  - If the EBV (EBER) test is done and is positive, the primary site should be assigned to C119 (nasopharynx, NOS) instead of C760, so that the Nasopharynx staging system can be used. Nasopharynx has a T0, for no evidence of primary tumor
- p16: p16 positive cancers in the head and neck are associated with oropharyngeal cancer. p16 is a surrogate marker for Human Papilloma Virus (HPV).
  - If the p16 test is done and positive (and EBV is negative or unknown), the primary site should be assigned to C109 (oropharynx, NOS) instead of C760, so that the Oropharynx staging system can be used. Oropharynx has a T0, for no evidence of primary tumor.
  - *Note*: p16 is the only test that can be used for this discriminator. If there is another HPV test that is positive, the p16 would still be negative for purposes of this data item.

# HISTOLOGICAL CLASSIFICATION OF ORAL CAVITY, OROPHARYNX AND SALIVARY GLAND TUMORS

- Oral Cavity is bathed in carcinogens for a lifetime – field effect to environmental factors, smoking, drinking
  - HPV Negative Squamous Cell Carcinoma
  - HPV Positive Squamous Cell Carcinoma
  - Oral Cavity Melanoma and Kaposi Sarcoma or Myofibroblastic Sarcoma

FACTORS: type of cancer, size of tumor, location of cancer, nodes, speech, grade, swallowing ability – HPV, EBV, Alcohol, Tobacco, Chew, Snuff, Vaping, other VIruses.

- Oropharynx is further back so more related to smoking than others – throat cancers
  - HPV Negative Squamous Cell Carcinoma
  - HPV Positive Squamous Cell Carcinoma
  - Lymphoid Neoplasms – Hodgkin, Burkitt, Follicular, Mantle Cell, T-Lymphoblastic Lymphomas

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FACTORS: type of cancer, size of tumor, location of cancer, nodes, speech, grade, swallowing ability – HPV, EBV, Alcohol, Tobacco, Chew, Snuff, Vaping, other VIruses.

4 Tumours of the oral cavity and mobile tongue
- WHO and TNM classifications
- Introduction
- Malignant surface epithelial tumours
  - Squamous cell carcinoma
- Oral potentially malignant disorders & oral epithelial dysplasia
  - Oral potentially malignant disorders
  - Oral epithelial dysplasia
  - Proliferative verrucous leukoplakia
- Papillomas
  - Squamous cell papilloma
  - Condyloma acuminatum
- Verruca vulgaris
- Multifocal epithelial hyperplasia

Tumours of uncertain histogenesis
- Congenital granular cell epulis
- Ectomesenchymal chondromyxoid tumour

Soft tissue and neural tumours
- Granular cell tumour
- Rhabdomyoma
- Lymphangioma
- Haemangioma
- Schwannoma and neurofibroma
- Kaposi sarcoma
- Myofibroblastic sarcoma

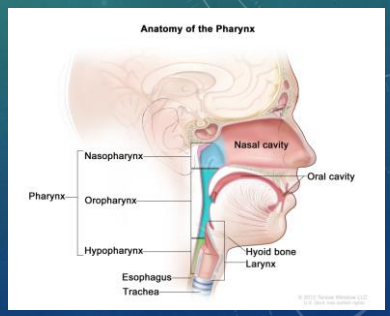
Oral mucosal melanoma

Salivary type tumours
- Mucoepidermoid carcinoma
- Pleomorphic adenoma

Haematolymphoid tumours
- Overview
- CD30-positive T-cell lymphoproliferative disorder
- Plasmablastic lymphoma
- Langerhans cell histiocytosis
- Extramedullary myeloid sarcoma

# HISTOLOGICAL CLASSIFICATION OF ORAL CAVITY, OROPHARYNX AND SALIVARY GLAND TUMORS

5 Tumours of the oropharynx (base of tongue, tonsils, adenoids)
- WHO and TNM classifications
- Introduction
- Squamous cell carcinoma
  - Squamous cell carcinoma, HPV-positive
  - Squamous cell carcinoma, HPV-negative
- Salivary gland tumours
  - Pleomorphic adenoma
  - Adenoid cystic carcinoma
  - Polymorphous adenocarcinoma
- Haematolymphoid tumours
  - Introduction
  - Hodgkin lymphoma
  - Burkitt lymphoma
  - Follicular lymphoma
  - Mantle cell lymphoma
  - T-lymphoblastic leukaemia/lymphoma
  - Follicular dendritic cell sarcoma

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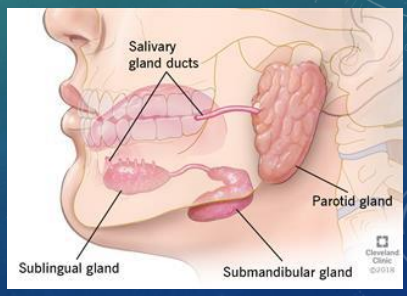
# HISTOLOGICAL CLASSIFICATION OF ORAL CAVITY, OROPHARYNX AND SALIVARY GLAND TUMORS

- Salivary Glands are glandular so they form into adenocarcinoma of various types – type of cancer, size of tumor, nodes, speech, symptoms, mets, grade
  - Types of cancerous (malignant) salivary gland tumors include:
  - Acinic cell carcinoma.
  - Adenocarcinoma.
  - Adenoid cystic carcinoma.
  - Clear cell carcinoma.
  - Malignant mixed tumor.
  - Mucoepidermoid carcinoma.
  - Oncocytic carcinoma.
  - Polymorphous low-grade adenocarcinoma
  - Mammary analogue secretory carcinoma - major or minor glands

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# HISTOLOGICAL CLASSIFICATION OF ORAL CAVITY, OROPHARYNX AND SALIVARY GLAND TUMORS

Histologic classification for salivary gland tumors in the WHO 4th Edition has 22 carcinomas, 11 benign tumors, 4 non-neoplastic epithelial lesions, 3 benign soft-tissue entities, and MALT Lymphoma, making it one of the most extensive classifications for tumors[7]

| **Malignant tumors** | | **Benign tumors** |
|---|---|---|
| Mucoepidermoid carcinoma | Secretory carcinoma | Pleomorphic adenoma |
| Adenoid cystic carcinoma | Sebaceous adenocarcinoma | Myoepithelioma |
| Acinic cell carcinoma | Carcinosarcoma | Basal cell adenoma |
| Polymorphous adenocarcinoma | Poorly differentiated carcinoma | Warthin tumor |
| Clear cell carcinoma | Undifferentiated carcinoma | Oncocytoma |
| Basal cell carcinoma | Large cell neuroendocrine carcinoma | Lymphadenoma |
| Intraductal carcinoma | Small cell neuroendocrine carcinoma | Cystadenoma |
| Adenocarcinoma, NOS | Lymphoepithelial carcinoma | Sialadenoma |
| Salivary duct carcinoma | Squamous cell carcinoma | Ductal papillomas |
| Myoepithelial carcinoma | Oncocytic carcinoma | Sebaceous adenoma |
| Epithelial-myoepithelial carcinoma | Uncertain malignant potential | Canalicular adenoma and other ductal adenomas |
| Carcinoma ex pleomorphic adenoma | Sialoblastoma | |
| **Non-neoplastic epithelial lesions** | **Benign soft tissue lesions** | **Hematolymphoid tumors** |
| Sclerosing polycystic adenosis | Hemangioma | Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma) |
| Nodular oncocytic hyperplasia | Lipoma/sialolipoma | |
| Lymphoepithelial sialadenitis | Nodular fasciitis | |
| Intercalated duct hyperplasia | | |

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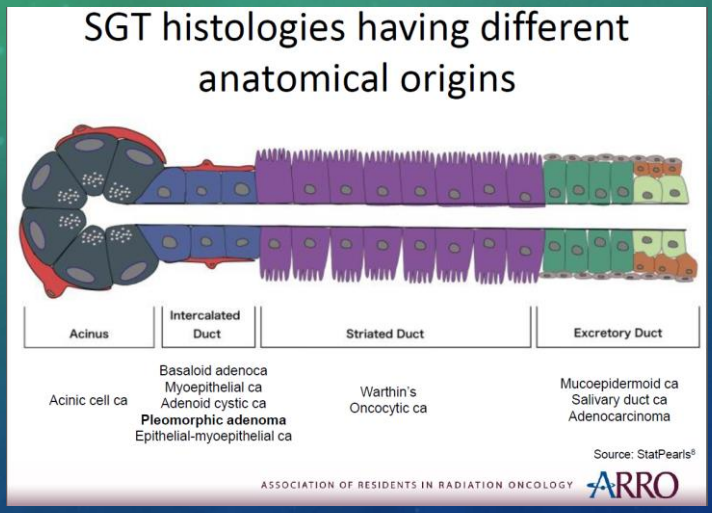
# HISTOLOGICAL CLASSIFICATION OF ORAL CAVITY, OROPHARYNX AND SALIVARY GLAND TUMORS

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# CODING GRADE FOR MOST HEAD AND NECK CANCERS GRADE TABLE 1 – ORAL CAVITY

| Schema ID# | Schema ID Name | AJCC ID | AJCC Chapter |
|---|---|---|---|
| 00071 | Lip | 7 | Oral Cavity |
| 00072 | Tongue Anterior | 7 | Oral Cavity |
| 00073 | Gum | 7 | Oral Cavity |
| 00074 | Floor of Mouth | 7 | Oral Cavity |
| 00075 | Palate Hard | 7 | Oral Cavity |
| 00076 | Buccal Mucosa | 7 | Oral Cavity |
| 00077 | Mouth Other | 7 | Oral Cavity |

| Code | Grade Description |
|---|---|
| 1 | G1: Well differentiated |
| 2 | G2: Moderately differentiated |
| 3 | G3: Poorly differentiated |
| 9 | Grade cannot be assessed (GX); Unknown |

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# CODING GRADE FOR MOST HEAD AND NECK CANCERS GRADE TABLE - OROPHARYNX

| Schema ID# | Schema ID Name | AJCC ID | AJCC Chapter |
|---|---|---|---|
| 00111 | Oropharynx (p16-) | 11.1 | Oropharynx (p16-) |

| Code | Grade Description |
|---|---|
| 1 | G1: Well differentiated |
| 2 | G2: Moderately differentiated |
| 3 | G3: Poorly differentiated |
| 4 | G4: Undifferentiated |
| 9 | Grade cannot be assessed (GX); Unknown |

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# CODING GRADE FOR MOST HEAD AND NECK CANCERS GRADE TABLE – SALIVARY GLANDS – TABLE 98

| | | | |
|---|---|---|---|
| 00080 | Major Salivary Glands | 8 | Major Salivary Glands |

| Code | Grade Description |
|---|---|
| A | Well differentiated |
| B | Moderately differentiated |
| C | Poorly differentiated |
| D | Undifferentiated, anaplastic |
| 9 | Grade cannot be assessed; Unknown |

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# WHEN IS A HEAD & NECK MOST LIKELY METASTATIC CANCER?

- A comparison of patients (N = 2,230) with index SCC of the oropharynx site and index SCC of non-oropharyngeal sites (i.e., oral cavity, larynx, and hypopharynx) was performed to determine the likelihood of developing second primary malignancies.
- The second primary malignancy rate was lower for patients with index oropharyngeal SCC than for patients with index non-oropharyngeal cancer (P < .001).
- Among patients with oropharyngeal SCC, former smokers had a 50% higher risk of second primary malignancy than never-smokers, and current smokers had a 100% higher risk than never-smokers (P trend = .008).
- These data suggest that patients who fit the typical HPV phenotype have a very low risk of second-primary malignancy

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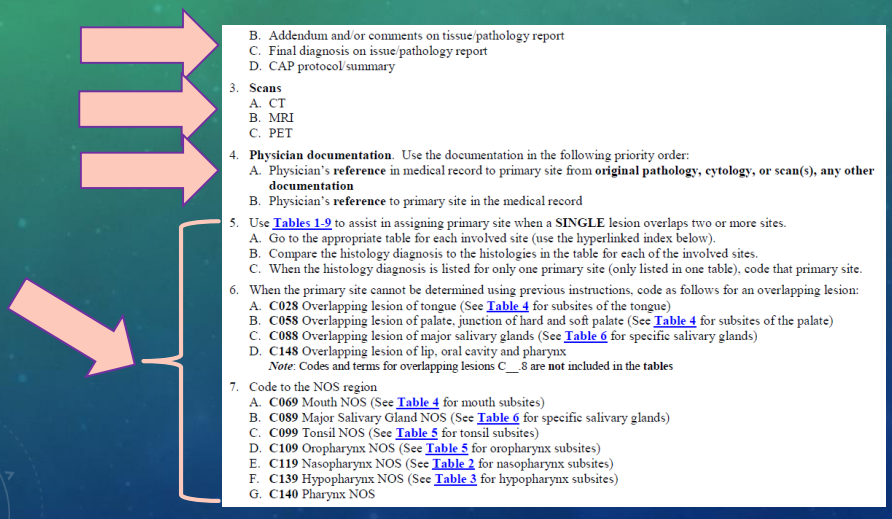
# HIGHLIGHTS FROM THE 2021 HEAD & NECK SOLID TUMORS MANUAL – MULTIPLE PRIMARY/HISTOLOGY

Determining Primary Site – Use Tables 1-9 which divides the H&N into manageable sized sections



B. Addendum and/or comments on tissue/pathology report
C. Final diagnosis on issue/pathology report
D. CAP protocol/summary

3. **Scans**
   A. CT
   B. MRI
   C. PET

4. **Physician documentation**. Use the documentation in the following priority order:
   A. Physician's **reference** in medical record to primary site from **original pathology, cytology, or scan(s), any other documentation**
   B. Physician's **reference** to primary site in the medical record

5. Use Tables 1-9 to assist in assigning primary site when a **SINGLE** lesion overlaps two or more sites.
   A. Go to the appropriate table for each involved site (use the hyperlinked index below).
   B. Compare the histology diagnosis to the histologies in the table for each of the involved sites.
   C. When the histology diagnosis is listed for only one primary site (only listed in one table), code that primary site.

6. When the primary site cannot be determined using previous instructions, code as follows for an overlapping lesion:
   A. **C028** Overlapping lesion of tongue (See Table 4 for subsites of the tongue)
   B. **C058** Overlapping lesion of palate, junction of hard and soft palate (See Table 4 for subsites of the palate)
   C. **C088** Overlapping lesion of major salivary glands (See Table 6 for specific salivary glands)
   D. **C148** Overlapping lesion of lip, oral cavity and pharynx
   *Note*: Codes and terms for overlapping lesions C__.8 are **not** included in the **tables**

7. Code to the NOS region
   A. **C069** Mouth NOS (See Table 4 for mouth subsites)
   B. **C089** Major Salivary Gland NOS (See Table 6 for specific salivary glands)
   C. **C099** Tonsil NOS (See Table 5 for tonsil subsites)
   D. **C109** Oropharynx NOS (See Table 5 for oropharynx subsites)
   E. **C119** Nasopharynx NOS (See Table 2 for nasopharynx subsites)
   F. **C139** Hypopharynx NOS (See Table 3 for hypopharynx subsites)
   G. **C140** Pharynx NOS

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# HIGHLIGHTS FROM THE 2021 HEAD & NECK SOLID TUMORS MANUAL – MULTIPLE PRIMARY/HISTOLOGY

Determining Primary Site – Use Tables 1-9 which divides the H&N into manageable sized sections

| Table Number | Table Title |
|---|---|
| Table 1 | Tumors of Nasal Cavity **C300** Paranasal Sinuses **C310-C313, C318, C319** |
| Table 2 | Tumors of Nasopharynx **C110, C111 (posterior wall of nasopharynx only), C112, C113, C118, C119** |
| Table 3 | Pyriform Sinus **C129** Tumors of Hypopharynx **C130-C132, C138, C139** Larynx **C320-C323, C328, C329** Trachea **C339** and Parapharyngeal Space **C139** |
| Table 4 | Tumors of Oral Cavity and mobile tongue **C020-C024, C028, C029, C030, C031, C039, C040, C041, C048, C049, C050-C052, C058, C059, C060-C062, C068, C069** |
| Table 5 | Tumors of Oropharynx **C100-C104, C108 C109** Base of Tongue **C019** Tonsils **C090, C091, C098, C099** Adenoids/pharyngeal tonsil only **C111** |
| Table 6 | Tumors of Salivary Glands **C079, C080, C081, C088, C089** |
| Table 7 | Tumors of Odontogenic and Maxillofacial Bone (Mandible **C411**, Maxilla **C410**) |
| Table 8 | Tumors of Ear **C301** and External auditory canal **C442** |
| Table 9 | Paraganglioma of Carotid body, Larynx, Middle Ear, Vagal nerve **C479** |
| Table 10 | Paired Sites |



# HIGHLIGHTS FROM THE 2021 HEAD & NECK SOLID TUMORS MANUAL – MULTIPLE PRIMARY/HISTOLOGY

**Table 4: Tumors of Oral Cavity and Mobile Tongue**

**Table 4** lists the more common histologies for the following head and neck subsites:

The **oral cavity category** includes the following:

**Mobile Tongue:**
- **C020** Dorsal surface of tongue NOS
- **C021** Border of tongue
- **C022** Ventral surface of tongue NOS
- **C023** Anterior 2/3 of tongue NOS
- **C024** Lingual tonsil
- **C028** Overlapping lesion of tongue
- **C029** Tongue NOS

**Gum:**
- **C030** Upper gum, maxillary gingiva, upper alveolar mucosa, upper alveolar ridge mucosa, upper alveolus, upper gingiva
- **C031** Lower gum mandibular gingiva, lower alveolar mucosa, lower alveolar ridge mucosa, lower alveolus, lower gingiva
- **C039** Gum NOS, gingiva NOS, alveolar mucosa NOS, alveolar ridge mucosa NOS, alveolar NOS periodontal tissue, tooth socket

**Floor of Mouth:**
- **C040** Anterior floor of mouth
- **C041** Lateral floor of mouth
- **C048** Overlapping lesion floor of mouth
- **C049** Floor of mouth NOS

**Palate:**
- **C050** Hard palate
- **C051** Soft palate
- **C052** Uvula
- **C058** Overlapping lesion of palate, junction of hard and soft palate
- **C059** Palate NOS, roof of mouth

**Other and unspecified parts of Mouth:**
- **C060** Cheek mucosa, buccal mucosa, internal cheek

# HIGHLIGHTS FROM THE 2021 HEAD & NECK SOLID TUMORS MANUAL – MULTIPLE PRIMARY/HISTOLOGY

**Table 5: Tumors of the Oropharynx, Base of Tongue, Tonsils, Adenoids**

**Table 5** lists the more common histologies for the following head and neck subsites:

**Oropharynx:**

**C100** Vallecula
**C101** Anterior surface of epiglottis
**C102** Lateral wall of oropharynx; lateral wall of nasopharynx
**C103** Posterior wall of oropharynx; posterior wall of nasopharynx
**C104** Brachial cleft
**C108** Overlapping lesion of oropharynx; junctional region of oropharynx
**C109** Oropharynx NOS; mesopharynx NOS; fauces NOS. Use this code only when the subsite has not been identified a subsite as the origin of the lesion.
*Note:* Code overlapping lesion of oropharynx; junctional region of oropharynx **C108** when a single tumor overlaps subsites of the oropharynx. For example, a single lesion which overlaps the vallecular and the anterior surface of the epiglottis.

**C019** Base of tongue

**Tonsils:**

**C090** Tonsillar fossa
**C091** Tonsillar pillar
**C098** Overlapping lesion of tonsil
**C099** Tonsil NOS
**C111** Adenoids/pharyngeal tonsil (does not include posterior wall of nasopharynx)

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the Hematopoietic Database.

**Column 1** contains specific and NOS histology terms.
- **Specific** histology terms **do not** have **subtypes/variants**
- **NOS** histology terms **do** have **subtypes/variants.**

**Column 2** contains **synonyms** for the specific or NOS term. Synonyms have the **same** histology **code** as the specific or NOS term.
**Column 3** contains **subtypes/variants** of the **NOS** histology. Subtypes/variants **do not** have the **same** histology code as the NOS.

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# HIGHLIGHTS FROM THE 2021 HEAD & NECK SOLID TUMORS MANUAL – MULTIPLE PRIMARY/HISTOLOGY

**Table 6: Tumors of Salivary Glands**

**Table 6** lists the more common histologies for the following head and neck subsites:
- **C079** Parotid gland, parotid NOS Stensen duct, parotid gland duct
- **C080** Submandibular gland, submaxillary gland, Wharton duct, submaxillary gland duct
- **C081** Sublingual gland; sublingual gland duct
- **C088** Overlapping lesion of major salivary glands
- **C089** Major salivary gland NOS; salivary gland NOS

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the Hematopoietic Database.
*Note:* Hematopoietic neoplasms are common in the major salivary glands.

**Column 1** contains specific and NOS histology terms.
- **Specific** histology terms **do not** have **subtypes/variants**
- **NOS** histology terms **do** have **subtypes/variants**

**Column 2** contains **synonyms** for the specific or NOS term. Synonyms have the **same** histology **code** as the specific or NOS term.
**Column 3** contains **subtypes/variants** of the **NOS** histology. Subtypes/variants **do not** have the **same** histology code as the NOS.

*Note 1:* **Salivary duct carcinoma** was **assigned** code **8500** because it resembles high-grade duct carcinoma as found in the breast. These tumors are very aggressive. Code **8500 only** when the diagnosis is **exactly salivary duct carcinoma.**
*Note 2:* Assign code **8140** when the diagnosis is **salivary gland adenocarcinoma**.

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**Table begins on next page**

# HIGHLIGHTS FROM THE 2021 HEAD & NECK SOLID TUMORS MANUAL – MULTIPLE PRIMARY/HISTOLOGY

The Multiple Primary Rules and Histology Coding Rules are Quite Simple if You Use the Tables Correctly

**Rule M4** Abstract **multiple primaries**[ii] when separate/non-contiguous tumors are present in sites with ICD-O **site** codes that **differ** at the second C**X**xx, and/or third characters Cx**X**x.
*Note 1:* Use this rule only for **multiple tumors.**
*Note 2:* Timing is irrelevant.
*Note 3:* Histology is irrelevant.

**Rule M5** Abstract **multiple primaries**[ii] when there are separate/non-contiguous tumors on both the **right** side and the **left** side of a paired site.
*Note 1:* See Table 10 for a list of **paired sites.**
*Note 2:* Use this rule only for **multiple tumors.**
*Note 3:* Timing is irrelevant.
*Note 4:* Histology is irrelevant.

**Rule M6** Abstract **multiple primaries**[ii] when the patient has a subsequent tumor after being **clinically disease-free** for **greater than five years** after the original diagnosis or last recurrence.
*Note 1:* **Clinically** disease-free means that there was **no evidence** of recurrence on follow-up.
- Scopes are NED
- Scans are NED

*Note 2:* When there is a recurrence less than or equal to five years of diagnosis, the "**clock**" starts over. The time interval is calculated from the **date of last recurrence.** In other words, the patient must have been disease-free for greater than five years from the date of the last recurrence.
*Note 3:* When it is **unknown/not documented** whether the patient had a recurrence, use **date of diagnosis** to compute the time interval.
*Note 4:* The physician may state this is a **recurrence,** meaning the patient had a previous head and neck tumor and now has another head and neck tumor. **Follow the rules**; do not attempt to interpret the physician's statement.

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# HIGHLIGHTS FROM THE 2021 HEAD & NECK SOLID TUMORS MANUAL – MULTIPLE PRIMARY/HISTOLOGY

The Multiple Primary Rules and Histology Coding Rules are Quite Simple if You Use the Tables Correctly

**Coding Histology**

*Note 1:* The priority is to code the most specific histology. **DO NOT USE BREAST HISTOLOGY CODING RULES FOR THIS SITE.**
*Note 2:* Only use this section for one or more histologies within a single tumor.
*Note 3:* Do not use this section in place of the Histology Rules.

1. **Code** the **most specific** histology or **subtype/variant, regardless of whether it is described as:**
   A. The majority or predominant part of tumor
   B. The minority of tumor
   C. A component
   *Example 1:* Diagnosis for a single tumor is adenocarcinoma 8140 with the majority or predominant part of tumor being enteric-type adenocarcinoma 8144. Code the subtype/variant: enteric-type adenocarcinoma 8144.
   *Example 2:* Diagnosis for a single tumor is squamous cell carcinoma 8070 with minority of tumor being spindle cell squamous cell carcinoma 8074. Code the subtype/variant: spindle cell squamous cell carcinoma 8074.
   *Example 3:* Diagnosis for a single tumor is sarcoma NOS 8800/3 with a component of leiomyosarcoma 8890/3. Code the subtype/variant: leiomyosarcoma 8890/3.

   *Note 1:* The terms above (A, B, C) must describe a **carcinoma** or **sarcoma** in order to code a histology described by those terms.
   *Example:* When the diagnosis is adenocarcinoma with an enteric-type **adenocarcinoma** component, code enteric-type adenocarcinoma 8144.
   *Negative Example:* When the diagnosis is simply adenocarcinoma with an enteric-type component, code adenocarcinoma NOS 8140. Do not assume this is enteric-type adenocarcinoma. This could be enteric-type differentiation or features.

   *Note 2:* When the most specific histology is described as differentiation or features, see #2.

2. Code the histology described as **differentiation** or **features/features of ONLY** when there is a specific ICD-O code for the "NOS with ____ features" or "NOS with ____ differentiation".
   *Note:* Do not code differentiation or features when there is no specific ICD-O code.

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# UNKNOWN HEAD AND NECK PRIMARY – UNDERSTANDING THE RATIONALE & CRITERIA TO ABSTRACT THESE CASES

**Table I**

First echelon lymph nodes for various primary sites

| | |
|---|---|
| Level 1 | Oral cavity, oropharynx |
| Level 2 | Oral cavity, oropharynx, larynx, nose, hypopharynx, parotid, nasopharynx |
| Level 3 | Oral cavity, oropharynx, larynx, hypopharynx, thyroid, nasopharynx |
| Level 4 | Larynx, thyroid, hypopharynx, oesophagus |
| Level 5 | Nasopharynx, hypopharynx, thyroid, oropharynx |
| Level 6 | Thyroid, larynx, hypopharynx, cervical oesophagus |

It should be also noted that patients presenting with supraclavicular lymphadenopathy may represent a different clinical entity, due to the potential for association with infra-clavicular neoplasms, such as lung cancer.

The first echelon lymph node or nodes, which are involved in SCC can act as an indicator for the potential origin of the index primary are shown here in Table I.

The lumps are usually located in level 2, followed by level 3, with bilateral involvement and other symptoms (i.e. pain and dysphagia) reported in less than 10 per cent.

The presence of cystic malignant metastases in level 2 is often considered to be a hallmark of human papilloma virus (HPV)-related squamous carcinoma, usually with subclinical primaries in the oropharynx.

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# UNKNOWN HEAD AND NECK PRIMARY – UNDERSTANDING THE RATIONALE & CRITERIA TO ABSTRACT THESE CASES

- CRITERIA FOR UNKNOWN PRIMARY TUMORS OF THE HEAD & NECK
  - Cervical Lymph Nodes Only Involved – no supraclavicular, solid organ, or other N3 or Distant Nodes Involved
  - Squamous Cell Carcinoma and Salivary Gland Carcinoma
  - DO NOT CODE C76.0 for any HPV-related oropharynx cancer, nasopharynx cancer, melanoma, thyroid carcinoma, sarcoma
  - **CANNOT BE EBV or HPV RELATED CANCER – MUST BE TESTED**
  - Allowed Specified Histologic Types are Annotated in Chapter 6 of AJCC Cancer Staging Manual
- KNOW YOUR LYMPH NODE REGIONS AND LEVELS FOR REFERENCE – ALWAYS.
  - Know Which Nodes were Removed, the Number of Nodes Examined, the Number of Nodes Positive, and the Size of Positive Nodes
  - Know if only FNA was Done and Follow Instructions for Scope of Surgery and Counting Nodes Pos/Nodes Exam
  - Note if any of the Lymph Nodes show Extra-Nodal Extension (ENE) and be sure to document and code this.
  - Cystic Malignant Nodal Mets in Level 2 nodes are considered a hallmark of HPV-related Squamous Cell Carcinoma
- If your case meets all criteria and no primary tumor can be found and no history of head and neck cancer – then this case qualifies as a true Unknown Primary Tumor of Head & Neck with (ONLY) positive Cervical Lymph Node Involvement.

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- Code primary site to C76.0 when cases meet all criteria – do not use code C14.8, C80.9, C02.8 or C08.8 as primary site code

# STAGING HEAD AND NECK CANCERS
# TIPS FOR AJCC TNM, SEER SUMMARY STAGE 2000 AND CODING THE VERY CONFUSING SSDIS

- ALWAYS FOLLOW THE BASICS – Clinical, Pathological, Tumor Size, Extension, Clinical and Pathological Nodes, Node Mobility, Size of Nodes, Extra Nodal Extension, Location of Nodes both Clinical and Pathological (are they regional or distant), Degree of Extra Nodal Extension, Grade(s), Post-Treatment Clinical, Post-Treatment Pathological, HPV
- ALWAYS FOLLOW THE BASICS – In-situ, Localized, Regional Direct Extension, Regional Nodes, Distant, Grades, HPV
- ALWAYS FOLLOW THE BASICS – Tumor Size, Extension, Grade, Regional Nodes, Metastasis, HPV
- PAY ATTENTION TO CLINICAL AND RESECTED METS – size, number, matting, laterality, mobility, resectability

- **Note 1:** There is evidence that human papilloma virus (HPV) plays a role in the pathogenesis of some cancers. HPV testing may be performed for prognostic purposes; testing may also be performed on metastatic sites to aid in determination of the primary site.
- **Note 2:** Record the results of any HPV testing performed on pathological specimens including surgical and cytological (from cell blocks) tissue from the primary tumor or a metastatic site, including lymph nodes. Do not record the results of blood tests or serology.
- **Note 3:** There are several methods for determination of HPV status. The most frequently used test is IHC for p16 expression which is surrogate marker for HPV infection. **Do not record the results of IHC p16 expression in this field.** The rest of the tests (based on ISH, PCR, RT-PCR technologies) detect the viral DNA or RNA. **This data item is only for HPV status determined by tests designed to detect viral DNA or RNA.**
- **Note 4:** HPV-type 16 refers to virus type and is different from p16 overexpression (p16+)

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# HEAD AND NECK – SITE SPECIFIC DATA ITEMS

- **Chapter 7 – Oral Cavity**
  - **00071:** Lip
  - **00072:** Tongue Anterior
  - **00073:** Gum
  - **00074:** Floor of Mouth
  - **00075:** Palate Hard
  - **00076:** Buccal Mucosa
  - **00077:** Mouth Other
- Chapter 6 – Cervical Lymph Nodes and Unknown Primary
  - 3926: Schema Discriminator 1: Occult Head and Neck Lymph Nodes (Primary site C760 only)
  - 3831: Extranodal Extension Head and Neck Clinical
  - 3832: Extranodal Extension Head and Neck Pathological
  - 3876: LN Head and Neck Levels I-III
  - 3877: LN Head and Neck Levels IV-V
  - 3878: LN Head and Neck Levels VI-VII
  - 3879: LN Head and Neck Other
  - 3883: LN Size

- Chapter 7 – Oral Cavity
  - 3831: Extranodal Extension Head and Neck Clinical
  - 3832: Extranodal Extension Head and Neck Pathological
  - 3883: LN Size
- Chapter 6 – Cervical Nodes and Unknown Primary
  - 3926: Schema Discriminator 1: Occult Head and Neck Lymph Nodes (Primary site C760 only)
  - 3831: Extranodal Extension Head and Neck Clinical
  - 3832: Extranodal Extension Head and Neck Pathological
  - 3876: LN Head and Neck Levels I-III
  - 3877: LN Head and Neck Levels IV-V
  - 3878: LN Head and Neck Levels VI-VII
  - 3879: LN Head and Neck Other
  - 3883: LN Size

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# HEAD AND NECK – SITE SPECIFIC DATA ITEMS

- Chapter 8 – Major Salivary Glands
  - **00080:** Major Salivary Glands
- Chapter 10 – Oropharynx HPV-Mediated (p16+)
  - 00100: Oropharynx HPV-Mediated (p16+)
- Chapter 11 – Oropharynx HPV-Negative (p16-)
  - 00111: Oropharynx
  - (p16-)

- Chapter 8 – Major Salivary Glands
  - 3831: Extranodal Extension Head and Neck Clinical
  - 3832: Extranodal Extension Head and Neck Pathological
  - 3883: LN Size
- Chapter 10 – Oropharynx HPV-Mediated (p16+)
  - 3926: Schema Discriminator 1 (Nasopharynx/PharyngealTonsil)
  - 3927: Schema Discriminator 2 (Oropharyngeal p16+)
  - 3831: Extranodal Extension Head and Neck Clinical
  - 3832: Extranodal Extension Head and Neck Pathological
  - 3883: LN Size
- Chapter 11 – Oropharynx HPV-Negative (p16-)
  - 3926: Schema Discriminator 1 (Nasopharynx/PharyngealTonsil)
  - 3927: Schema Discriminator 2 (Oropharyngeal p16-)
  - 3831: Extranodal Extension Head and Neck Clinical
  - 3832: Extranodal Extension Head and Neck Pathological
  - 3883: LN Size

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# H&N BIOPSY OF REGIONAL LYMPH NODE

- When is it treatment and when is it not treatment? Surg/Rad and Surg/Syst Sequence
  - Starting 2021 the 01 FNA/Core node biopsy is finally not counted as surgery.
- When do you code the lymph node procedure and when not?
  - Be sure it is a regional not distant node before you code it.
- What about the CoC fields for Sentinel Lymph Node Removal?
  - Follow the CoC instructions
- How do you code lymph nodes examined and lymph nodes positive
  - 00 none examined 98 none postiive
- 95 FNA/Core examined 00 positive if the FNA/Core is Neg –
  - You may not have to code procedure
  - 95 FNA/Core examined 95 positive if the FNA/Core is POSITIVE
  - Count otherwise

- Was the Biopsy of a REGIONAL node or Distant Node?
- Type of Biopsy – Scope Reg LN Surg
- 01 - FNA/Core
- 02 – Excisional
- 4, 5, 6 Lymphadenectomy

- Which Dates to Fill in if only a node biopsy?
- When do you fill in both RX Date Surg and Most Definitive
- When do you fill in Surg Oth/Reg/Dist

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# NEW & EMERGING TUMOR MARKERS AND MOLECULAR GENETIC TESTING FOR TARGETING TREATMENTS

CAP Protocols for Head and Neck Biomarker Reporting

1. HPV Testing
    1. P16 Expression (IHC)
    2. HPV-DNA by (ISH)
    3. HPV E6/E7 mRNA (ISH)
    4. HPV DNA (PCR)
    5. HPV E6/E7 mRNA (RT-PCR)
2. EBV Testing
    1. EBV Early mRNA (EBER) (ISH)
    2. NUT Midline Carcinoma
    3. NUT Expression (IHC)
    4. NUT Rearrangements (FISH)
    5. BRD4-NUT Fusion (RT-PCR
    6. Other NUT Fusion (RT-PCR)
3. Salivary Gland Carcinoma
    1. EWSR1 Rearrangements (FISH)
    2. EWSR1-ATF1 Fusion (RT-PCR)
    3. Other EWSR1 Fusion (RT-PCR)

- Mammary Analogue Secretory Carcinoma
    - ETV6 Rearrangements (FISH)
    - ETV6-NTRK3 Fusion (RT-PCCR)
- Mucoepidermoid Carcinoma
    - MEML2 Rearrangements (FISH)
    - CRTC1-MAML2 Fusion (RT-PCR)
    - CRTC3-MAML2 Fusion (RT-PCR)
- Adenoid Cystic Carcinoma
    - MYB Expression (IHC)
    - MYB Rearrangements (FISH)
    - MYB-NFIB Fusion (FISH)
- Salivary Duct Carcinoma
    - HER2 (ERBB23) – IHC
    - HER2 (ERBB2) – FISH
    - Androgen Receptor (IHC)

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# STANDARD TREATMENT REGIMENS & GUIDELINES

- Oral cavity and oropharyngeal cancers that are linked with HPV10 tend to have a better outcome than those that are HPV negative (HPV-). Clinical trials are ongoing to examine type and amount of chemo and/or radiation to administer for HPV+ versus HPV- cancers without reducing survival based on p16.
- TUMOR FOCALITY MAY BE AN IMPORTANT DECISION TREE FOR ANY HEAD AND NECK CANCER
- KEY FACTORS – LOCION and SIZE OF PRIMARY TUMOR, HISTOLOGY, NUMBER and SIZE OF LYMPH NODES, EXTRANODAL EXTENSION, SYMPTOMS, PATIENT STATUS, OTHER OPTIONS AVAILABLE
- SURGERY – COMPLETE EXCISION OF PRIMARY TUMOR WITH NODES SAMPLED/NECK DISSECTION
- LYMPH NODE PROCEDURE – ASSESSES LEVEL, LOCATION, SIZE, EXTRANODAL EXTENSION, ETC.
- RADIATION – High Dose - NEW TECHNIQUES TARGET THE FOCUS OF THE RADIATION BEAM OR Brachytherapy
- CHEMOTHERAPY – Platinum-based agents – cisplatin/carboplatin, 5FU, Cytoxan or cyclophoasphamide, docxorubicin (Adriamycin), docetaxel (Taxotere), methotrexate, Bleomycin, Hydroxyurea
- IMMUNOTHERAPY – erlotiinib (Tarceva), bevacizumab (Avastin), Cetuximab, keytruda, Nivolumab
- PDT – photodynamic therapy - this involves using a drug that cancer cells absorb. The drug is then exposed to special laser light source which “turns it on” so the drug then kills the cancer cells. PDT is less damaging than surgery and may be used in earl7y cancers and recurrent cancers in H&N.

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# NEW & EMERGING TREATMENTS AND TECHNIQUES

- Treatment in a clinical trial1 should be considered for any type or stage of cancer in the head and neck areas. This way people can get the best treatment available now and may also get the new treatments that are thought to be even better.
- TP53 gene mutation - The protein produced by this gene (p53) normally helps keep cells from growing too much and helps to destroy cells that are too damaged to be fixed. Changes in the TP53 gene can lead to increased growth of abnormal cells and cancer. Recent studies suggest that tests to find these gene changes might help find oral and oropharyngeal cancers early.
- Immunotherapy – pembrolizumab (Keytruda) – approved mid-2019 for first line metastatic/unresectable, Cetuximab or Erbitux was approved in 2006, Nivolumab following progression on platinum-based therapy, Nivolumab has also been approved along the same lines.
- Prevention – for people at high risk - Tarceva (Erlotinib) keeps the receptor EGFR from signaling cells to grow which may prevent cancers or treat early cancers or pre-cancers from becoming malignant. Metformin may have similar action to help keep pre-cancerous changes from turning into cancers.
- Prevention – Gardisil 9 – HPV Vaccine approved on 6/9/2020 to prevent HPV infection thus preventing H&N cancers

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# QUESTIONS


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