2018 Updates for Neoplasms of the Thyroid

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FLccSC LMS – CEU Quiz – FCDS IDEA

- 2017 - Florida Changed How FCDS Awards CEUs for FCDS Webcasts
- 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

2018 - A Year for Major Changes to Cancer Registry Data Standards

- New ICD-O-3 Histology Code & Behavior Codes
- New Histology Coding Rules and Tools
- New Reportable Cancers
- 2018 Solid Tumor MP/H Rules
- 2018 Hematopoietic MP/H Rules
- Cancer Staging Updates
  - SS2018
  - Grade Coding
  - Site-Specific Data Items
  - AJCC TNM 8th ed.
  - 2018 SEER EOD
- EDITS v18
- STORE Manual
- 2018 FCDS DAM
2018 - A Year for Major Changes to Cancer Registry Data Standards

Presentation Outline

- Introduction to Neoplasms of the Thyroid
- Anatomy and Physiology of the Thyroid Gland
- Signs, Symptoms and Cancer Risk
- Thyroid Cancer Diagnostic Workup
- Biomolecular & Genetic Testing - Thyroid
- Changes to ICD-O-3 Histology Codes - Thyroid
- 2018 Grade Fields and Codes - Thyroid
- 2018 MP/H Rules - Thyroid - Important!!
- Anatomic Staging & Site-Specific Data Items
- Treatment Guidelines - Thyroid
- Text Documentation
- Practice Cases - Pending
- Questions

http://safetyca.info
Introduction – GLOBOCAN 2018

Source: Global Cancer Statistics 2018

Introduction – United States

Figure 3. Leading Sites of New Cancer Cases and Deaths – 2018 Estimates

American Cancer Society – 2018 Cancer Facts & Figures
Introduction

Thyroid Cancer Statistics - CAUTION

https://statecancerprofiles.cancer.gov/
Why Thyroid Cancer and Why Now ???

Anatomy of Thyroid Gland

Source: NIH and Terese Windslow LLC, 2012
Anatomy of Thyroid Gland

Anatomy of Parathyroid Glands
Anatomy of Thyroid - Lymphatics

- Patterns of Nodal Mets for Thyroid Cancers begin in the Central Compartment VI then involve the Lateral Compartments III and IV then most commonly to the Contralateral Neck – different than most H&N cancers which have varying patterns of nodal spread.

Signs, Symptoms and Risk Factors

**Symptoms – Thyroid**

- Lump found in the neck
- Swelling in the neck
- Pain in the front of the neck
- Persistent hoarseness or voice changes
- Swallowing difficulty
- Persistent cough not due to a cold
Benign Nodules and Thyroid Goiter

- Goiter – An abnormally large thyroid gland – some are diffuse enlarged and some nodular with one or more nodules present.
- Many reasons for development of goiter - most are not cancer.
- Main reason for development of a goiter is hormone imbalance due to nutritional deficit (diet low in iodine) or iodine absorption related disease.
- Most nodules are benign cysts filled with colloid – about 17/18 of every 20, and even most solid nodules are benign.
- Most nodules that overproduce hormones are benign.
- So, most thyroid cancers present with few symptoms other than a lump in the neck or incidental findings in workup for another medical problem – not because of symptoms.

Cold/Warm or Hot Nodule

- A cold nodule indicates that the cells within the nodule do not produce thyroid hormone resulting in no uptake of the iodine contrast when a thyroid scan is performed.
- About 85% of thyroid nodules are cold nodules.
- About 85% of cold nodules are benign.
- About 10% of thyroid nodules are warm nodules.
- About 90% of warm nodules are benign.
- A hot nodule is a nodular region that takes up large amounts of radioactive iodine relative to the rest of the thyroid gland on thyroid scan, hence it is visualized as a “hot spot” on the scan.
- Hot nodules indicate a over-active thyroid gland.
- Hot nodules are rarely cancerous.
- Hot nodules make up about 5% of findings
- About 95% of hot nodules are benign.
Multiple Endocrine Neoplasia (MEN)

- Family history of Hashimoto's thyroiditis
- Family history of benign thyroid nodule or goiter
- Symptoms of hyperthyroidism or hypothyroidism
- Pain or tenderness associated with a nodule
- A soft, smooth, mobile nodule
- Multi-nodular goiter without a predominant nodule (lots of nodules, not one main nodule)
- "Warm" nodule on thyroid scan (produces normal amount of hormone)
- Simple cyst on an ultrasound
Suspect Malignant Disease

- Age less than 20
- Age greater than 70
- Male gender
- New onset of swallowing difficulties
- New onset of hoarseness
- History of external neck irradiation during childhood
- Firm, irregular, and fixed nodule
- Presence of cervical lymphadenopathy (swollen, hard lymph nodes in the neck)
- Previous history of thyroid cancer
- Nodule that is "cold" on scan (shown in picture above, meaning the nodule does not make hormone)
- Solid or complex on an ultrasound

Cancer Screening - Thyroid

- November 2016 AHRQ Recommendations to USPSTF: “Although ultrasonography of the neck using high-risk sonographic characteristics plus follow-up cytology from fine needle aspiration can reasonably identify thyroid cancers, it is unclear if population-based or targeted screening can decrease mortality or improve important patient health outcomes.”
- “More importantly, screening results in the identification indolent thyroid cancers, and treatment of these over-diagnosed cancers can pose real patient harms.”
Over-Diagnosis of Thyroid Cancer

- Over-diagnosis of thyroid cancers contributes to increased incidence of thyroid cancers worldwide, which is already a serious public health problem.
- A great number of medical tests, helping to detect thyroid cancer, may result in an epidemic of diagnosis.
- A dramatic increase in the number of detected cases of thyroid cancer may be associated with a great number of neck ultrasounds, fine-needle aspirations and incidental findings during examination.
- Over-diagnosis can lead to overtreatment.
- Many patients undergoing surgery and receiving radioactive iodine as part of their therapy may never have developed clinical disease.
- Mortality due to this pathology remains relatively stable.
- It is important to differentiate stationary cancers from potentially aggressive diseases.


Diagnostic Workup - Thyroid

- Physical exam and history
- Laryngoscopy
- Blood hormone studies
- Blood chemistry studies
- Ultrasound exam
- Computed tomography scan
- Fine-needle aspiration biopsy of the thyroid
- Surgical excision
- Gene Panel

Source: Textbook – Surgical Oncology – Thyroid Cancer, pp 539-561
Most Common Types of Thyroid Cancer

- **Papillary Thyroid Cancer**
  - 75% to 85% of all thyroid cancers
  - Histology often miscoded by registrars who don’t know the rules to code for papillary thyroid cancers
  - Women in the 20-55-year age group are more likely to get thyroid cancer than men.
  - Children with previous history of thyroid cancer children suffering from thyroid cancer
  - Patients who have undergone previous radiation to thyroid. These tumors are still often still well-differentiated and slow-growing.

- **Follicular Thyroid Cancer**
  - 15% of thyroid cancer cases.
  - Women over 50 are more likely to get thyroid cancer than men.
  - Thyroglobulin, a tumor marker, for 50% of all the proteins of the thyroid gland.
  - Thyroglobulin can be used as a tumor marker for well-differentiated follicular thyroid cancer.

Less Common Types of Thyroid Cancer

- **Medullary Thyroid Cancer**
  - 3% of all thyroid cancer cases.
  - **Familial MTC** - RET proto-ongene positive cancers a familial form of medullary thyroid cancer.
  - **Sporadic MTC** is not a familial cancer but can be paraneoplastic syndrome in people with MEN2

- **Anaplastic Thyroid Cancer** is a very aggressive form of thyroid cancer with a very poor prognosis
  - 2% of all thyroid cancers
  - They all exhibit aggressive behavior with less than 1 year survival not uncommon
Clinical Features - Epithelial Thyroid Malignancy

Table 1: Clinical Features of Epithelial Thyroid Malignancies

<table>
<thead>
<tr>
<th>Feature and Cell of Origin</th>
<th>PTC</th>
<th>FTC</th>
<th>MTC (C cell)</th>
<th>ATC</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cell of origin</td>
<td>Follicular</td>
<td>Follicular</td>
<td>Parafollicular</td>
<td>Follicular</td>
</tr>
<tr>
<td>Percentage of all thyroid cancers</td>
<td>80%–85%</td>
<td>10%–15%</td>
<td>3%–5%</td>
<td>1%–2%</td>
</tr>
<tr>
<td>Most common age group</td>
<td>Third to fourth decade</td>
<td>Fourth to sixth decade</td>
<td>Fourth to sixth decade</td>
<td>&gt;65 years</td>
</tr>
<tr>
<td>Gender predilection (female-to-male ratio)</td>
<td>2.5:1</td>
<td>3:1</td>
<td>3:2</td>
<td>3:1</td>
</tr>
<tr>
<td>Familial inheritance</td>
<td>5%</td>
<td>5%</td>
<td>25%</td>
<td>...</td>
</tr>
<tr>
<td>Common sites of metastasis</td>
<td>Lymph nodes</td>
<td>Lungs and bone</td>
<td>Liver</td>
<td>Lungs</td>
</tr>
<tr>
<td>Prognosis (10-year survival)</td>
<td>95%–98%</td>
<td>90%–95%</td>
<td>60%–80%</td>
<td>&lt;10%</td>
</tr>
</tbody>
</table>


Revised Classifications

- Papillary Neoplasms of Thyroid – remove the word ‘carcinoma’ from the diagnosis – entirely
- EFVPTC – encapsulated follicular variant of papillary thyroid carcinoma – now 10-20% of dx
- NIFTP – noninvasive follicular thyroid neoplasm with papillary-like nuclear features
- Will affect care and treatment of more than 45,000 patients every year – worldwide
- NCI and CDC will be monitoring changes to new terminology and treatment guidelines very closely
Inadequate FNA Specimen & Gene Testing

- Approximately 20% of fine-needle aspirations (FNA) of thyroid nodules have indeterminate cytology, most frequently Bethesda category III or IV.
- Diagnostic surgeries can be avoided for these patients if the nodules are reliably diagnosed as benign without surgery.
- A new multigene genomic classifier test (112-gene test) demonstrated a high sensitivity and reasonably high specificity when identifying cancer-related point mutations, gene fusions and gene expression alterations.
- Negative test results may avoid diagnostic surgery in up to 61% of patients with Bethesda III to IV indeterminate nodules, and up to 82% of all benign nodules with indeterminate cytology.
- Information on specific genetic alterations obtained from FNA may help inform individualized treatment of patients with a positive test result.

Inadequate FNA Specimen & Gene Testing


Genomic Taxonomy of Thyroid Cancers

CAP Biomarker Checklist – Thyroid

- BRAF Mutation (V600E, K601E, Other)
- TERT Mutation (1-124 or 1-146, Other)
- NRAS Mutation – Q61R
- HRAS Mutation – Q61R, G12V
- KRAS Mutation – G12D
- AKT1 Mutation – E17K
- TP53 Mutation
- PIK3CA Mutation – H1047R
- CTNNB1 Mutation – S33a
- RET Mutation – M918T and Mutation Type
- ALK Rearrangement – STRN/ALK or EML4/ALK
- NTRK1 Rearrangement – NTRK1/TPM3 or NTRK1/TFG
- NTRK3 Rearrangement – NTRK3/ETV6
- RET Rearrangement – RET/PTC1 or RET/PTC3
- PPAR gamma Rearrangement – PAX8/PPAR gamma or CREB3L2/PPAR gamma
- Other Markers

ICD-O-3 Updates - 2018

ICD-O-3 Coding Resources

- ICD-O-3 Manual – use your current manual
- ICD-O-3 Errata & 2011 Updates
  - http://www.who.int/classifications/icd/updates/icd03updates/en/
- ICD-O-3 Updates for 2018
- 2018 Solid Tumor MP/H Rules
  - https://seer.cancer.gov/tools/solidtumor
- Hematopoietic Database On Line
  - https://seer.cancer.gov/seertools/hemelymph/
- 2018 Site-Specific Grade Instructions
- 2018 SEER Site/Type Validation List

New Reporting Requirements - Thyroid

- The changes to classification of thyroid neoplasms will result in incorrect and inconsistent coding by medical records and billing departments for thyroid.
- Some may be coded as benign, some malignant, some borderline malignant – wide variation seen.
- Include benign and borderline ICD-10-CM codes while coders learn about coding new classifications.
  - D34 – benign neoplasm of thyroid gland
  - D44 – neoplasm of uncertain behavior thyroid gland
  - E04.1 – single thyroid nodule
  - E04.2 – multiple thyroid nodules
  - E04.2 – multinodular goiter
ICD-O-3 Updates - Thyroid

<table>
<thead>
<tr>
<th>New code/term</th>
<th>8339/3</th>
<th>Follicular thyroid carcinoma (FTC), encapsulated angioinvasive (C73.9)</th>
<th>Y</th>
</tr>
</thead>
<tbody>
<tr>
<td>New Term</td>
<td>8343/3</td>
<td>Invasive encapsulated follicular variant of papillary thyroid carcinoma (invasive EFVPTC) (C73.9)</td>
<td>Y</td>
</tr>
<tr>
<td>New Term</td>
<td>8343/3</td>
<td>Encapsulated follicular variant of papillary thyroid carcinoma, NOS (EFVPTC, NOS) (C73.9)</td>
<td>Y</td>
</tr>
<tr>
<td>New Term</td>
<td>8345/3</td>
<td>Medullary thyroid carcinoma (C73.9)</td>
<td>Y</td>
</tr>
</tbody>
</table>

Grade Coding Manual

https://apps.naaccr.org/ssdi/list/
Grade Coding Manual – Schema ID

Clinical Grade - the grade of a solid primary tumor before any treatment. Treatment may include surgical resection, systemic therapy, radiation therapy, or neoadjuvant therapy. NOTE: All surgical procedures are not treatment, e.g. TURB and endoscopic biopsies.

Pathological Grade - the grade of a solid primary tumor that has been surgically resected and for which no neoadjuvant therapy was administered. If AJCC pathological staging is being assigned, the tumor must have met the surgical resection requirements in the AJCC manual. This may include the grade from the clinical workup, as all information from diagnosis (clinical staging) through the surgical resection is used for pathological staging.

Post-Therapy Grade - the grade of a solid primary tumor that has been resected following neoadjuvant therapy. If AJCC post-therapy staging is being assigned, the tumor must have met the surgical resection requirements for yp in the AJCC manual. Neoadjuvant therapy must meet guidelines or standards, and not be that given for variable or unconventional reasons as noted in the AJCC manual.
2018 Grade - Parathyroid

Grade 25

<table>
<thead>
<tr>
<th>Schema ID</th>
<th>Schema ID Name</th>
<th>ACR ID</th>
<th>ACR Chapter</th>
</tr>
</thead>
<tbody>
<tr>
<td>Parathyroid</td>
<td>75</td>
<td>Parathyroid</td>
<td></td>
</tr>
</tbody>
</table>

Note 1: Clinical grade is not listed.
Note 2: Assign the highest grade from the primary tumor assessed during the clinical time frame.
Note 3: Code C and D take priority over A-D.
Note 4: Code 9 when:
- Grade from primary site is not documented
- Clinical workup is not done (for example, cancer is an incidental finding during surgery for another condition)
- Grade checked "not applicable" on CAP Protocol (if available) and no other grade information is available
Note 5: If there is only one grade available and it cannot be determined if it is clinical, pathological, or after non-adjacent therapy, assign as a clinical grade or code unknown (U) if pathological grade, and blank for post-thyroid grade.

<table>
<thead>
<tr>
<th>Code</th>
<th>Grade Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>2</td>
<td>Low-grade round monomorphic nuclei with only mild to moderate nuclear size variation, indistinct nuclei, and chromatin characteristics resembling those of normal parathyroid or adrenals</td>
</tr>
<tr>
<td>3</td>
<td>High-grade, more pleomorphic, with a nuclear size variation greater than 4.0; prominent nuclear membrane irregularities; chromatin alterations, including hyperchromasia or margination of chromatin, and prominent nucleoli; high-grade tumors show several discrete confluent areas with nuclear changes</td>
</tr>
<tr>
<td>A</td>
<td>Well-differentiated</td>
</tr>
<tr>
<td>B</td>
<td>Moderately differentiated</td>
</tr>
<tr>
<td>C</td>
<td>Poorly differentiated</td>
</tr>
<tr>
<td>D</td>
<td>Undifferentiated, anaplastic</td>
</tr>
<tr>
<td>0</td>
<td>Grade cannot be assigned. Unknown</td>
</tr>
</tbody>
</table>

2018 Solid Tumor MP/H Rules

Published June 2018 – but still had MAJOR changes in October 2018 – be sure you have the correct set of rules

Solid Tumor Rules
Effective with Cases Diagnosed 1/1/2018 and Forward

Published June 2018

Editors:
- Lois Dicker, CTR, NCIA/SER
- Carol Hama Johnson, BS, CTR (Retired), Consultant
- Suzanne Adams, BA, CTR (DIT, Inc.)
- Sarika Nagaria, MD, PhD, CTR, NCIA/SER

Suggested citation:
General Instructions

- TEXT ONLY RULES INCLUDE:
  - General Instructions PLUS
  - 10 Sets of Solid Tumor MP/H Rules
  - Each Module includes Multiple Sections (Notes/Site/MP/Histology)
- Code subtypes/variants when definitively described (no modifiers)
- Do Not Use Ambiguous Terminology to Code Histology
- Ambiguous terminology is used to determine “case reportability”
- Ambiguous terminology is not to be used to determine histology
- Ambiguous terminology such as “with features of”, etc. are no longer used to determine a subtype OR to determine which histology should be coded. See the following histology rules for instructions on coding multiple histologies.
- Use the Histology (H) Rules to determine when to use or not use any ambiguous terminology when an ambiguous term is used to describe a histologic type -- sometimes you use the ambiguous term to code a subtype or variant or mixed histology -- and sometimes you do not.

General Instructions

- Introduction
- Changes from 2007 MPH Rules
- Definitions
- Equivalent or Equal Terms
- Terms that are NOT Equivalent or Equal
- Table and Instructions for Coding Primary Site
- Table: Specific Histologies, NOS and Subtypes
- Table: Combination/Mixed Histology Codes
- Table: Histologies Not Reportable for This Site
- Illustrations
- Multiple Primary Rules
- Histology Coding Rules
General Instructions

How to Use the Solid Tumor Rules

1. Use the following site-specific rules for tumors diagnosed 1/1/2018 and forward:
   - Malignant CNS and Peripheral Nerves
   - Head and neck
   - Non-Malignant CNS
   - Kidney
   - Breast
   - Cervix
   - Urinary sites

2. Use the following site-specific rules for tumors diagnosed 1/1/2007 through 12/31/2018:
   - Non-malignant melanoma of the skin (not updated for 2018)
   - Other sites (not updated for 2018) for solid tumors which occur in primary sites not covered by the site-specific rules.

3. 2007 MPR Rules and 2018 Solid Tumor Rules are used based on date of diagnosis.
   - Tumors diagnosed 01/01/2007 through 12/31/2017: Use 2007 MPR Rules
   - Tumors diagnosed 01/01/2018 and later: Use 2018 Solid Tumor Rules

4. An original tumor diagnosed before 1/1/2018 and a subsequent tumor diagnosed 01/1/2018 or later in the same primary site: Use the 2018 Solid Tumor Rules.

5. The Solid Tumor Rules are not used to determine case reportability, stage, or tumor grade.

General Instructions

How to Use the Histology Rules

1. Rules are divided into two sections: Single Tumor and Multiple Tumors. Abstracted as a Single Primary
   - Each section is a complete set of rules.

2. Within each section, the rules are hierarchical. Use the first rule that applies and STOP. Do not continue through the rules.
   - A list of terms which cannot be used to code histology unless they are in the histology rules.
   - Do not code histologies or subtypes not described by ambiguous terms.

3. Most likely:
   - Most likely
   - Probable
   - Suspected
   - Consistent with
   - Apparent

4. Stop:
   - Apparent
   - Consistent with

5. Note: Histology described by ambiguous terminology is coded as "X" when a case is accessioned based on ambiguous terminology and no other histology information is available documented.
Multiple Primary Rules – Remember: Most People Have Only One Cancer

Some People or Their Families Have More Than One Cancer

https://www.curetoday.com/journey/cancer-guides/at-diagnosis/
### 2018 Solid Tumor Rules – Other Sites

<table>
<thead>
<tr>
<th>Column 1: Required Histology</th>
<th>Column 2: Combined with Histology</th>
<th>Column 3: Combination Term</th>
<th>Column 4: Code</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gyn malignancies with two or more of the histologies in column 2</td>
<td>Clear cell Endometroid Mucinous Papillary Serous Squamous Transitional (Bruner)</td>
<td>Mixed cell adenocarcinoma</td>
<td>8323</td>
</tr>
<tr>
<td>Papillary and Follicular</td>
<td>Follicular</td>
<td>Papillary carcinoma, follicular variant</td>
<td>8340</td>
</tr>
<tr>
<td>Medullary</td>
<td>Papillary</td>
<td>Mixed medullary-follicular carcinoma</td>
<td>8346</td>
</tr>
<tr>
<td>Medullary</td>
<td>Papillary</td>
<td>Mixed medullary-papillary carcinoma</td>
<td>8347</td>
</tr>
</tbody>
</table>

**Rule M6 Follicular and papillary** tumors in the thyroid within 60 days of diagnosis are a single primary. *

**Rule M10** Tumors diagnosed more than one (1) year apart are multiple primaries.

**Rule M15** An invasive tumor following an in situ tumor more than 60 days after diagnosis is a multiple primary.

**Rule M17** Tumors with ICD-O-3 histology codes that are different at the first (xxxx), second (xxxx) or third (xxxx) number are multiple primaries.

### 2018 Solid Tumor Rules - Thyroid

<table>
<thead>
<tr>
<th>Column 1: Required Histology</th>
<th>Column 2: Combined with Histology</th>
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<td>Papillary</td>
<td>Mixed medullary-papillary carcinoma</td>
<td>8347</td>
</tr>
</tbody>
</table>

**Rule H14** Code papillary carcinoma of the thyroid to papillary adenocarcinoma, NOS (8260).

**Rule H15** Code follicular and papillary carcinoma of the thyroid to papillary carcinoma, follicular variant (8340).

**Rule H30** Code the appropriate combination/mixed code (Table 2) when there are multiple specific histologies or when there is a non-specific histology with multiple specific histologies.
2018 SEER Summary Stage

Purpose of Staging
- Biochemical Tumor Markers
- Molecular Tumor Markers
- Genetic Mutations/Variations
- Risk Stratification

Thyroid Cancer Staging – SS2018

ENDOCRINE SYSTEM
THYROID
- 6005.8700, 8720.8790, 9706.8701
- C739
- C740 Thyroid gland

SUMMARY STAGE
- Localized only (localized, NOS)
- Confined to thyroid, NOS
- Into thyroid capsule, but not beyond
- Multiple fixed confined to thyroid

3 Regional lymph node(s) involved only
- Level I
  - Level IA - Submandibular (submandibular), sublingual
  - Level IB - Submandibular (submandibular)
- Level II - Upper jugular
  - Jugulodigastric (submaxillary)
  - Upper deep cervical
- Level III - Middle jugular
  - Middle deep cervical
- Level IV - Lower jugular
  - Jugulo-cervical (jugulo-cervical)
  - Nerve deep cervical
  - Vertebrobasilar
- Level V - Pectoral triangle group
  - Pectoral deep cervical
  - Level VA - Spinal accessory

Source: SEER Summary Staging Manual 2018
**Thyroid Cancer Staging – SS2018**

- Level VI: Anterior compartment group
  - Larynx
  - Parathyroid
  - Thyroid gland
  - Trachea
  - Skeletal muscle

- Level VII: Superior mediastinal group
  - Esophagus
  - Recurrent laryngeal nerve
  - Trachea
  - Trachea
  - Thyroid gland

- Level VIII: Paratracheal and pretracheal groups

**Parathyroid Cancer Staging – SS2018**

**PARATHYROID**

- SUMMARY STAGE
  - Tumor, intrathyroidal, noninvasive
  - Any size, intrathyroidal, noninvasive
  - Any size, intrathyroidal, noninvasive

- Localized only (localized, NOS)
  - Confined to parathyroid
  - Extension to soft tissue

- Regional by direct extension only
  - Adjacent lymph nodes
  - Esophagus
  - Parathyroid
  - Thyroid gland

- Regional by both direct extension and regional lymph nodes involved
  - Codes (2) + (3)

- Distant site(s) lymph node(s) involved
  - Distant node(s) (including further contiguous extension)
    - Gross extrathyroidal extension involving
      - Bone
      - Mediastinal tissues

- Parathyroid fossa
- Skeletal muscle, other than strap or sternocleidomastoid muscle
- Distant lymph node(s), NOS
- Distant metastasis, NOS
  - Carcinomatous
  - Distant mets WITH OR WITHOUT distant lymph node(s)

- Unknown if extension or metastasis
ParaThyroid Cancer Staging – SS2018

- Deep cervical, NOS
- Facial
  - Buccal (buccal)
  - Mandible
  - Nasal
- Internal jugular, NOS
- Parapharyngeal
- Parotid
  - Intracanalicular
  - Intracranial
  - Periglandular
  - Parotid extracanicular
- Reteonuncular (manoid)
- Retrostapedial
- Subcapsular
  - Regional lymph node(s), NOS
  - Lymph node(s), NOS

4 Regional by ROTH direct extension AND regional lymph node(s) involved

- Codes (2) + (3)

Thyroid Staging – AJCC TNM Criteria

- Chapter 73 – Differentiated & Anaplastic Carcinoma
  - Papillary Carcinoma (8260/3)
  - Follicular Carcinoma (8330/3)
  - Hurthle Cell Carcinoma (8290/3)
  - NIFTP (8343/2) – non-invasive
  - EFVPTC (8343/2) – non-invasive
  - Anaplastic/Undifferentiated Carcinoma (8020/3)

- Chapter 74 – Medullary Carcinoma
  - Medullary Carcinoma (8345/3)
  - Mixed Medullary and Follicular Carcinoma (8346/3)
  - Mixed Medullar and Papillary Carcinoma (8347/3)

- Chapter 75 – Parathyroid
  - See List in AJCC Manual
Thyroid Staging – AJCC TNM Criteria

- **AJCC Prognostic Stage Group is Further Divided by:**
  - **Histology**
    - Papillary & Follicular
    - Anaplastic/Undifferentiated
    - Medullary
    - Mixed Medullary
  - **Behavior**
    - Non-Invasive Histology (/2)
    - Invasive Histology (/3)
  - **Age at Diagnosis**
  - **Restaged at Recurrence for Risk Assessment**

---

Major Changes - Thyroid

**Table 1. Major Changes to the AJCC/TNM Staging of Differentiated and Anaplastic Thyroid Cancers in the Eighth Edition**

<table>
<thead>
<tr>
<th>Differentiated</th>
<th>1. The age cutoff used for staging was increased from 45 to 55 years of age at diagnosis.</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>2. Minor extrathyroidal extension detected only on histological examination was removed from the definition of T3 disease and therefore has no impact on either T category or overall stage.</td>
</tr>
<tr>
<td></td>
<td>3. N1 disease no longer upstages a patient to stage III. If &lt;55 years of age at diagnosis, N1 disease is stage I; if ≥55 years of age, N1 disease is stage II.</td>
</tr>
<tr>
<td></td>
<td>4. T3a is a new category for tumors ≥4 cm confined to the thyroid gland.</td>
</tr>
<tr>
<td></td>
<td>5. T3b is a new category for tumors of any size demonstrating gross extrathyroidal extension into strap muscles (sternothyroid, sternothyroid, thyrohyoid, or omohyoid muscles).</td>
</tr>
<tr>
<td></td>
<td>6. Level VI lymph nodes, previously classified as lateral neck lymph nodes (N1b), were reclassified as central neck lymph nodes (N1a) to be more anatomically consistent and because level VI presented significant coding difficulties for tumor registrars, clinicians, and researchers.</td>
</tr>
<tr>
<td></td>
<td>7. In differentiated thyroid cancer, the presence of distant metastases in older patients is classified as stage IVB disease rather than stage IVC disease. Distant metastasis in anaplastic thyroid cancer continues to be classified as stage IVC disease.</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Anaplastic</th>
<th>1. Unlike previous editions where all anaplastic thyroid cancers were classified as T4 disease, anaplastic cancers will now use the same T definitions as differentiated thyroid cancer.</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>2. Intrathyroidal disease is stage IVA, gross extrathyroidal extension or cervical lymph node metastases is stage IVB, and distant metastases are stage IVC.</td>
</tr>
</tbody>
</table>
# Major Changes - Thyroid

<table>
<thead>
<tr>
<th>Distant metastasis</th>
<th>Gross ETE present?</th>
<th>Structures involved with gross ETE</th>
<th>T category</th>
<th>N category</th>
<th>Stage</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;55 years</td>
<td>No</td>
<td>Any or none</td>
<td>Any</td>
<td>Any</td>
<td>I</td>
</tr>
<tr>
<td></td>
<td>Yes</td>
<td>Any or none</td>
<td>Any</td>
<td>Any</td>
<td>II</td>
</tr>
<tr>
<td>≥55 years</td>
<td>No</td>
<td>No</td>
<td>≤4 cm (T1–2)</td>
<td>N0/Nx/N1a</td>
<td>I</td>
</tr>
<tr>
<td></td>
<td>Yes</td>
<td>Only strap muscle (T3b)</td>
<td>Any</td>
<td>Any</td>
<td>II</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Subcutaneous, larynx, trachea,</td>
<td>Any</td>
<td>Any</td>
<td>III</td>
</tr>
<tr>
<td></td>
<td></td>
<td>esophagus, recurrent laryngeal</td>
<td>Any</td>
<td>Any</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>nerve (T4a)</td>
<td>Any</td>
<td>Any</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Prevertebral fascia, encasing</td>
<td>Any</td>
<td>Any</td>
<td>IVA</td>
</tr>
<tr>
<td></td>
<td></td>
<td>major vessels (T4b)</td>
<td>Any</td>
<td>Any</td>
<td>IVB</td>
</tr>
<tr>
<td></td>
<td>Yes</td>
<td>Any or none</td>
<td>Any</td>
<td>Any</td>
<td></td>
</tr>
</tbody>
</table>

## AJCC TNM – T Category Codes

**Definition of Primary Tumor (T)**

*For Papillary, Follicular, Poorly differentiated, Hurthle cell and Anaplastic Thyroid Carcinoma*

<table>
<thead>
<tr>
<th>T Category</th>
<th>T Criteria</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>T1</td>
<td>Tumor ≤ 2 cm in greatest dimension limited to the thyroid</td>
</tr>
<tr>
<td>T1a</td>
<td>Tumor ≤ 1 cm in greatest dimension limited to the thyroid</td>
</tr>
<tr>
<td>T1b</td>
<td>Tumor &gt; 1 cm but ≤ 2 cm in greatest dimension, limited to the thyroid</td>
</tr>
<tr>
<td>T2</td>
<td>Tumor &gt; 2 cm but ≤ 4 cm in greatest dimension limited to the thyroid</td>
</tr>
<tr>
<td>T3*</td>
<td>Tumor &gt; 4 cm limited to the thyroid, or gross extrathyroidal extension invading only strap muscles</td>
</tr>
<tr>
<td>T3a*</td>
<td>Tumor &gt; 4 cm limited to the thyroid</td>
</tr>
<tr>
<td>T3b*</td>
<td>Gross extrathyroidal extension invading only strap muscles (sternohyoid, sternothyroid, thyrohyoid, or omohyoid muscles) from a tumor of any size</td>
</tr>
<tr>
<td>T4</td>
<td>Includes gross extrathyroidal extension into major neck structures</td>
</tr>
<tr>
<td>T4a</td>
<td>Gross extrathyroidal extension invading subcutaneous soft tissues, larynx, trachea, esophagus, or recurrent laryngeal nerve from a tumor of any size</td>
</tr>
<tr>
<td>T4b</td>
<td>Gross extrathyroidal extension invading prevertebral fascia or encasing carotid artery or mediastinal vessels from a tumor of any size</td>
</tr>
</tbody>
</table>

Note: All categories may be subdivided: (s) solitary tumor and (m) multifocal tumor (the largest tumor determines the classification).
### AJCC TNM – N & M Category Codes

**Definition of Regional Lymph Node (N)**

<table>
<thead>
<tr>
<th>N Category</th>
<th>N Criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>NX</td>
<td>Regional lymph nodes cannot be assessed</td>
</tr>
<tr>
<td>N0</td>
<td>No evidence of regional lymph nodes metastasis</td>
</tr>
<tr>
<td>N0a*</td>
<td>One or more cytological or histologically confirmed benign lymph node</td>
</tr>
<tr>
<td>N0b*</td>
<td>No radiologic or clinical evidence of locoregional lymph node metastasis</td>
</tr>
<tr>
<td>N1*</td>
<td>Metastasis to regional nodes</td>
</tr>
<tr>
<td>N1a*</td>
<td>Metastasis to level VI or VII (pretracheal, paratracheal, or prelaryngeal/Delphian or upper mediastinal) lymph nodes. This can be unilateral or bilateral disease.</td>
</tr>
<tr>
<td>N1b*</td>
<td>Metastasis to unilateral, bilateral, or contralateral lateral neck lymph nodes (Levels I, II, III, IV, or V) or retropharyngeal lymph nodes</td>
</tr>
</tbody>
</table>

**Definition of Distant Metastasis (M)**

<table>
<thead>
<tr>
<th>M Category</th>
<th>M Criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>M0</td>
<td>No distant metastasis</td>
</tr>
<tr>
<td>M1</td>
<td>Distant metastasis</td>
</tr>
</tbody>
</table>

### AJCC TNM – Stage Group Derivation

**Differentiated thyroid cancer**

- **When age at diagnosis is...**
  - < 55 yrs
    - Any T
      - Any N
        - M0: I
  - ≥ 55 yrs
    - T1
      - N0/NX
        - M0: I
    - T1
      - N1
        - M0: II
    - T2
      - N0/NX
        - M0: I
    - T2
      - N1
        - M0: II
    - T3a/T3b
      - Any N
        - M0: II
    - T4a
      - Any N
        - M0: III
    - T4b
      - Any N
        - M1: IVB
- **And N is...**
  - Any N
    - M0: I
  - Any N
    - M1: II
- **And M is...**
  - M0: I
  - M1: II
  - M1: IVB
  - M0: III
  - M1: IVB
  - M0: IVB
  - M1: IVC

**Anaplastic thyroid cancer**

- **T is...**
  - T1-T3a
    - N0/NX
      - M0: IVA
  - T1-T3a
    - N1
      - M0: IVB
  - T3b
    - Any N
      - M0: IVB
  - T4
    - Any N
      - M0: IVB
  - Any T
    - Any N
      - M1: IVC
AJCC TNM – Medullary Carcinoma

Primary tumor (pT) for medullary thyroid carcinomas:
- T2*: Advanced disease
- T4a: Moderately advanced disease; tumor of any size with gross extrathyroidal extension into the nearby tissues of the neck, including subcutaneous soft tissue, larynx, trachea, esophagus or recurrent laryngeal nerve
- T4b: Very advanced disease; tumor of any size with extension toward the spine or into nearby large blood vessels, invading the prevertebral fascia or encasing the carotid artery or mediastinal vessels

Regional lymph node (pN):
- N0: No evidence of regional lymph node metastasis
- N1*: Metastasis to regional nodes
  - N1a*: Metastasis to level VI or VII (pretracheal, paratracheal, prelaryngeal / Delphian or upper paratracheal)
- N1b*: Metastasis to bilateral or contralateral lateral neck lymph nodes (levels I, II)

Distant metastasis (M):
- M0: No distant metastasis
- M1: Distant metastasis

Medullary thyroid cancer:

<table>
<thead>
<tr>
<th>Stage</th>
<th>T</th>
<th>N</th>
<th>M</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stage I</td>
<td>T1</td>
<td>N0</td>
<td>M0</td>
</tr>
<tr>
<td>Stage II</td>
<td>T2</td>
<td>N0</td>
<td>M0</td>
</tr>
<tr>
<td>Stage III</td>
<td>T3</td>
<td>N0</td>
<td>M0</td>
</tr>
<tr>
<td>Stage III</td>
<td>T1 - 3</td>
<td>N1a</td>
<td>M0</td>
</tr>
<tr>
<td>Stage IVA</td>
<td>T4a</td>
<td>any N</td>
<td>M0</td>
</tr>
<tr>
<td>Stage IVB</td>
<td>T4b</td>
<td>any N</td>
<td>M0</td>
</tr>
<tr>
<td>Stage IVC</td>
<td>any T</td>
<td>any N</td>
<td>M1</td>
</tr>
</tbody>
</table>

Introduction to SSDI Manual

https://apps.naaccr.org/ssdi/list/
Types of Site Specific Data Items

- Prognostic Factors “Required for Stage Grouping” (All Cases)
  - Not ALL SSDIs Labeled “Required for Stage Grouping” are actually required for staging.
  - Some “Required for Stage Grouping” Items have “Prognostic Significant” and are Required.

- Additional Factors Recommended for Clinical Care (CoC/NCDB and SEER)

- Emerging Factors for Clinical Care (Web Only – Not Required)

- May Include Molecular or Protein Biomarkers, Genetic Markers, Lab Test Value, Interpretation of Lab Value, Clinical Factors such as Size of Lymph Node, Alternate Staging such as FIGO, Measured Depth of Invasion (Breslow Depth), Site Specific Grade Detail (Gleason), Cytogenetics, Immunohistochemistry, Surgical Margin Details, MSI or Microsatellite Instability and More

- You may not see the SSDIs that clinicians reference and think are important today...the reason is that it takes time for cancer registry standards to catch up with present day technology and testing – particularly for genetic factors.

- Your Cancer Program can define any additional SSDIs you would like to capture for your physicians – genetic markers for lung for example - approve these through your Cancer Committee and carefully define user-defined instructions and codes
Types of Site Specific Data Items

- Molecular Genetics still minimally addressed in 2018 SSDIs
- 2018 SSDIs support TNM data - not biomarkers/molecular markers or genetics
- Most evaluate genetic mutations and/or protein surface markers
- Some have targeted therapy(s) associated with mutation

- Chromosomal Abnormality(s) – Mutation
- Biochemical Abnormality
- Genetic/DNA Mutation

- Prognostic
- Diagnostic
- Predictive
- Tumor Burden
- Pharmacodynamics
- Recurrence Monitoring

2018 SSDIs - Thyroid

- Only One SSDI – Schema Discriminator 1
- A schema discriminator is used to discriminate between thyroid gland and thyroglossal duct tumors with primary site code C739: Thyroid Gland.
- Code the site in which the tumor arose.
  - Thyroid gland (see code 1)
    - Subsites include: Thyroid, NOS
  - Thyroglossal duct (see code 2)

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
<th>AJCC Disease ID</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Thyroid gland</td>
<td>73.1: Thyroid: Differentiated</td>
</tr>
<tr>
<td></td>
<td>Thyroid, NOS</td>
<td>73.2: Thyroid: Anaplastic</td>
</tr>
<tr>
<td></td>
<td></td>
<td>74: Thyroid: Medullary</td>
</tr>
<tr>
<td>2</td>
<td>Thyroglossal duct cyst</td>
<td>n/a (not TNM staged)</td>
</tr>
</tbody>
</table>
Treatment Guidelines - Thyroid

- Most types are highly curable cancers – Factors
  - Patient Age
  - Size of Tumor
  - Type of Tumor
  - (Histology, Behavior and Grade)
  - Stage or Extent of Disease – How far has the tumor spread?

- New Tyrosine Kinase Inhibitors (TKI inhibitor) have shown to shrink tumors and slow growth but no cure

- Primary Treatment is remove all tumor at surgery plus nodes to ensure they are free of cancer. Then the patient is treated with radioactive iodine (I-131) to destroy remaining thyroid tissue or for latter stages of disease.

- Hormone Replacement Therapy is always given after thyroidectomy to bring back into balance the hormones normally produced by the thyroid AND to keep the body from producing thyroid stimulating hormones.

- Long-Term Follow Up Plan Must Be Part of Every Treatment Plan

Treatment Guidelines – No Treatment

- Papillary Thyroid Cancers – Active Surveillance
- Some physicians may not even biopsy a small tumor
- Avoids Overtreatment of Cancers Unlikely to Cause Medical Problems...
  - Incidental Nodule(s) Found During W/U for Other Reason
  - Small and Asymptomatic Thyroid Cancers
  - Suspected But Not Proven Thyroid Cancers
- ...however, patients and physicians and family members may insist on treatment even when it is felt to not be necessary and may lead to harmful side effects
- Patients need support they have made a sound treatment decision for their cancer – not a stupid or crazy decision.
Treatment Guidelines - Surgery

- Papillary / Follicular Histology – Local/Regional Disease
  - Lobectomy with Level VI Central Node Dissection
  - Total Thyroidectomy with Level VI Central Node Dissection
  - Radioactive Iodine Therapy – I-131
  - Thyroid Suppression Therapy with Thyroid-Stimulating Hormone (TSH)
  - External Beam Radiation Therapy

- Anaplastic Thyroid Cancer
  - Surgery – Lobectomy or Total Thyroidectomy with Central Node Dissection
  - External Beam Radiation Therapy
  - Systemic Therapy or Target Therapy – Sorafenib, Lenvatinib

- Medullary Thyroid Cancer – Localized Disease
  - Total Thyroidectomy with Nodes plus External Beam Radiation Therapy

- Medullary Thyroid Cancer – Locally Advanced/Metastatic
  - Targeted Therapy – Sorafenib, Lenvatinib, Vandetanib, Cabozantinib
  - Palliative Chemotherapy

Treatment Guidelines – Target Drugs

- Usually only given for metastatic disease or when patient has anaplastic or medullary histology.
- May 25, 2018 - Dabrafenib–Trametinib Combination
  - Anaplastic Histology Only
  - Dabrafenib (Tafinlar) – code as chemo per SEER*Rx
    - Inhibits enzyme over-produced by BRAF V600 gene mutation
  - Trametinib (Mekinist) – code as chemo per SEER*Rx
    - Blocks one of resistance pathways making Tafinlar more effective

<table>
<thead>
<tr>
<th>Table 3: U.S. FDA-approved MTTs for Thyroid Cancers</th>
</tr>
</thead>
<tbody>
<tr>
<td>MTT</td>
</tr>
<tr>
<td>-------</td>
</tr>
<tr>
<td>Vandelatib</td>
</tr>
<tr>
<td>Cabozantinib</td>
</tr>
<tr>
<td>Sorafenib</td>
</tr>
<tr>
<td>Lenvatinib</td>
</tr>
</tbody>
</table>

Note.—FGFR = fibroblast growth factor receptor, PDGFR = platelet-derived growth factor receptor.
NCRA Informational Text Guidelines for Abstracts – Thyroid – Not Available in the Current Set for Text

Follow Typical Recommendations for Complete Documentation with Special Focus on Histology, Capsular Invasion, Extra-thyroid Extension, Nodes and Extranodal Extension, and Mets as well as completely documenting Treatment Given

Be aware there are multiple new histology codes, etc.
Staging Practice - Pending

References

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