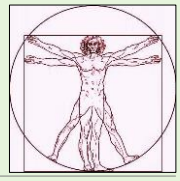
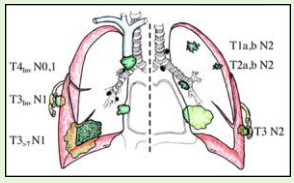
Neoplasms of the LUNG and PLEURA



2015-2016 FCDS Educational Webcast Series

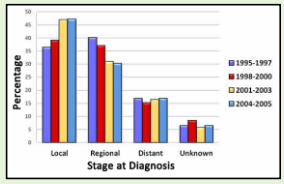
Steven Peace, BS, CTR

September 19, 2015



2015 Focus

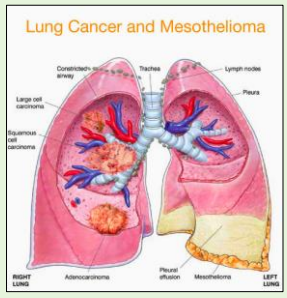
- Anatomy
- SSS 2000
- MPH Rules
- AJCC TNM



1

Presentation Outline

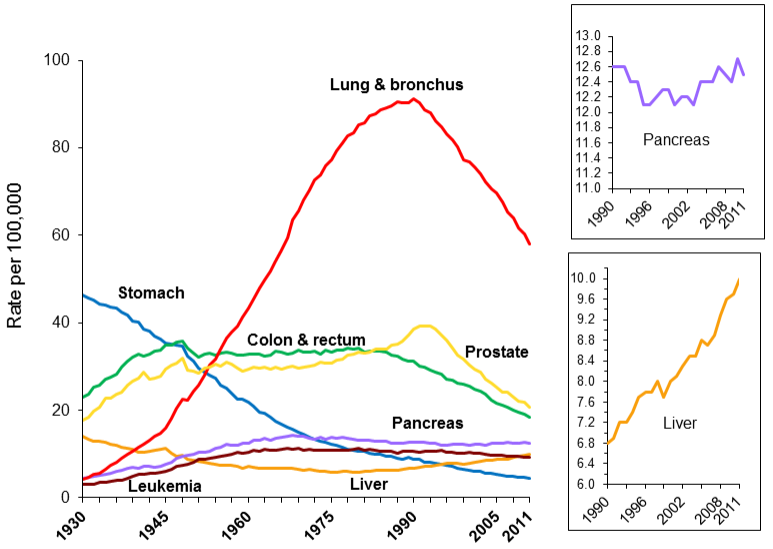
- Overview of Neoplasms of the Lung and Pleura
- Classification of Lung and Pleural Neoplasms
- Anatomy of the Lung and Pleura
- MPH Rules – Quick Review
- Anatomic Staging – SS2000
- Anatomic Staging – AJCC TNM
- Non-Anatomic Site-Specific Factors
- Practice Cases – Live Polling
- Text Documentation



Source: https://www.mesothelioma.com

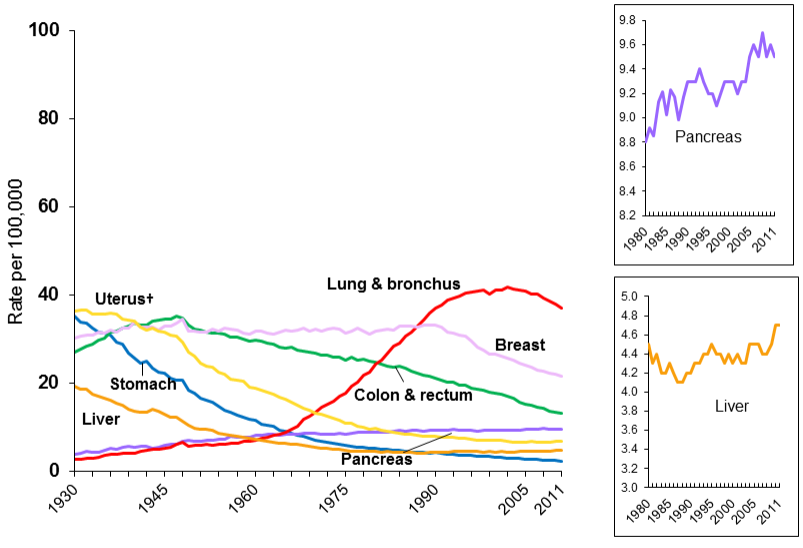
2

Trends in Cancer Death Rates* Among Men, US,1930-2011



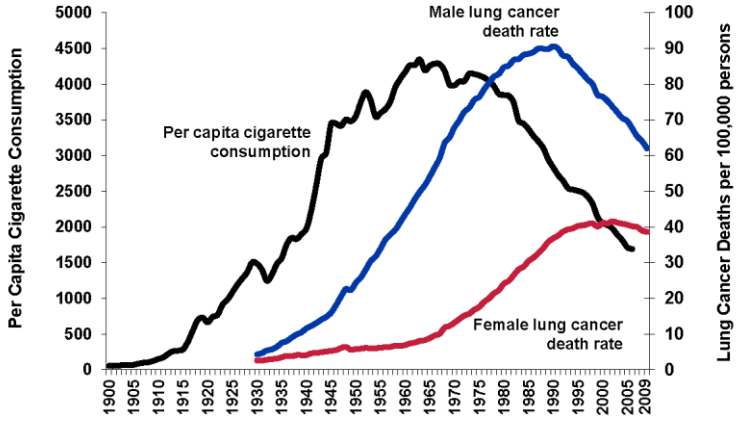
*Age-adjusted to the 2000 US standard population.
Source: National Center for Health Statistics, Centers for Disease Control and Prevention, 2014.

Trends in Cancer Death Rates* Among Women, US,1930-2011



*Age-adjusted to the 2000 US standard population.
†Uterus includes uterine corpus and uterine cervix combined.
Source: National Center for Health Statistics, Centers for Disease Control and Prevention, 2014.

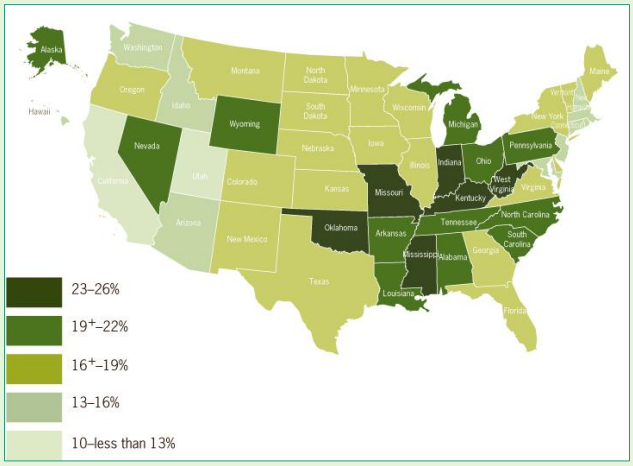
Trends in Tobacco Use and Lung Cancer Death Rates* in the US



*Age-adjusted to 2000 US standard population.

Source: Death rates: US Mortality Data, 1960-2009, US Mortality Volumes, 1930-1959, National Center for Health Statistics, Centers for Disease Control and Prevention. Cigarette consumption: US Department of Agriculture, 1900-2007.

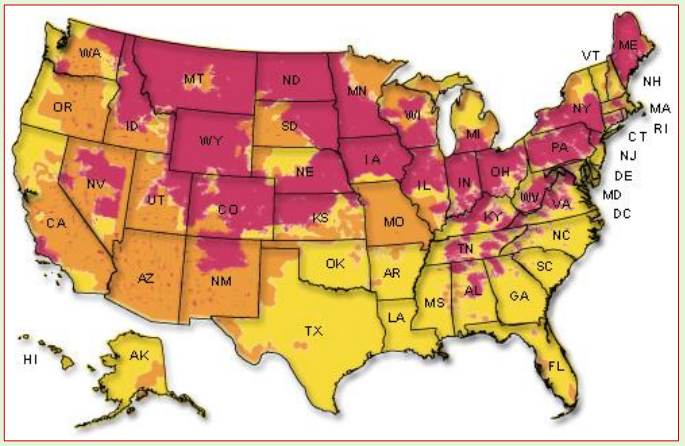
U.S. Adult Smoking Rates



Adult Smoking Rates - http://www.cdc.gov

6

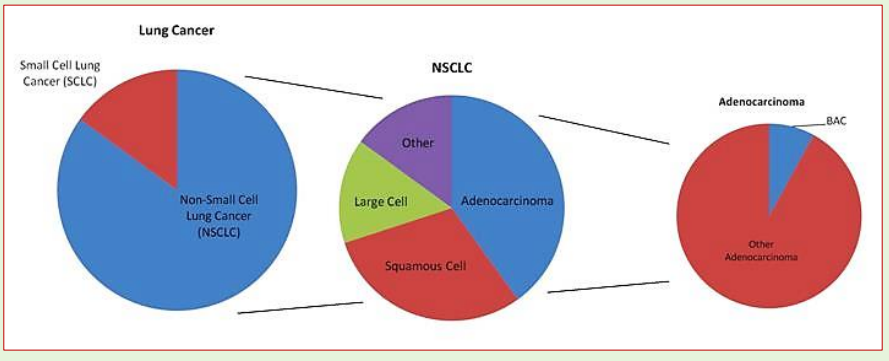
Radon Gas



http://pillartopost.com/epa

Classification of Lung Neoplasms

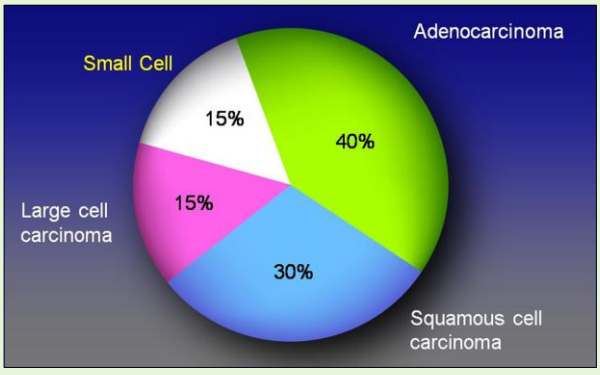
Based on WHO Classification, 3rd ed. (2004)



http://stageiv.files.wordpress.com

Classification of Lung Neoplasms

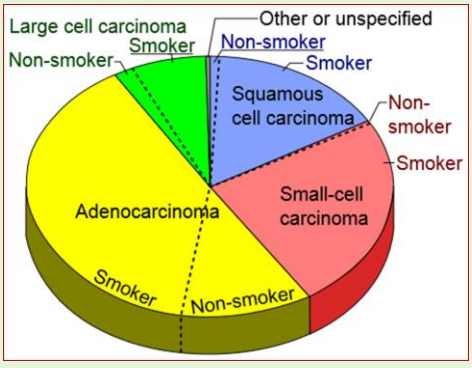
Based on WHO Classification, 3rd ed. (2004)



Wahbah M, et al. Ann Diagn Pathol. 2007; 110:89-96

9

Association with Smoking

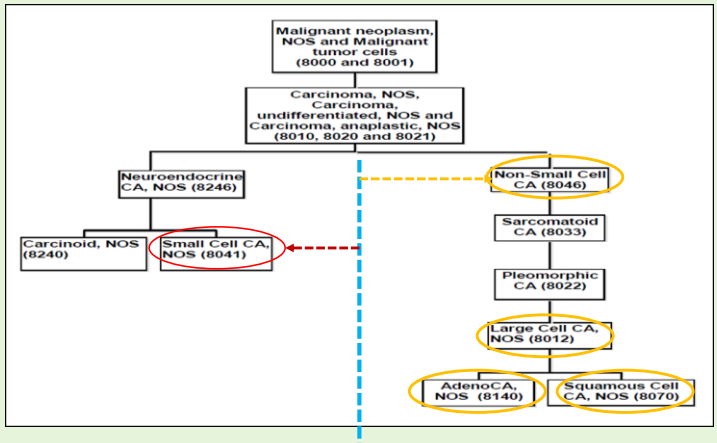


Kenfield SA, Wei EK, Stampfer MJ, Rosner BA, Colditz GA (2008), Tobacco Control 17 (3): 198-204

10

Classification of Lung Neoplasms

Based on WHO Classification, 3rd ed. (2004)



11

Characteristics of SCC of Lung

- Most **arise centrally** within main, lobar, segmental or subsegmental bronchi
- Peripheral tumors **often undergo central necrosis** with cavitation
- Tumor **growth usually intrabronchial and peribronchial**
- **Slow growing neoplasm** with late metastatic spread
- **Better prognosis than adenocarcinoma**

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Characteristics of AdenoCA of Lung

- More **easily detected on imaging**
- **More common in women and non-smokers**
- Most **arise in the smaller peripheral airways**
- Central tumors **more likely to have lymph node metastasis**
- More often **associated with pleural effusion(s) and distant metastasis**

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Characteristics of Small Cell CA of Lung

- 15-25% of all lung cancers and **almost exclusive to smokers**
- TNM Staging can be used but **VALG Staging System** is often used by MDs
  - ✓ **Limited disease** – confined to 1 side of thorax - includes contralateral LN
  - ✓ **Extensive disease** – beyond boundaries noted above
- **Most arise centrally with large hilar mass and bulky mediastinal nodes**
- Characteristic **rapid growth with early development of diffuse metastasis**
  - Metastasis include brain and CNS, bone marrow, liver, adrenal(s) and bone
- **Associated with paraneoplastic syndrome and poor survival**

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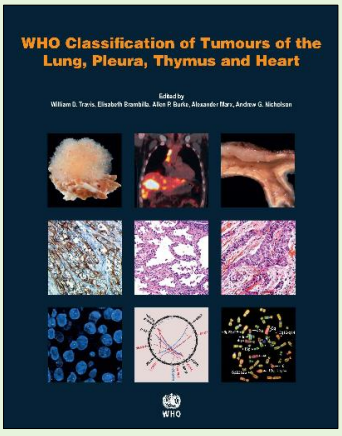
Characteristics of Non-Small Cell CA

- "**Non-Small Cell Lung Cancer is a generic term (aka; NSCLC)**.
- **Use of the term NSCLC should be avoided as a single diagnostic term**.
- The following terms are acceptable when IHC can distinguish the two:
  - **"NSCLC favor adenocarcinoma" or**
  - **"NSCLC favor squamous cell carcinoma"**
- EGFR testing is strongly recommended in all "NSCLC favor adenocarcinoma"

15

2015 WHO Classification, 4th ed.



Highlights

- Multi-Disciplinary Correlation
- Invasive Neoplasm classified according to predominant subtype
- Stop Using the Term "BAC" and "bronchio-alveolar carcinoma"
- Replace BAC with 5 new adenocarcinoma subtypes
  - Add "in situ" classification
  - Add "minimally invasive"
- Add genetic test/markers
  - EGFR, Alk, KRAS, TTF-1, p40
- SCC with minor changes

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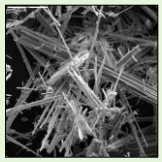
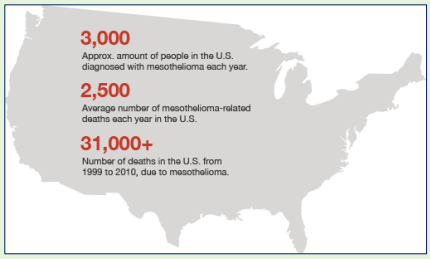
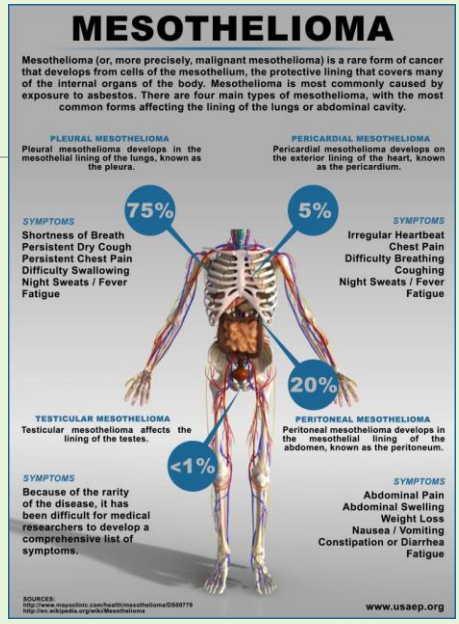
Mesothelioma

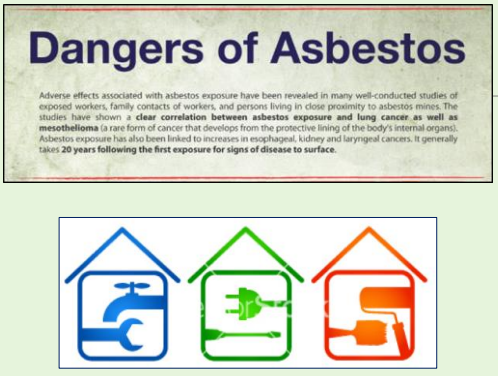


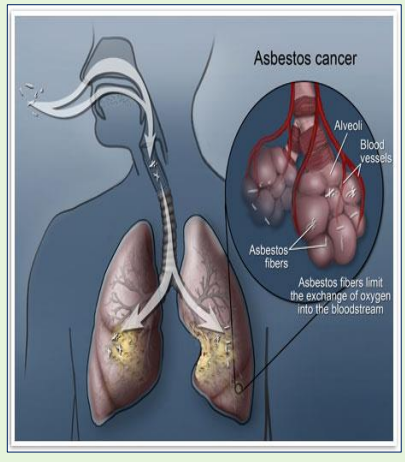




Sources: http://www.mesothelioma.com and http://www.usaep.org

Asbestos





http://www.mesothelioma.com/asbestos-cancer

Lung Anatomy

http://www.omnimedicalsearch.com/conditions-diseases/images/lung-cancer.jpg

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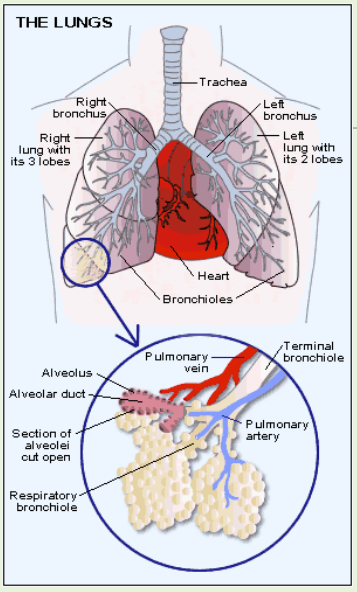
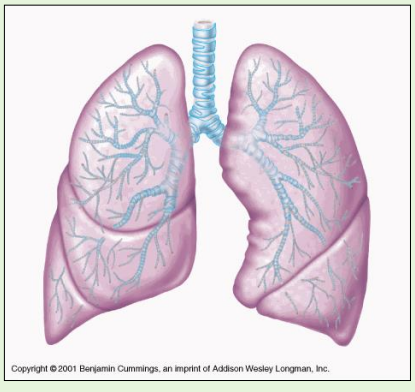
Lung Anatomy





http://www.damav.com/mare/lung/ http://legacy.owensboro.kctcs.edu

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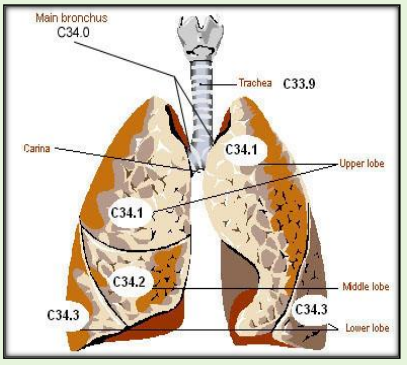
Lung Anatomy

C34.0 Main bronchus
C34.1 Upper lobe, lung
C34.2 Middle lobe, lung
(right lung only)
C34.3 Lower lobe, lung
C34.8 Overlapping lesion
C34.9 Lung, NOS



Source: SEER Training: ICD-O-3 Site Codes

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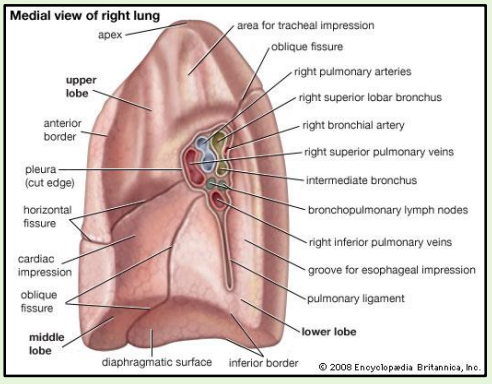
Lung Anatomy

The hilum is the space in each lung where the bronchus and blood vessels enter the lung.

The apex is the rounded area at the top of each lung.

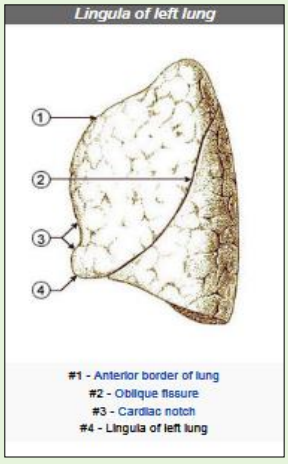


Source: 2008 Encyclopedia Britannica, Inc. on-line

22

Lung Anatomy

The lingula, found only in the left lung, is a projection of the upper lobe of the left lung thought to be a remnant of an ancient middle lobe of the left lung.



Source: SEER Training: ICD-O-3 Site Codes

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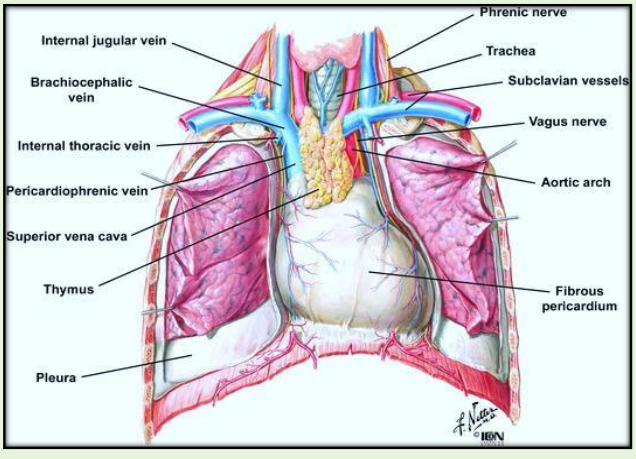
Lung Anatomy



Source: Springer Images. Figure adapted from Atlas of Human Anatomy, 2nd ed. Contents of the superior and middle mediastinum. http://www.springerimages.com/Images/MedicineAndPublicHealth/1-10.1007_978-1-60327-372-5_4-9

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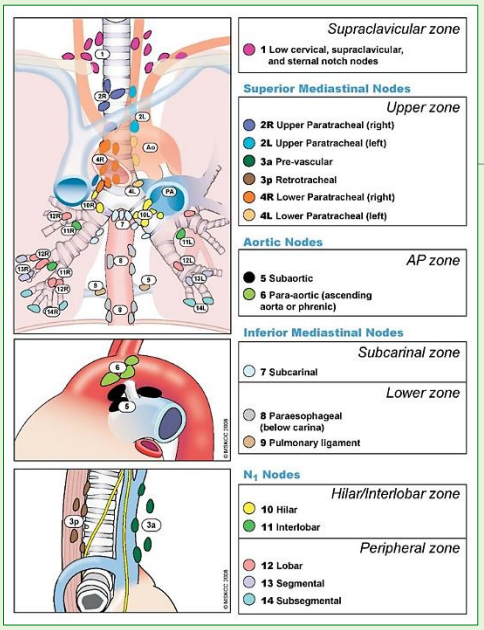

Lung Anatomy

NX Regional lymph nodes cannot be assessed

N0 No regional lymph node metastases

N1 Metastasis in ipsilateral peribronchial and/or ipsilateral hilar lymph nodes and intrapulmonary nodes, including involvement by direct extension

N2 Metastasis in ipsilateral mediastinal and/or subcarinal lymph node(s)

N3 Metastasis in contralateral mediastinal, contralateral hilar, ipsilateral or contralateral scalene, or supraclavicular lymph node(s)

IASLC lymph node map from Memorial Sloan-Kettering Cancer Center, 2009

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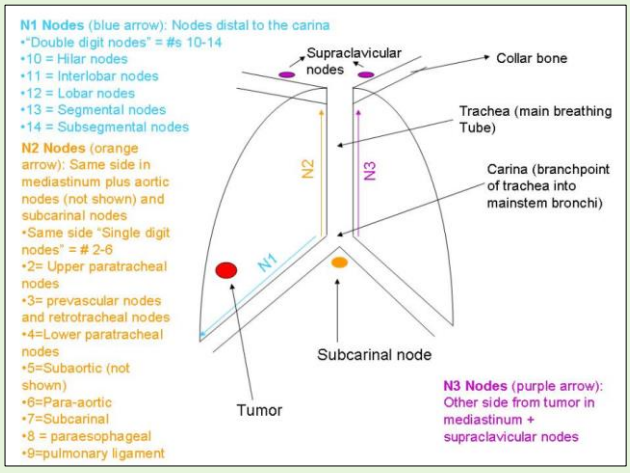
Lung Anatomy



Source: http://cancergrace.org/lung/files/2010/04/simplified-staging.jpg

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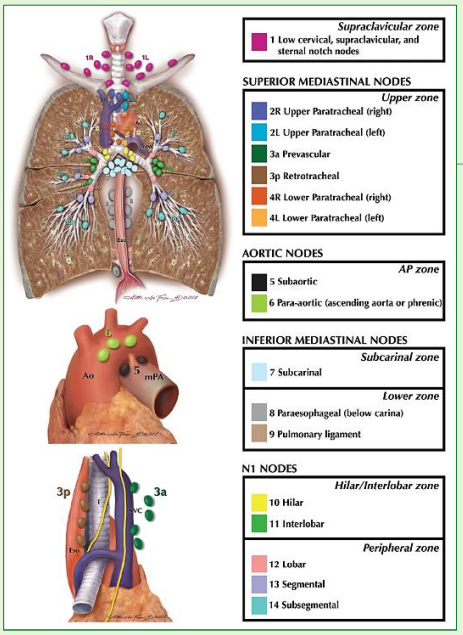

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IASLC lymph node map - WHO Classification of Tumours of the Lung, 2015

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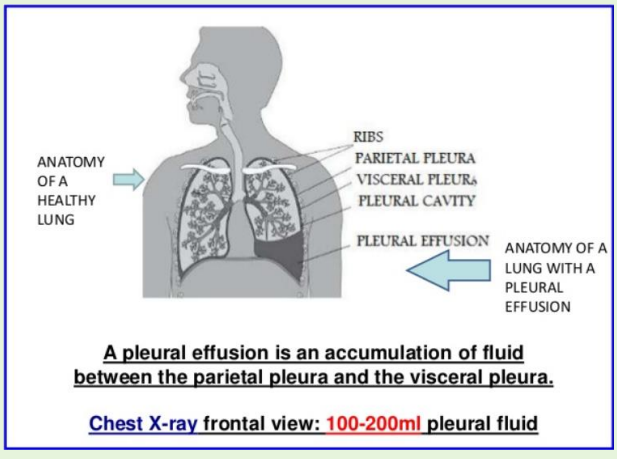
Lung Anatomy



Source: www.slideshare.net/pleuraleffusion/drmahesh

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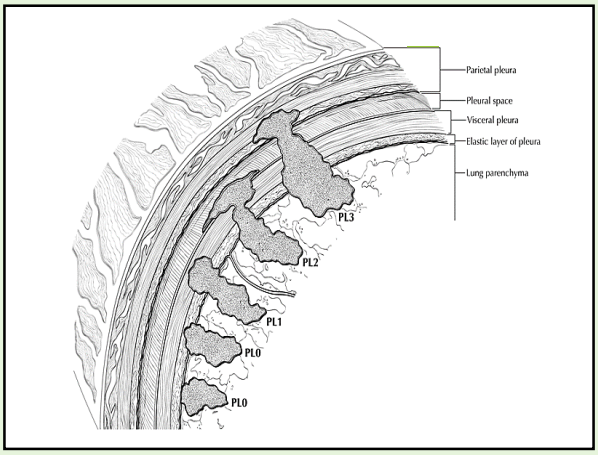
Lung Anatomy



Collaborative Stage Data Collection System, Part I Section II – Lab Tests, Tumor Markers, SSFs

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Genetic Abnormalities

Table 1.07 Major genetic changes in lung cancer

Alterations	Small cell carcinoma (%)	Adenocarcinoma (%)	Squamous cell carcinoma (%)
Mutation			
BRAF	0	< 5	0
EGFR Caucasian	< 1	10–20	< 1
Asian	< 5	35–45	< 5
ERBB2/HER2	0	< 5	0
KRAS Caucasian	< 1	15–35	< 5
Asian	< 1	5–10	< 5
PIK3CA	< 5	< 5	5–15
RB	> 90	5–15	5–15
TP53	> 90	30–40	50–80
Amplification			
EGFR	< 1	5–10	10
ERBB2/HER2	< 1	< 5	< 1
MET	< 1	< 5	< 5
MYC	20–30	5–10	5–10
FGFR1	< 1	< 5	15–25
Gene rearrangement			
ALK	0	5	< 1
RET	0	1–2	0
ROS1	0	1–2	0
NTRK1	0	< 1	0
NRG1	0	< 1	0

EMERGING TARGETED AGENTS FOR PATIENTS WITH GENETIC ALTERATIONS

Genetic Alteration (ie, Driver event)	Available Targeted Agents with Activity Against Driver Event in Lung Cancer
BRAF* V600E mutation	vemurafenib[1] dabrafenib[2]
***MET* amplification**	crizotinib[3,4]
***ROS1* rearrangements**	crizotinib[5]
***HER2* mutations**	trastuzumab[6] (category 2B) afatinib[7] (category 2B)
***RET* rearrangements**	cabozantinib[8] (category 2B)

*Non-V600E mutations have variable kinase activity and response to these agents.

Source: WHO Classification of Tumours of the Lung, Pleura, Thymus and Heart, 2015 and NCCN Guidelines NSCLCv7.2015

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Biomarkers and Genetic Abnormalities

CK7 and CK20 – CKs are cytokeratins, markers common to carcinomas. Sometimes when the site of origin of an adenocarcinoma isn't certain, CK7 and CK20 can help. The typical lung pattern is CK7+ and CK20-.

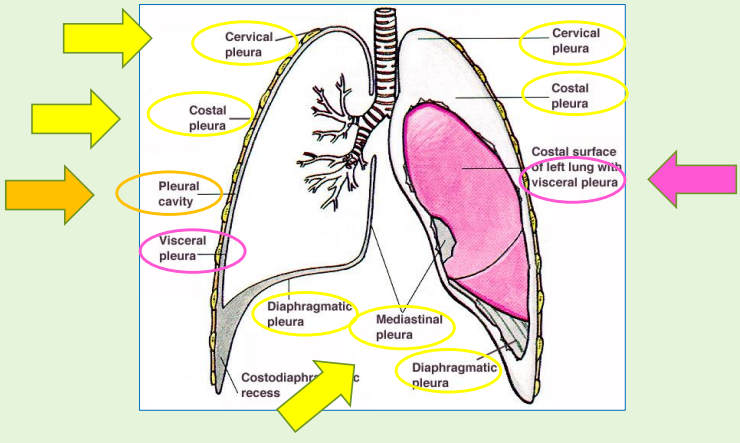
TTF-1 (thyroid transcription factor-1) – 85% of adenocarcinoma of the lung and a minority of squamous cell cancers will bear this marker. Most non-lung cancers do not express TTF-1 (prominent exceptions include thyroid cancer and cancers of the female genitourinary tract).

p63 – A marker for squamous cell carcinoma

Genetic Abnormality: EGFR mutation, K-RAS mutation, ALK rearrangement, EML4/ALK translocation. *(Note: genetic testing for these mutations can help predict efficacy of agents; erlotinib (tarceva), gefitinib (iressa) and crizotinib)*

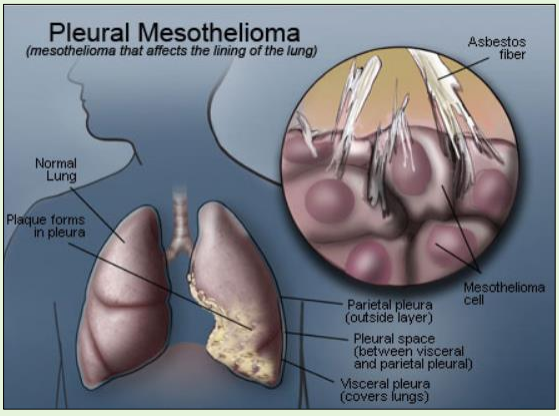
Source: NCCN and GRACE (Global Resource for Advancing Cancer Education) 31

Pleura Anatomy



Source: http://www.depure.org/learning-further-about-anatomy-of-lung/basic-anatomy-of-lung/ 32

Pleural Mesothelioma



Source: http://www.mesothelioma.com/images/pleural-mesothelioma.jpg

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Lung MPH Rules Terms and Definitions

Based on WHO Classification, 3rd ed. (2004)

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Lung Equivalent Terms, Definitions, Charts, Tables and Illustrations
C340-C349
(Excludes lymphoma and leukemia M9590-9989 and Kaposi sarcoma M9140)

Introduction
Use these rules only for cases with primary lung cancer.

Lung carcinomas may be broadly grouped into two categories, small cell and non-small cell carcinoma.
Frequently a patient may have two or more tumors in one lung and may have one or more tumors in the contralateral lung. The physician may biopsy only one of the tumors. Code the case as a single primary (See Rule M1, Note 2) unless one of the tumors is proven to be a different histology. It is irrelevant whether the other tumors are identified as cancer, primary tumors, or metastases.

Equivalent or Equal Terms
- Low grade neuroendocrine carcinoma, carcinoid
- Tumor, mass, lesion, neoplasm (for multiple primary and histology coding rules only)
- Type, subtype, predominantly, with features of, major, or with ___differentiation

Obsolete Terms for Small Cell Carcinoma (Terms that are no longer recognized)
- Intermediate cell carcinoma (8044)
- Mixed small cell/large cell carcinoma (8045) (Code is still used; however current accepted terminology is combined small cell carcinoma)
- Oat cell carcinoma (8042)
- Small cell anaplastic carcinoma (No ICD-O-3 code)
- Undifferentiated small cell carcinoma (No ICD-O-3 code)

Definitions

Adenocarcinoma with mixed subtypes (8255): A mixture of two or more of the subtypes of adenocarcinoma such as acinar, papillary, bronchoalveolar, or solid with mucin formation.

Adenosquamous carcinoma (8560): A single histology in a single tumor composed of both squamous cell carcinoma and adenocarcinoma.

Bilateral lung cancer: This phrase simply means that there is at least one malignancy in the right lung and at least one malignancy in the left lung. Do not base multiple primary decision on this phrase; bilateral does not mean this is a single primary. Use the multiple primary rules to decide whether to code bilateral lung cancers as a single or multiple primary.

Combined small cell carcinoma (8045): A small cell carcinoma that is combined with a non-small cell carcinoma. The combinations are small cell and adenocarcinoma, or squamous cell carcinoma, or large cell carcinoma.

Lung Terms and Definitions

Lung Equivalent Terms, Definitions, Charts, Tables and Illustrations
C340-C349
(Excludes lymphoma and leukemia M9590-9989 and Kaposi sarcoma M9140)

Large cell carcinoma (8012): Large cell is a diagnosis that is used when the tumor is a non-small cell carcinoma that is undifferentiated. Because the tumor is undifferentiated, the pathologist cannot find glandular (adeno), or squamous differentiation.

Large cell neuroendocrine carcinoma (8013): A non-small cell carcinoma with neuroendocrine differentiation proven by immunohistochemical stain, currently classified as large cell carcinoma. These tumors require further study before being included as a separate category in a histologic classification.

Most invasive: The tumor with the greatest continuous extension.

Neuroendocrine carcinoma (8246): Neuroendocrine carcinoma is a group of carcinomas that include typical carcinoid tumor and small cell carcinoma. Code the specific histology when given. Code neuroendocrine carcinoma, NOS (8246) when no specific histology is documented.

Non-small cell carcinoma (8046): The term non-small cell is used two ways, as a group term describing all carcinomas that are not small cell; and as a default diagnosis when there isn't enough tissue to classify the tumor beyond the exclusion of small cell.

Pancoast tumor: An anatomic designation (not a specific histology) for a lung cancer that starts in the upper lobe of the lung and extends outward to destroy the ribs and vertebrae. The tumor may compress or directly invade the brachial plexus (nerve bundles) of the neck, causing pain. Pancoast tumor may also be called **superior sulcus tumor**.

Pleomorphic carcinoma (8022): A poorly differentiated non-small cell carcinoma (squamous cell carcinoma, adenocarcinoma, or large cell carcinoma) containing spindle cells and/or giant cells or, a carcinoma containing only spindle cells and giant cells. These fall under the general category of **sarcomatoid carcinoma**.

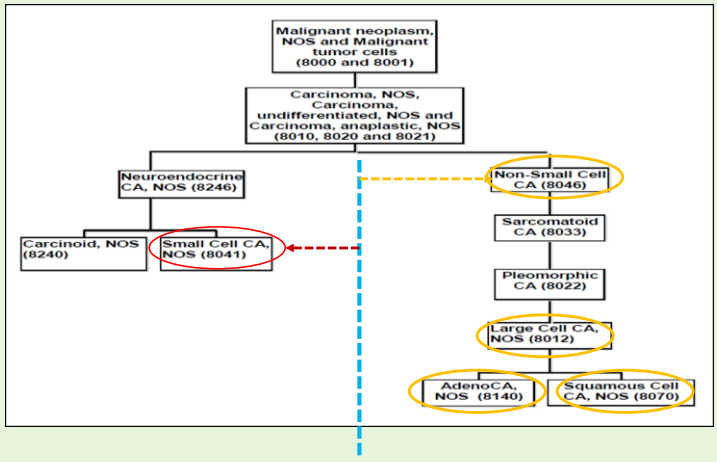
Sarcomatoid carcinoma: A group of tumors that are non-small cell in type and contain spindle cells and/or giant cells. Depending on the histologic features the tumor may be designated: pleomorphic carcinoma (8022); spindle cell carcinoma (8032); giant cell carcinoma (8031), carcinosarcoma (8980); or pulmonary blastoma (8972)

Small cell carcinoma: Malignant epithelial tumor consisting of small cells. There are many types of lung cancer, but most can be categorized into one of two basic types, "small cell carcinoma" or "non-small cell carcinoma"

Undifferentiated carcinoma (8020): A high grade malignancy lacking glandular structures or other specific features that can be used to better classify the tumor. Undifferentiated carcinoma is used by pathologists when they believe the tumor is a carcinoma (not lymphoma, melanoma, or sarcoma) but they are not sure if the tumor is small cell or non-small cell.

Classification of Lung Neoplasms

Based on WHO Classification, 3rd ed. (2004)



37

Lung MPH Rules
Multiple Primary Rules

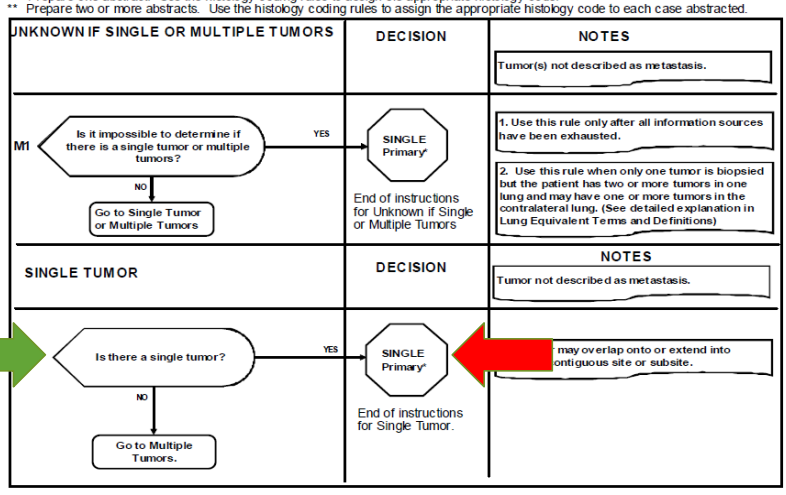
Based on WHO Classification, 3rd ed. (2004)

38

Lung Multiple Primary Rules - Flowchart

(C340 - C349)
(Excludes lymphoma and leukemia M9590-9989 and Kaposi sarcoma M9140)

* Prepare one abstract. Use the histology coding rules to assign the appropriate histology code.
** Prepare two or more abstracts. Use the histology coding rules to assign the appropriate histology code to each case abstracted.



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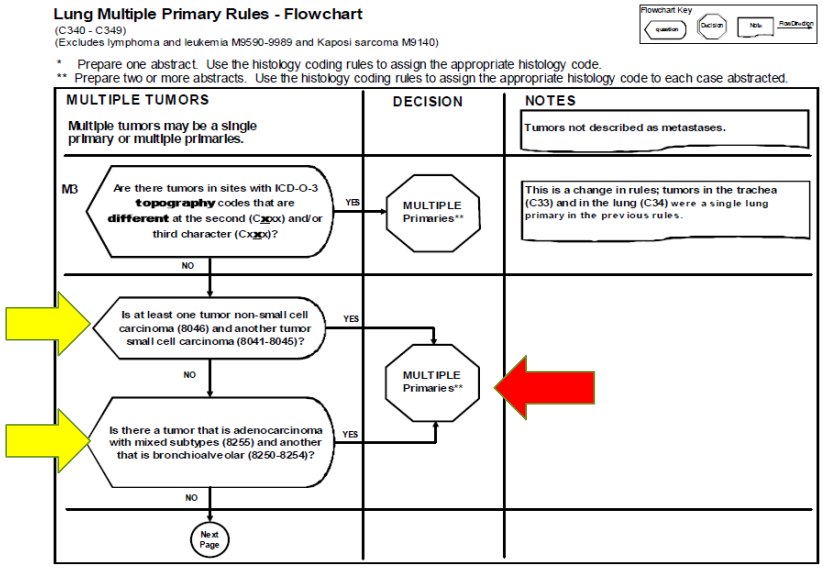

Lung Multiple Primary Rules - Flowchart

(C340 - C349)
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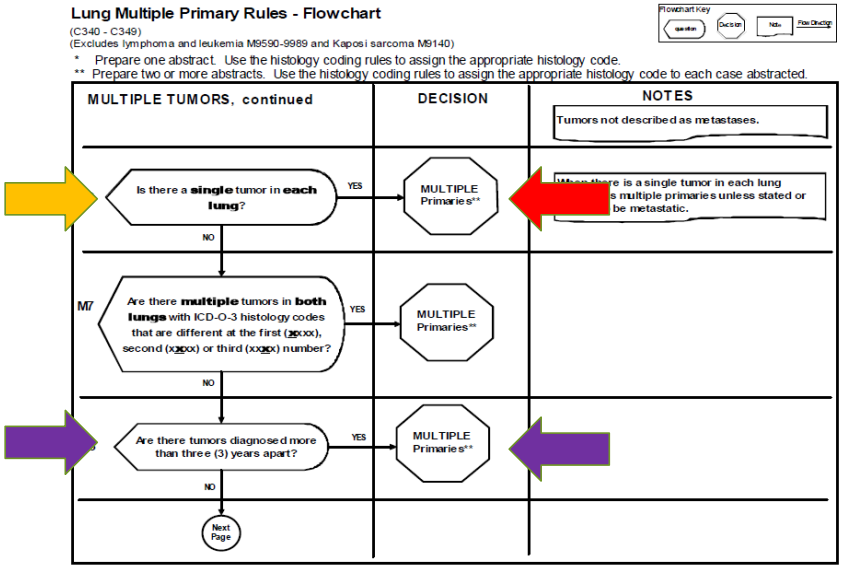

Lung Multiple Primary Rules - Flowchart

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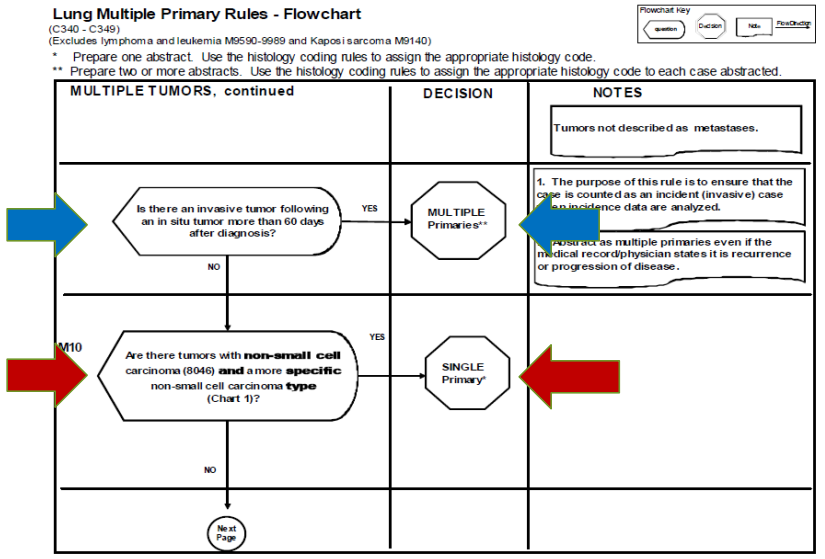
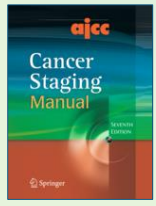


Quick Review: How to Use the AJCC Cancer Staging Manual, 7th ed.

FCDS ANNUAL CONFERENCE
ST PETERSBURG, FLORIDA
JULY 30, 2015

Steven Peace, CTR



Purchase and Ordering Information

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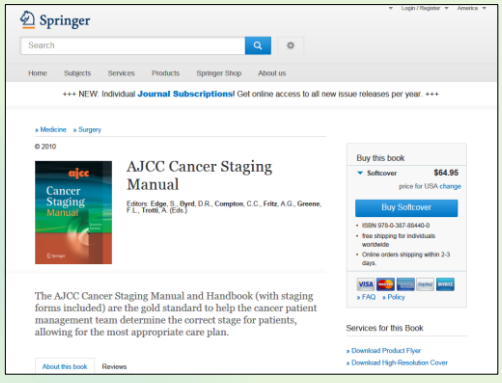

- AJCC Cancer Staging Manual – 7th edition, 2010
- COST: $64.95
- ISBN: 978-0-387-88440-0

- Required - Florida Mandate
  - FCDS will not purchase
  - Facility may purchase
  - Individual may purchase

- Also Required to Purchase 8th Edition in 2016-2017

- https://cancerstaging.org
- http://springer.com
- 1-800-SPRINGER

http://www.springer.com/us/book/9780387884400

Chapter Outline and Contents

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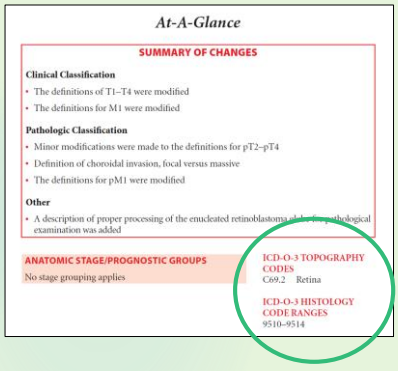
Staging at a Glance	Summary of anatomic stage/prognostic grouping
Changes in Staging	Table summarizing changes in staging from the 6th edition
Introduction	Overview of factors affecting staging and outcome
Anatomic Considerations	o Primary Tumor o Regional lymph nodes o Metastatic sites
Rules for Classification	o Clinical o Pathologic
Prognostic Features	Identification and discussion of non-anatomic prognostic factors
Definitions of TNM	T: Primary tumor N: Regional lymph nodes M: Distant metastasis
Anatomic Stage Prognostic Groups	
Prognostic Factors (SSFs)	a. Required for staging b. Clinically significant
Grade	
Histopathologic Type	
Bibliography	
Staging Form	

AJCC Cancer Staging Manual, 7th ed. – Chapter 1, Table 1.10, p.14

Identifying Neoplasms by Chapter

46



At-A-Glance

SUMMARY OF CHANGES

Clinical Classification

- The definitions of T1–T4 were modified
- The definitions for M1 were modified

Pathologic Classification

- Minor modifications were made to the definitions for pT2–pT4
- Definition of choroidal invasion, focal versus massive
- The definitions for pM1 were modified

Other

- A description of proper processing of the enucleated retinoblastoma [illegible] pathological examination was added

ANATOMIC STAGE/PROGNOSTIC GROUPS

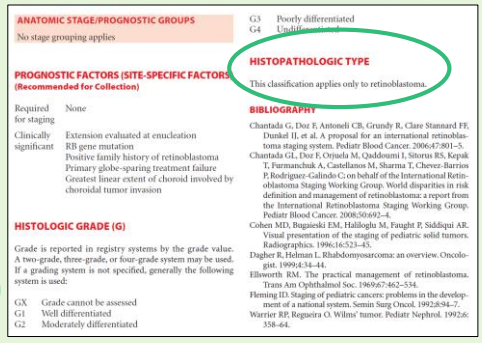
No stage grouping applies

ICD-O-3 TOPOGRAPHY CODES

C69.2 Retina

ICD-O-3 HISTOLOGY CODE RANGES

9510–9514



ANATOMIC STAGE/PROGNOSTIC GROUPS

No stage grouping applies

PROGNOSTIC FACTORS (SITE-SPECIFIC FACTORS)
(Recommended for Collection)

Required for staging	None
Clinically significant	Extension evaluated at enucleation RB gene mutation Positive family history of retinoblastoma Primary globe-sparing treatment failure Greatest linear extent of choroid involved by choroidal tumor invasion

HISTOLOGIC GRADE (G)

Grade is reported in registry systems by the grade value. A two-grade, three-grade, or four-grade system may be used. If a grading system is not specified, generally the following system is used:

GX	Grade cannot be assessed
G1	Well differentiated
G2	Moderately differentiated
G3	Poorly differentiated
G4	Undifferentiated

HISTOPATHOLOGIC TYPE

This classification applies only to retinoblastoma.

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Ellsworth RM. The practical management of retinoblastoma. Trans Am Ophthalmol Soc. 1969;67:462–534.

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Warrier RP, Reguera O. Wilms' tumor. Pediatr Nephrol. 1992;6:358–64.

AJCC Cancer Staging Manual, 7th ed. – Chapter 52, Retinoblastoma, p.561 and p.564

Neoplasms Not in the AJCC Manual

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- **Not all types of cancer are AJCC-stage able.**
- **Use the Primary Site Codes** listed at the beginning of each chapter in the AJCC Cancer Staging Manual. **Use the List of Histopathologic Types** in each chapter are toward the end of each chapter and are used **as a guide** to indicate the cancer types which can be AJCC-staged using that staging scheme.
- *Histologic Types listed as inclusions (or not listed – because they are exclusions) for each individual chapter should NOT be AJCC-staged using that chapter.*
- **Note:** Some chapters are specifically limited to certain cancer types only with a certain anatomic site (such as skin melanomas). Some chapters are specifically limited to certain histologic types 9590-9729 regardless of primary site. This site and/or histology limitation does not limit coding for the primary site here.

Read the Chapter Introduction, Anatomy and Rules Before You Start

48

- These 3 sections, Chapter Introduction, Anatomy and Rules, are often overlooked or skimmed. This is where most of your questions will be answered…not in the coding section.
- The Rules for Classification instruction you as to which diagnostic and staging tests, imaging, biopsy, sentinel or resected nodes, etc. can and should be used when assigning clinical or pathologic TNM.
- Sometimes the Cancer Staging Form and/or the AJCC Chapter includes anatomic drawings to help clarify local/regional anatomy.
- Always review the Prognostic Features as this will help you identify which laboratory tests, symptoms, or other factors are important for staging

Refer Directly to the AJCC Manual

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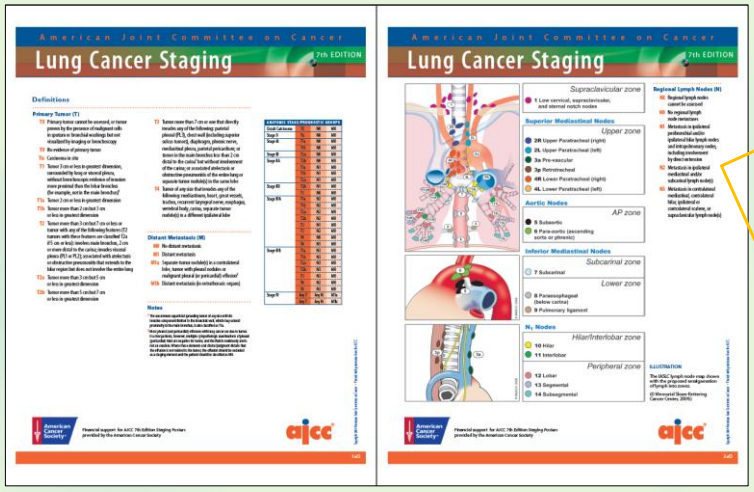
- Once you have sized up the case and read through all of the imaging studies, diagnostic biopsy (any type), resection including lymph nodes if done, neo-adjuvant therapy status, operative report, consultations, etc. Be sure to include and annotate in text the physician stage if provided as a component of your assessment.
- PLEASE - DO NOT JUST USE THE PHYSICIAN STAGE WITHOUT ASSESSING THE CORRECTNESS. You may have additional information in the medical record that was not available or not included in the physician or pathologist assignment of TNM or AJCC Stage Group.
- Use the Definitions of TNM Section in the Chapter to assign the most appropriate T, N, and M values using the AJCC Staging Manual Instructions and Rules for assignment. Remember the Down-Staging Rule when assigning stage when for some cancers or cases you may not be able to assign a value as precisely as you did in CS Ext or CS LN.
- Make sure you include any prognostic factors (anatomic or non-anatomic) that are required for determining the correct Anatomic Stage/Prognostic Groups – most are SSFs may also include; age, histologic type, grade, as well as other non-anatomic SSFs.
- Assign the appropriate Anatomic Stage/Prognostic Group using T, N, M and SSFs.

Free AJCC TNM Staging Poster

50



SAMPLE ONLY

AJCC TNM Staging Form

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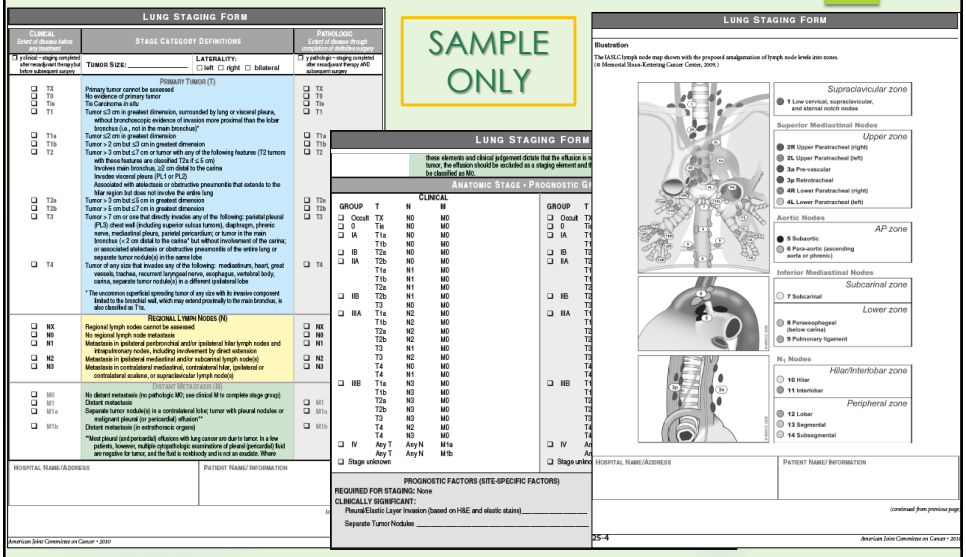
SAMPLE ONLY



Other Helpful Information

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TNM Help

- AJCC 6th ed. & 7th ed. Help
- Introduction Help
- Abbreviated Chapter
- Explanatory Notes
- Common Questions
- FREE !

http://cancerstaging.blogspot.com/2005/02/explanatory-notes-specific-anatomical.html

Cancer Staging Basics

1. Where did the cancer start (primary site)?
2. Where did the cancer go (how far did it spread)?
3. How did the cancer get to the other organ or structure?
4. What is the SS2000 and AJCC TNM for this cancer?

➢ Incorporate SSFs Required for Staging but not for SS2000

53

Cancer Staging Basics

There are three components to AJCC Cancer Stage and to assign Summary Stage:

- Where and how big the original mass or primary tumor is = T
- Which nodes the cancer has spread to including how many positive = N
- Whether the cancer has spread to 1 or more distant site(s) = M

The T, N, and M information is joined to assign a Summary Stage and an AJCC "Stage Group" (now called **Anatomic Stage/Prognostic Group** with addition of genetic and bio-molecular tumor markers and other prognostic factors in the AJCC 7th edition)

All cancers must be assigned a Summary Stage – SS2000

All cancers are assigned clinical stage – verify histology inclusion for TNM Chapter

Surgically resected cancers can be assigned pathologic stage – verify inclusion list

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Criteria Used to Stage Lung Cancer

What To Look For & Document When Reviewing Lung Cancer Cases

Physical Exam – paraneoplastic syndrome, nerve or vessel obstruction

CT Chest – tumor location, tumor size, nodes, pleural effusion

CT Abdomen – liver or adrenal mets

CT/MRI Brain – brain mets

Pathology Report(s) – Resection of Primary and Nodal Status

Pathology Report(s) – Extension to/thru visceral pleura

Pathology Report(s) – Extension to parietal pleura

Cytology Report(s) – Pleural Fluid (blood/exudate)

Genetic Abnormalities – EGFR, KRAS, BRAF, ALK

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SS2000

BRONCHUS AND LUNG
C34.0-C34.3, C34.8-C34.9
C34.0 Main bronchus (including carina, hilus of lung) <>
C34.1 Upper lobe (including lingula), lung <>
C34.2 Middle, lung <>
C34.3 Lower lobe, lung <>
C34.8 Overlapping lesion of lung <>
C34.9 Lung, NOS <>
<> Laterality must be coded for this site (except carina and hilus of lung).

SUMMARY STAGE

0 In situ: Noninvasive; intraepithelial

1 Localized

Confined to carina
Confined to hilus of lung
Confined to the main stem bronchus ≥2.0 cm from carina
Confined to the main stem bronchus, NOS
Extension from other parts of the lung to main stem bronchus ≥2.0 cm from carina[#]
Extension from other parts of the lung to main stem bronchus, NOS[##]
Single tumor confined to one lung

Localized, NOS

7 Distant site(s)/node(s) involved

Distant lymph node(s):
Cervical, NOS
Contralateral/bilateral hilar (bronchopulmonary) (proximal lobar) (pulmonary root)
Contralateral/bilateral mediastinal
Scalene (inferior deep cervical), ipsilateral or contralateral
Supraclavicular (transverse cervical), ipsilateral or contralateral
Other distant lymph node(s)

Extension to:
Abdominal organs
Adjacent rib[##]
Contralateral lung
Contralateral main stem bronchus
Heart[##]
Pericardial effusion (malignant or NOS)
Pleural effusion (malignant or NOS)
Skeletal muscle
Skin of chest
Sternum
Vertebra(e)
Visceral pericardium[##]

Further contiguous extension

Separate tumor nodule(s) in different lobe[#*]

Separate tumor nodule(s) in contralateral lung

Metastasis

56

Steps to Assign SS2000

Three summary stage groups can be ruled out quickly: in situ, distant, and localized

FIRST

In situ

1. Rule out in situ stage disease. Carcinomas and melanomas are the only types of cancer that can be classified as in situ. Only carcinomas have a basement membrane. Sarcomas are never described as in situ. A pathologist must examine the primary organ and state that the tumor is in situ. If the cancer is anything except a carcinoma or melanoma, it cannot be in situ.
2. If there is any evidence of invasion (or extension to), nodal involvement or metastatic spread, the case is not in situ even if the pathology report so states. This is a common error in staging cervical cancer where the path report states that the cancer is "in situ with microinvasion"—such a case would be staged as localized.

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Steps to Assign SS2000

Three summary stage groups can be ruled out quickly: in situ, distant, and localized

SECOND

Distant

3. Rule out distant disease. If metastases can be documented, there is no need to spend a great deal of time identifying local or regional spread. If distant metastases are recorded on x-ray or needle biopsy, the stage is already determined and the patient does not need to undergo a lot of other tests.
4. Hematopoietic diseases, such as leukemia and multiple myeloma, are considered disseminated or distant at time of diagnosis.
5. Rule out distant spread by reading the operative report for comments about seeding, implants, liver nodules, or other indications of metastases. Read diagnostic reports for references to distant disease.
6. If nodes, organs, or adjacent tissues are not specifically mentioned in the description of the various categories, attempt to cross-reference the term you have with those outlined. If there is no match, assume the site in question represents distant disease.

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Steps to Assign SS2000

Three summary stage groups can be ruled out quickly: in situ, distant, and localized

THIRD

Localized

7. Rule out that the cancer is "confined to the organ of origin." In order for a lesion to be classified as localized, it must not extend beyond the outer limits of the organ and there must be no evidence of metastases anywhere else.
8. Terms such as "blood vessel invasion" or "perineural lymphatic invasion" do not necessarily indicate that the cancer has spread beyond the primary organ. If tumor at the primary site has invaded lymph or blood vessels, there is the potential for malignant cells to be transported throughout the body. Step 1 (invasion), has occurred, but not necessarily steps 2 (transport of cancer cells) and 3 (growth at the secondary site). The case may still be localized.
9. Vascular invasion within the primary is not a determining factor in changing the stage unless there is definite evidence of tumor at distant sites.

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SS2000

BRONCHUS AND LUNG
C34.0-C34.3, C34.8-C34.9
C34.0 Main bronchus (including carina, hilus of lung) <>
C34.1 Upper lobe (including lingula), lung <>
C34.2 Middle, lung <>
C34.3 Lower lobe, lung <>
C34.8 Overlapping lesion of lung <>
C34.9 Lung, NOS <>
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Extension from other parts of the lung to main stem bronchus, NOS[##]
Single tumor confined to one lung

Localized, NOS

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Distant lymph node(s):
Cervical, NOS
Contralateral/bilateral hilar (bronchopulmonary) (proximal lobar) (pulmonary root)
Contralateral/bilateral mediastinal
Scalene (inferior deep cervical), ipsilateral or contralateral
Supraclavicular (transverse cervical), ipsilateral or contralateral
Other distant lymph node(s)

Extension to:
Abdominal organs
Adjacent rib[##]
Contralateral lung
Contralateral main stem bronchus
Heart[##]
Pericardial effusion (malignant or NOS)
Pleural effusion (malignant or NOS)
Skeletal muscle
Skin of chest
Sternum
Vertebra(e)
Visceral pericardium[##]

Further contiguous extension

Separate tumor nodule(s) in different lobe[#*]

Separate tumor nodule(s) in contralateral lung

Metastasis

60

Steps to Assign SS2000

Three summary stage groups can be ruled out quickly:
in situ, distant, and localized
FINAL (if needed)

Regional

10. If in situ, local and distant categories have been ruled out, the stage is regional.
11. For carcinomas, if there are lymph nodes involved with the tumor, the stage is at least regional.
12. For tissues, structures, and lymph nodes, assume ipsilateral unless stated to be contralateral or bilateral.

Unknown if Extension or Metastasis

13. If there is not enough information in the record to categorize a case, it must be recorded as unstageable.

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SS2000

2 Regional by direct extension only

Atelectasis/obstructive pneumonitis

Extension to:
- Blood vessel(s) (major):
  - Aorta***
  - Azygos vein
  - Pulmonary artery or vein
  - Superior vena cava (SVC syndrome)
- Brachial plexus from superior sulcus###***
- Carina from lung
- Chest (thoracic) wall***
- Diaphragm***
- Esophagus
- Main stem bronchus <2.0 cm from carina
- Mediastinum, extrapulmonary or NOS
- Nerve(s):
  - Cervical sympathetic (Horner's syndrome)
  - Phrenic
  - Recurrent laryngeal (vocal cord paralysis)
  - Vagus
- Pancoast tumor (superior sulcus syndrome)###***
- Parietal (mediastinal) pleura***
- Parietal pericardium###
- Pericardium, NOS
- Pleura, NOS
- Pulmonary ligament
- Trachea
- Visceral pleura

Multiple masses/separate tumor nodule(s) in the SAME lobe#*
Multiple masses/separate tumor nodule(s) in the main stem bronchus

Tumor of main stem bronchus <2.0 cm from carina*

3 Regional IPSILATERAL regional lymph node(s) involved only

REGIONAL Lymph Nodes

- Aortic [above diaphragm], NOS:
  - Peri/para-aortic, NOS:
    - Ascending aorta (phrenic)
  - Subaortic (aortico-pulmonary window)
- Bronchial
- Carinal (tracheobronchial) (tracheal bifurcation)
- Hilar (bronchopulmonary) (proximal lobar) (pulmonary root)
- Intrapulmonary, NOS:
  - Interlobar
  - Lobar
  - Segmental
  - Subsegmental
- Mediastinal, NOS:
  - Anterior
  - Posterior (tracheoesophageal)
- Pericardial
- Peri/parabronchial
- Peri/paraesophageal
- Peri/paratracheal, NOS:
  - Azygos (lower peritracheal)
- Pre- and retrotracheal, NOS:
  - Precarinal
- Pulmonary ligament
- Subcarinal

Regional lymph node(s), NOS

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

Codes (2) + (3)

5 Regional, NOS

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SS2000

Note 1: "Bronchopneumonia" is not the same thing as "obstructive pneumonitis" and should not be coded as such.
Note 2: Assume tumor ≥2 cm from carina if lobectomy, segmental resection, or wedge resection is done.
Note 3: If no mention is made of the opposite lung on a chest x-ray, assume it is not involved.
Note 4: Ignore pleural effusion which is negative for tumor. Assume that a pleural effusion is negative if a resection is done.
Note 5: If at mediastinoscopy/x-ray, the description is "mass," "adenopathy," or "enlargement" of the mediastinum or of any of the lymph nodes listed under Regional Lymph Nodes (see page 150), assume that at least regional lymph nodes are involved.
Note 6: The words "no evidence of spread" and/or "remaining examination negative" are sufficient information to consider regional lymph nodes negative in the absence of any statement about nodes.
Note 7: "Vocal cord paralysis," "superior vena cava syndrome," and "compression of the trachea or the esophagus" are classified as mediastinal lymph node involvement unless there is a statement of involvement by direct extension from the primary tumor.

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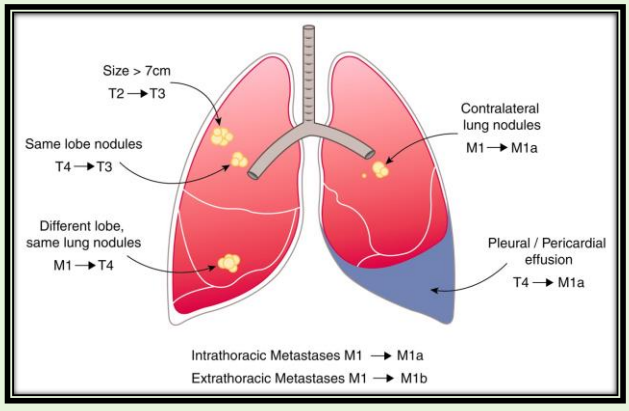
AJCC Staging for Lung Cancer



Nair A et al. Radiographics 2011;31:215-238

64

Clinical Classification - cTNM

- Based on Evidence Acquired Before Any Treatment
  - Physical Exam
  - Imaging (CT Scan, PET Scan)
  - Laboratory Tests
  - Thoracentesis
  - Endoscopy with ultrasound or other "directed" biopsy (core/FNA)
    - Bronchoscopy (EBUS)
    - Esophagoscopy (EUS)
    - Mediastinoscopy
    - Thoracoscopy (VATS without resection of primary tumor)
  - Exploratory Thoracotomy

65

Pathologic Classification - pTNM

- Includes Any Evidence Acquired Before Any Treatment PLUS
- Pathologic Assessment of Resected Primary Tumor (to highest pT) *
- Pathologic Assessment of Regional Lymph Nodes (to highest pN) *
- Isolated Tumor Cells (ITCs) in Lymph Node(s) are Classified N0 or M0
  - pN0
  - pN0(i-)
  - pN0(i+)
  - pN0(mol-)
  - pN0(mol+)
- pM can be either cM or pM when the T and/or N categories are valid

* *"Pathologic staging depends on the proven anatomic extent of disease, whether or not the primary lesion has been completely removed. If a biopsied primary tumor technically cannot be removed…and if the highest T and N categories or the M1 category of the tumor can be confirmed microscopically, the criteria for pathologic classification and staging have been satisfied without total removal of the primary cancer."*

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Post-Neoadjuvant p Classification - ypTNM

- **Best Demonstrates Need for Accurate Clinical Stage** when the first cancer surgery follows radiation therapy, chemotherapy, hormones, immunologic agents meant to alter the tumor behavior, size, extension, lymph node status, etc. resulting in down-stage of disease at time of first surgery and with some current regimens showing no primary tumor and negative nodes at surgery.

- *Patient must have received planned presurgical therapy(s):*
  - *Radiation Therapy (any modality)*
  - *Chemotherapy*
  - *Hormone(s)*
  - *Biologic Agent (BRM/Immuno)*
  - *Combination of above*

- *Patient must have post-therapy excision of primary site and nodes sufficient to meet the criteria to assign AJCC Stage Pathologic Classification or pTNM.*

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Definitions for T, N, M

Table 1. Definitions for T, N, M*

T	Primary Tumor
TX	Primary tumor cannot be assessed, or tumor proven by the presence of malignant cells in sputum or bronchial washings but not visualized by imaging or bronchoscopy
T0	No evidence of primary tumor
Tis	Carcinoma in situ
T1	Tumor ≤3 cm in greatest dimension, surrounded by lung or visceral pleura, without bronchoscopic evidence of invasion more proximal than the lobar bronchus (i.e., not in the main bronchus)[a]
T1a	Tumor ≤2 cm in greatest dimension
T1b	Tumor >2 cm but ≤3 cm in greatest dimension
T2	Tumor >3 cm but ≤7 cm or tumor with any of the following features:[b] Involves main bronchus, ≥2 cm distal to the carina Invades visceral pleura Associated with atelectasis or obstructive pneumonitis that extends to the hilar region but does not involve the entire lung
T2a	Tumor >3 cm but ≤5 cm in greatest dimension
T2b	Tumor >5 cm but ≤7 cm in greatest dimension
T3	Tumor >7 cm or one that directly invades any of the following: chest wall (including superior sulcus tumors), diaphragm, phrenic nerve, mediastinal pleura, parietal pericardium; or tumor in the main bronchus <2 cm distal to the carina[a] but without involvement of the carina; or associated atelectasis or obstructive pneumonitis of the entire lung or separate tumor nodule(s) in the same lobe
T4	Tumor of any size that invades any of the following: mediastinum, heart, great vessels, trachea, recurrent laryngeal nerve, esophagus, vertebral body, carina; separate tumor nodule(s) in a different ipsilateral lobe

N	Regional Lymph Nodes
NX	Regional lymph nodes cannot be assessed
N0	No regional lymph node metastasis
N1	Metastasis in ipsilateral peribronchial and/or ipsilateral hilar lymph nodes and intrapulmonary nodes, including involvement by direct extension
N2	Metastasis in ipsilateral mediastinal and/or subcarinal lymph node(s)
N3	Metastasis in contralateral mediastinal, contralateral hilar, ipsilateral or contralateral scalene, or supraclavicular lymph node(s)

M	Distant Metastasis
MX	Distant metastasis cannot be assessed
M0	No distant metastasis
M1	Distant metastasis
M1a	Separate tumor nodule(s) in a contralateral lobe; tumor with pleural nodules or malignant pleural (or pericardial) effusion[c]
M1b	Distant metastasis

[a]The uncommon superficial spreading tumor of any size with its invasive component limited to the bronchial wall, which may extend proximally to the main bronchus, is also classified as T1.
[b]T2 tumors with these features are classified T2a if ≤5 cm or if size cannot be determined, and T2b if >5 cm but ≤7 cm.
[c]Most pleural (and pericardial) effusions with lung cancer are due to tumor. In a few patients, however, multiple cytopathologic examinations of pleural (pericardial) fluid are negative for tumor, and the fluid is nonbloody and is not an exudate. Where these elements and clinical judgment dictate that the effusion is not related to the tumor, the effusion should be excluded as a staging element and the patient should be classified as T1, T2, T3, or T4.

*Used with permission. Goldstraw P, Crowley J, Chansky K, et al. The IASLC Lung Cancer Staging Project: Proposals for the revision of the TNM stage groupings in the forthcoming (seventh) edition of the TNM classification of malignant tumors. J Thorac Oncol 2007;2:706-714.

Source: NCCN Guidelines NSCLCv7.2015

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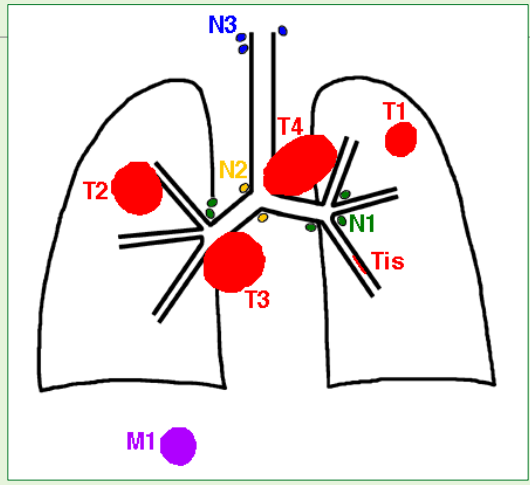
Definitions for T, N, M



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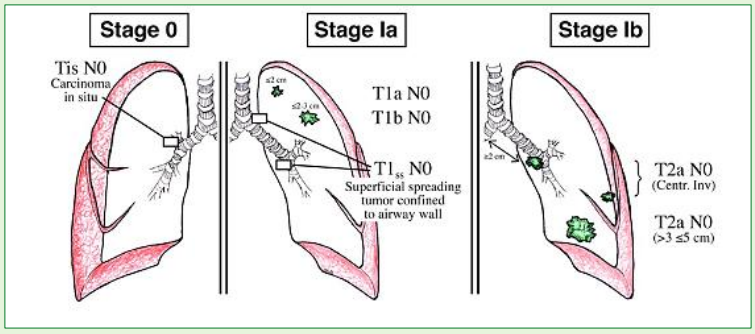
Anatomic Stage/Prognostic Groups



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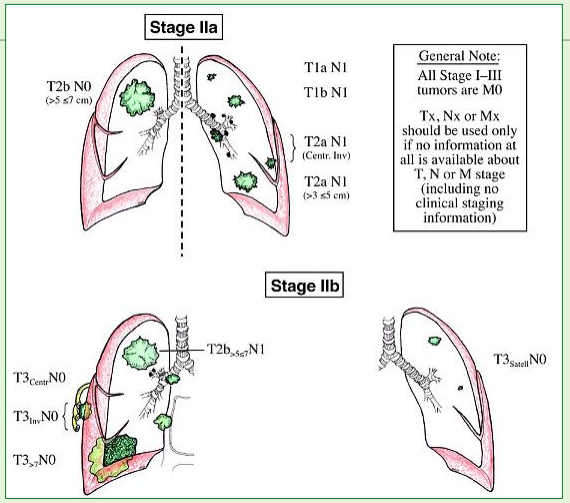
Anatomic Stage/Prognostic Groups



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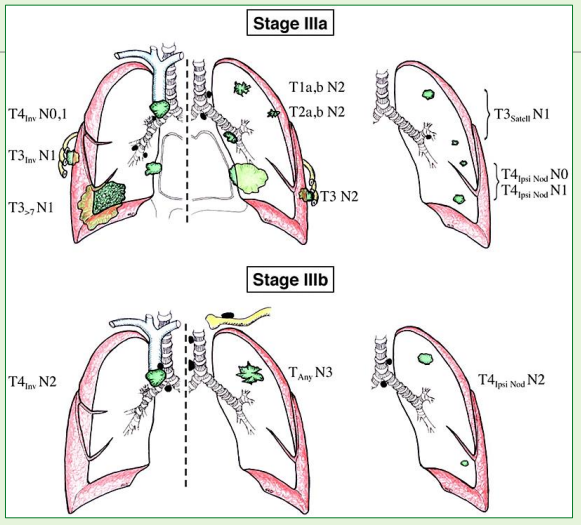
Anatomic Stage/Prognostic Groups



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Atelectasis and Pleural Effusion

Atelectasis is the collapse or closure of the lung that may affect part or all of one lung.

Atelectasis (lung collapse) may be caused by pleural effusion or may be caused by other factors that impair function of part or all of one lung.

Pleural effusion is excess fluid that accumulates between the two pleural layers, the fluid filled (pleural) space that surrounds the lungs.

Extensive amounts of pleural fluid can impair breathing by limiting the expansion of the lungs.

Pleural effusion in lung cancer is presumed malignant even following negative cytology from thoracentesis.

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Small Cell Lung Cancer VALG Stage

Veterans Administration Lung Study Group's (VALG) Staging Classification for Small Cell Lung Cancer

Limited-Stage: AJCC (7th edition) Stage I-III (excludes most T3-T4 due to multiplicity of tumors in same lung – cannot radiate for local control)

Extensive-Stage: AJCC (7th edition) Stage IV and most T3-T4

Still use AJCC TNM when can be more specific. But, most clinicians will refer to the VALG "limited" or "extensive" when assessing for treatment options, particularly for inclusion/exclusion of XRT to chest when T3-T4.

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IMIG Staging - Pleural Mesothelioma

International Mesothelioma Interest Group (IMIG) Staging System for Diffuse Malignant Pleural Mesothelioma*

T Primary Tumor

TX Primary tumor cannot be assessed
T0 No evidence of primary tumor
T1 Tumor limited to the ipsilateral parietal pleura with or without mediastinal pleura and with or without diaphragmatic pleural involvement
T1a No involvement of the visceral pleura
T1b Tumor also involving the visceral pleura
T2 Tumor involving each of the ipsilateral pleural surfaces (parietal, mediastinal, diaphragmatic, and visceral pleura) with a least one of the following:
-Involvement of the diaphragmatic muscle
-Extension of tumor from visceral pleura into the underlying pulmonary parenchyma
T3 Locally advanced but potentially resectable tumor. Tumor involving all of the ipsilateral pleural surfaces (parietal, mediastinal, diaphragmatic, and visceral pleura), with at least one of the following:
-Involvement of the endothoracic fascia
-Extension into the mediastinal fat
-Solitary, completely resectable focus of tumor extending into the soft tissues of the chest wall
-Nontransmural involvement of the pericardium
T4 Locally advanced technically unresectable tumor. Tumor involving all of the ipsilateral pleural surfaces (parietal, mediastinal, diaphragmatic, and visceral pleura) with at least one of the following:
-Diffuse extension or multifocal masses of tumor in the chest wall, with or without associated rib destruction
-Direct transdiaphragmatic extension of the tumor to the peritoneum
-Direct extension of tumor to the contralateral pleura
-Direct extension of the tumor to mediastinal organs
-Direct extension of tumor into the spine
-Tumor extending through to the internal surface of the pericardium with or without a pericardial effusion or tumor Involving the myocardium

N Regional Lymph Nodes

NX Regional lymph nodes cannot be assessed
N0 No regional lymph node metastasis
N1 Metastasis to the ipsilateral bronchpulmonary or hilar lymph nodes
N2 Metastases in the subcarinal lymph node or the ipsilateral mediastinal lymph nodes including the ipsilateral internal mammary and peridiaphragmatic nodes
N3 Metastasis in contralateral mediastinal, contralateral internal mammary, ipsilateral or contralateral supraclavicular lymph nodes

M Distant Metastasis

M0 No distant metastasis
M1 Distant metastasis

Stage Grouping

Stage	T	N	M
I	T1	N0	M0
IA	T1a	N0	M0
IB	T1b	N0	M0
II	T2	N0	M0
III	T1, T2	N1	M0
	T1, T2	N2	M0
	T3	N0, N1, N2	M0
IV	T4	Any N	M0
	Any T	N3	M0
	Any T	Any N	M1

*Used with the permission of the American Joint Committee on Cancer (AJCC), Chicago, Illinois. The original and primary source for this information is the AJCC Cancer Staging Manual, Seventh Edition (2010), published by Springer Science+Business Media, LLC (SBM). (For complete information and data supporting the staging tables, visit www.springer.com.) Any citation or quotation of this material must be credited to the AJCC as its primary source. The inclusion of this information herein does not authorize any reuse or further distribution without the expressed, written permission of Springer SBM, on behalf of the AJCC.

Source: IMIG Staging System for AJCC TNM, 7th ed. and NCCN Guidelinesv2.2015Mesothelioma 75

Case 1 – Case Vignette

HISTORY: 65 year old black male admitted with chest pain, cough, hoarseness and partial vocal cord paralysis. Hx of 1ppd smoker x 50yrs

CHEST X-RAY: 7.5cm mass noted in right mainstem bronchus

CT CHEST: 7.5cm mass right mainstem bronchus with supraclavicular node.

CT-GUIDED CORE BX RIGHT LUNG TUMOR MASS: Poorly differentiated squamous cell carcinoma. p63 and CK5 positive, Napsin and TTF1 neg - c/w squamous cell carcinoma of lung origin. *(positive immunohistochemistry for p63 and CK5 supports the diagnosis of squamous cell carcinoma. Negative immunohistochemistry for Napsin and TTF-1 argues against adenocarcinoma.)*

ULTRASOUND-GUIDED CORE BX SUPRACLAVICULAR MASS: positive for metastatic squamous cell carcinoma of pulmonary origin.

FINAL DX: Biopsy-proven unresectable squamous cell carcinoma of right lung with vocal cord paralysis and positive supraclavicular lymph node on FNA.

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Case 2 – Case Vignette

HISTORY: 70-year-old female developed right pleural effusion in January of 2015. Thoracentesis with bloody pleural fluid. Cytology showed no tumor cells. Patient was admitted and found to have a right pleural effusion with a pleural based mass and these were biopsied. Preliminary diagnosis between adenocarcinoma or mesothelioma. Pathology will do a TTF-1 and if positive, then more likely this is lung primary. If TTF-1 is negative, then we will have to make sure there is no other primary source of pleural effusion. She is a nonsmoker. Secondary smoke exposure - husband and father.

CT CHEST/ABD/PELVIS: nonspecific hilar and mediastinal lymph nodes. Soft tissue mass in RLL lung size 3.5 x 2.5cm. Extensive abnormal right pleural thickening with large right pleural effusion. Abdomen and pelvis – neg

PROCEDURE: Mini Thoracotomy with VATS wedge resection RLL lung.

RLL LUNG WEDGE RESECTION: moderately differentiated adenocarcinoma typical of lung primary with extensive visceral pleural invasion. TTF1 and CK7 positive and CK20 negative. This type of lung adenocarcinoma is sometimes referred to as "pseudomesotheliomatous" adenocarcinoma.

FINAL DX: Adenocarcinoma of lung, right lower lobe, stage IV.

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Case 3 – Case Vignette

HISTORY: 61 yr old Vietnamese man, smoker, with lung mass noted on CT. He has had repeated bouts of bronchopneumonia treated with antibiotics. He complains of shortness of breath, 15 pound weight loss, and mental status change. Admitted for workup and start of treatment.

CT CHEST/ABD/PELVIS: Large mass obstructing right upper lobe lung measuring at least 6cm with large mediastinal mass 5cm x 6cm in size. Large right-sided pleural effusion noted. Multiple cysts noted in liver.

MRI BRAIN: Diffuse 4th ventricle involvement with large cerebellar mass

BRONCHOSCOPY WITH BIOPSY: right upper lobe lung tumor, biopsy with small cell neuroendocrine (oat cell) carcinoma. CK7 +, Chromogranin + with SY38 positive consistent with small cell carcinoma of lung origin.

THORACENTESIS: pleural fluid + for malignant cells

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Case 4 – Case Vignette

HISTORY: 55 yr old white female, non-smoker, with lung mass seen on routine chest x-ray. No clinical symptoms or complaints. Admitted for workup and surgical treatment for left upper lobe lung cancer.

CT CHEST: 3cm tumor in left upper lobe lung no lymphadenopathy.

FNA LEFT LUNG TUMOR: non small cell carcinoma, favor adenocarcinoma

VATS WEDGE RESECTION LUL LUNG WITH NODE SAMPLING: moderately differentiated adenocarcinoma 2.5 x 2.8cm in size, wedge resection, with no involvement of surgical margins. 3 hilar lymph nodes sampled, 1 node with micrometastasis noted on IHC.

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Case 5 – Case Vignette

HISTORY: 59 year old white male firefighter with recently diagnosed unresectable mesothelioma of lung. Seen in ER then admitted with chest pain and shortness of breath prior to starting chemotherapy. Overall patient status is quite poor. Patient was discharged to hospice.

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Wrap Up – Final Q&A