Thoracic Imaging

Interlobar fissures

• The minor fissure separates the right upper lobe (RUL) from the right middle lobe

(RML) and is seen on both the frontal and lateral views as a fine horizontal line.

• The major (oblique) fissures are seen only on the lateral radiograph as oblique lines.

On the right, the major fissure separates the RUL and RML from the right lower lobe.

On the left, the major fissure separates the left upper lobe from the left lower lobe.

• The azygos fissure is an accessory fissure present in less than 1% of patients, seen in the presence of an azygos lobe.

An azygos lobe is an anatomic variant where the right upper lobe apical or posterior segments are encased in their own parietal and visceral pleura.

• Atelectasis is loss of lung volume due to decreased aeration.

Atelectasis is synonymous with collapse.

• **Direct signs of atelectasis** are from lobar volume loss and include:

• Air bronchograms are *not* seen in atelectasis when the cause of the atelectasis is central bronchial obstruction, but air bronchograms can be seen in subsegmental atelectasis.

Subsegmental atelectasis is caused by obstruction of small peripheral bronchi, usually by secretions.

• Subsegmental atelectasis and mild fever are both commonly encountered in postsurgical patients, although it has been proposed that there is no causative relationship between atelectasis and postoperative fever.

Mechanisms of atelectasis

• **Obstructive** atelectasis occurs when alveolar gas is absorbed by blood circulating through alveolar capillaries but is not replaced by inspired air due to bronchial obstruction.

Obstructive atelectasis can cause lobar atelectasis, which is complete collapse of a lobe.

Obstructive atelectasis occurs more quickly when the patient is breathing supplemental oxygen since oxygen is absorbed from the alveoli more rapidly than nitrogen.

In general, obstructive atelectasis is associated with volume loss. In critically ill ICU patients, however, there may be rapid transudation of fluid into the obstructed alveoli, causing

superimposed consolidation.

In children, airway obstruction is most often due to an aspirated foreign object.

In contrast to adults, the affected side becomes hyperexpanded in children due to a ball-valve effect.

Subsegmental atelectasis is a subtype of obstructive atelectasis commonly seen after surgery or general illness, due to mucus obstruction of the small airways.

• **Relaxation** (passive) atelectasis is caused by relaxation of lung adjacent to an intrathoracic lesion causing mass effect, such as a pleural effusion, pneumothorax, or pulmonary mass.

• **Adhesive** atelectasis is due to surfactant deficiency.

Adhesive atelectasis is seen most commonly in neonatal respiratory distress syndrome, but can also be seen in acute respiratory distress syndrome (ARDS).

• **Cicatricial** atelectasis is volume loss from architectural distortion of lung parenchyma by fibrosis.

Lobar atelectasis

• Lobar atelectasis is usually caused by central bronchial obstruction (obstructive atelectasis), which may be secondary to mucus plugging or an obstructing neoplasm.

If the lobar atelectasis occurs acutely, mucus plugging is the most likely cause.

If lobar atelectasis is seen in an outpatient, an obstructing central tumor must be ruled out.

• Lobar atelectasis, or collapse of an entire lobe, has characteristic appearances depending on which of the five lobes is collapsed.

• Each of the five lobes tends to collapse in a predictable direction.

• The *luftsichel* (*air-sickle* in German) sign of left upper lobe collapse is a crescent of air seen on the frontal radiograph, which represents the interface between the aorta and the hyperexpanded superior segment of the left lower lobe.

• It is important to recognize left upper lobe collapse and not mistake the left lung opacity for pneumonia, since a mass obstructing the airway may be the cause of the lobar atelectasis.

Right upper lobe atelectasis

• *The reverse S sign of Golden* is seen in right upper lobe collapse caused by an obstructing mass.

The central convex margins of the mass form a reverse S. Although the sign describes a *reverse* S, it is also commonly known as *Golden's S* sign.

Similar to left upper lobe collapse, a right upper lobe collapse should raise concern for an underlying malignancy, especially with a Golden's S sign present.

• The juxtaphrenic peak sign is a peridiaphragmatic triangular opacity caused by diaphragmatic traction from an inferior accessory fissure or an inferior pulmonary ligament.

Left lower lobe atelectasis

• In left lower lobe collapse, the heart slightly rotates and the left hilum is pulled down.

• The *flat waist* sign describes the flattening of the left heart border as a result of downward shift of hilar structures and resultant cardiac rotation.

Right lower lobe atelectasis

Right lower lobe atelectasis is the mirror-image of left lower lobe atelectasis.

• The collapsed lower lobe appears as a wedge-shaped retrocardiac opacity.

• The findings of right middle lobe atelectasis can be subtle on the frontal radiograph.

Silhouetting of the right heart border by the collapsed medial segment of the middle lobe may be the only clue.

The lateral radiograph shows a wedge-shaped opacity anteriorly.

Round atelectasis

• Round atelectasis is focal atelectasis with a round morphology that is *always* associated with an adjacent pleural abnormality (e.g., pleural effusion, pleural thickening or plaque,

pleural neoplasm, etc.).

• Round atelectasis is most common in the posterior lower lobes.

• All five of the following findings must be present to diagnose round atelectasis:

1) Adjacent pleura must be abnormal.

2) Opacity must be peripheral and in contact with the pleura.

3) Opacity must be round or elliptical.

4) Volume loss must be present in the affected lobe.

5) Pulmonary vessels and bronchi leading into the opacity must be curved — this is the *comet tail* sign.

Patterns of lung disease

Essential anatomy

Secondary pulmonary lobule (SPL)

• The secondary pulmonary lobule (SPL) is the elemental unit of lung function.

• Each SPL contains a central artery (the aptly named centrilobular artery) and a central bronchus, each branching many times to ultimately produce acinar arteries and

respiratory bronchioles.

On CT, the centrilobular artery is often visible as a faint dot. The centrilobular bronchus is not normally visible.

The acinus is the basic unit of gas exchange, containing several generations of branching respiratory bronchioles, alveolar ducts, and alveoli.

There are generally 12 or fewer acini per secondary lobule.

• Pulmonary veins and lymphatics collect in the periphery of each SPL.

• Connective tissue, called interlobular septa, encases each SPL.

Thickening of the interlobular septa can be seen on CT and suggests pathologic enlargement of either the venous or lymphatic spaces, as discussed on subsequent pages.

• Each SPL is between 1 and 2.5 cm in diameter.

Abnormalities of the secondary pulmonary lobule

Consolidation and ground glass

• Consolidation and ground glass opacification are two very commonly seen patterns of lung disease caused by abnormal alveoli.

The alveolar abnormality may represent either filling of the alveoli with fluid or incomplete alveolar aeration.

• consolidation can be described on either a chest radiograph or CT, while ground glass is generally reserved for CT.

• Although consolidation often implies pneumonia, both consolidation and ground glass are nonspecific findings with a broad differential depending on chronicity (acute versus chronic) and distribution (focal versus patchy or diffuse).

Consolidation

• Consolidation is histologically due to complete filling of affected alveoli with a liquidlike substance (commonly remembered as *blood, pus, water,* or *cells*).

• Pulmonary vessels are **not** visible through the consolidation on an unenhanced CT.

• *Air bronchograms* are often present if the airway is patent. An air bronchogram represents a lucent air-filled bronchus (or bronchiole) seen within a consolidation.

• Consolidation causes silhouetting of adjacent structures on conventional radiography.

• **Acute consolidation** is most commonly due to pneumonia, but the differential includes:

Pneumonia (by far the most common cause of acute consolidation).

Pulmonary hemorrhage (primary pulmonary hemorrhage or aspiration of hemorrhage).

Acute respiratory distress syndrome (ARDS), which is noncardiogenic pulmonary edema seen in critically ill patients and thought to be due to increased capillary permeability.

Pulmonary edema may cause consolidation, although this is an uncommon manifestation.

• The differential diagnosis of **chronic consolidation** includes:

Bronchioloalveolar carcinoma mucinous subtype, a form of adenocarcinoma.

Organizing pneumonia, which is a nonspecific response to injury characterized by granulation polyps which fill the distal airways, producing peripheral rounded and nodular consolidation.

Ground glass opacification (GGO)

• Ground glass opacification is histologically due to either partial filling of the alveoli (by blood, pus, water, or cells), alveolar wall thickening, or reduced aeration of alveoli (atelectasis).

• Ground glass is usually a term reserved for CT. CT shows a hazy, gauze-like opacity, through which pulmonary vessels are still visible.

The term ground glass was originally described for unenhanced CT as enhanced vessels are visible in consolidation as well; however, in common practice ground glass is used for any type of CT.

• As with consolidation, air bronchograms may be present.

• **Acute ground glass** opacification has a similar differential to acute consolidation, since many of the entities that initially cause partial airspace filling can progress to completely fill the airspaces later in the disease.

The differential of acute ground glass includes:

Pulmonary edema, which is usually dependent.

Pneumonia. Unlike consolidation, ground glass is more commonly seen in atypical pneumonia such as viral or *Pneumocystis jiroveci* pneumonia.

Pulmonary hemorrhage.

Acute respiratory distress syndrome (ARDS).

• **Chronic ground glass** opacification has a similar but broader differential diagnosis compared to chronic consolidation. In addition to all of the entities which may cause chronic consolidation, the differential diagnosis of chronic ground glass also includes:

Bronchioloalveolar carcinoma, which tends to be focal or multifocal.

Organizing pneumonia, typically presenting as rounded, peripheral chronic consolidation.

Chronic eosinophilic pneumonia, usually with an upper-lobe predominance.

Idiopathic pneumonias, which are a diverse group of inflammatory responses to pulmonary injury.

Hypersensitivity pneumonitis (HSP), especially the subacute phase. HSP is a type III hypersensitivity reaction to inhaled organic antigens. In the subacute phase there is ground glass,

centrilobular nodules, and mosaic attenuation.

Alveolar proteinosis, an idiopathic disease characterized by alveolar filling by a proteinaceous substance.

The distribution is typically central, with sparing of the periphery.

Chronic eosinophilic pneumonia, an inflammatory process characterized by eosinophils causing alveolar filling in an upper-lobe distribution.

Diffuse but central-predominant ground glass

The differential diagnosis for ground glass in a central distribution includes:

Pulmonary edema.

Alveolar hemorrhage.

Pneumocystititis jiroveci pneumonia.

Alveolar proteinosis.

Peripheral ground glass or consolidation

The differential diagnosis for **peripheral consolidation or ground glass** includes:

Organizing pneumonia.

Chronic eosinophilic pneumonia, typically with an upper lobe predominance.

Atypical or viral pneumonia.

Pulmonary edema. Peripheral pulmonary edema tends to be noncardiogenic in etiology, such as edema triggered by drug reaction. Peripheral consolidation/ground glass is unusual for

cardiogenic pulmonary edema.

Interlobular septal thickening – smooth

Conditions that dilate the pulmonary veins cause smooth interlobular septal thickening.

• By far the most common cause of **smooth interlobular septal thickening** is pulmonary edema; however, the differential diagnosis for smooth interlobular septal thickening is identical to the differential for central ground glass:

Pulmonary edema (by far the most common cause of smooth interlobular septal thickening).

Pulmonary alveolar proteinosis.

Pulmonary hemorrhage.

Atypical pneumonia, especially *Pneumocystis jiroveci* pneumonia.

Interlobular septal thickening – nodular, irregular, or asymmetric

Nodular, irregular, or asymmetric septal thickening tends to be caused by processes that infiltrate the peripheral lymphatics, most commonly lymphangitic carcinomatosis and sarcoidosis:

Lymphangitic carcinomatosis is tumor spread through the lymphatics.

Sarcoidosis is an idiopathic, multi-organ disease characterized by noncaseating granulomas, which form nodules and masses primarily in a lymphatic distribution.

Crazy paving

• *Crazy paving* describes interlobular septal thickening with superimposed ground glass opacification, which is thought to resemble the appearance of broken pieces of stone.

• Although nonspecific, this pattern was first described for alveolar proteinosis, where the ground glass opacification is caused by filling of alveoli by proteinaceous material and the interlobular septal thickening is caused by lymphatics taking up the same material.

• The differential diagnosis for **crazy paving** includes:

Alveolar proteinosis.

***Pneumocystis jiroveci* pneumonia**.

Organizing pneumonia.

Bronchioloalveolar carcinoma, mucinous subtype.

Lipoid pneumonia, an inflammatory pneumonia caused by a reaction to aspirated lipids.

Acute respiratory distress syndrome.

Pulmonary hemorrhage.

Approach to multiple nodules

Centrilobular nodules

• Centrilobular nodules represent opacification of the centrilobular bronchiole (or less commonly the centrilobular artery) at the center of each secondary pulmonary lobule.

• On CT, multiple small nodules are seen in the centers of secondary pulmonary lobules.

Centrilobular nodules never extend to the pleural surface. The nodules may be solid or of ground glass attenuation, and range in size from tiny up to a centimeter.

• Centrilobular nodules may be caused by infectious or inflammatory conditions.

• **Infectious** causes of **centrilobular nodules** include:

Endobronchial spread of tuberculosis or atypical mycobacteria. Atypical mycobacteria are a diverse spectrum of acid-fast mycobacteria that do not cause tuberculosis.

The typical pulmonary manifestation of atypical mycobacteria is a low-grade infection typically seen in elderly women, most commonly caused by *Mycobacterium avium-intracellulare.*

Bronchopneumonia, which is spread of infectious pneumonia via the airways.

Atypical pneumonia, especially **mycoplasma pneumonia**.

• The two most common **inflammatory** causes of **centrilobular nodules** include hypersensitivity pneumonitis (HSP) and respiratory bronchiolitis interstitial lung disease (RB-ILD), both exposure-related lung diseases.

More prominent centrilobular nodules are suggestive of HSP.

Perilymphatic nodules

Perilymphatic nodules:

• Perilymphatic nodules follow the anatomic locations of pulmonary lymphatics, three locations in the lung:

1) Subpleural.

2) Peribronchovascular.

3) Septal (within the interlobular septa separating the hexagonal secondary pulmonary lobules).

• Sarcoidosis is by far the most common cause of perilymphatic nodules, typically with an upper-lobe distribution. The nodules may become confluent creating the *galaxy* sign.

The differential of **perilymphatic nodules** includes:

Sarcoidosis.

Pneumoconioses (silicosis and coal workers pneumoconiosis) are reactions to inorganic dust inhalation.

The imaging may look identical to sarcoidosis with perilymphatic nodules, but there is usually a history of exposure (e.g. a sandblaster who develops silicosis).

Lymphangitic carcinomatosis.

Random nodules

Random nodules:

Schematic of the secondary pulmonary lobule (top left

image) demonstrates nodules distributed randomly

throughout the SPL.

Schematic of the lungs (bottom left image) demonstrates

nodules scattered randomly.

Some of the nodules are in close contact with the pleural surface.

Axial CT (top right image) demonstrates multiple random

nodules. Some of the nodules abut the pleural surface.

This was a case of metastatic colon cancer.

Randomly distributed nodules usually occur via hematogenous spread and have an angiocentric distribution.

The differential of random nodules includes:

Hematogenous metastases.

Septic emboli. Embolic infection has a propensity to cavitate but early emboli may be irregular or solid.

Pulmonary Langerhans's cell histiocytosis (PLCH), a smoking-related lung disease that progresses from airway-associated and random nodules to irregular cysts.

PLCH is usually distinguishable from other causes of random nodules due to the presence of cysts and non-angiocentric

distribution.

• A ***miliary*** **pattern** is innumerable tiny random nodules disseminated hematogenously, suggestive of the appearance of millet seeds.

The differential of miliary nodules includes:

Disseminated tuberculosis.

Disseminated fungal infection.

Disseminated hematogenous metastases.

Tree-in-bud nodules

Tree-in-bud nodules are multiple small nodules connected to linear branching structures, which resembles a budding tree branch in springtime as seen on CT.

The linear branching structures represent the impacted bronchioles, which are normally invisible on CT, and the nodules represent impacted terminal bronchioles.

Tree-in-bud nodules are due to mucus, pus, or fluid impacting bronchioles and terminal bronchioles.

• **Tree-in-bud nodules** are almost always associated with small airways infection, such as endobronchial spread of tuberculosis. The differential of tree-in-bud nodules includes:

Mycobacteria tuberculosis and **atypical mycobacteria**.

Bacterial pneumonia.

Aspiration pneumonia.

Airway-invasive aspergillus. Aspergillus is an opportunistic fungus with several patterns of disease.

The airway-invasive pattern is seen in immunocompromised patients and may present either as bronchopneumonia or small airways infection.

Cavitary and cystic lung disease

Solitary cavitary nodule/mass

A cavitary lesion has a thick, irregular wall, often with a solid mural component.

Although the findings of benign and malignant cavitary nodules overlap, a maximum wall thickness of ≤4 mm is usually benign and a wall thickness >15 mm is usually malignant.

Spiculated margins also suggest malignancy.

• A **solitary cavitary lesion** is most likely cancer or infection.

Primary bronchogenic carcinoma. While both squamous cell and adenocarcinoma can cavitate, squamous cell cavitates more frequently. Small cell carcinoma is never known to cavitate.

Tuberculosis classically produces an upper-lobe cavitation.

Multiple cavitary nodules

Multiple cavitary lesions are typically vascular or spread through the vascular system:

Septic emboli.

Vasculitis, including Wegener granulomatosis, which is especially prone to cavitate.

Metastases, of which squamous cell carcinoma and uterine carcinosarcoma are known to cavitate.

Cystic lung diseases

• A cyst is an air-containing lucency with a thin, nearly imperceptible wall.

In general, cystic lung disease is usually due to a primary airway abnormality.

• The differential diagnosis for multiple lung cysts includes:

• The differential for a single cyst includes:

Lymphangioleiomyomatosis (LAM), a diffuse cystic lung disease caused by smooth muscle proliferation of the distal airways.

LAM causes uniformly distributed, thin-walled cysts in a diffuse

distribution. It is classically associated with chylous effusion, as demonstrated in the above right case.

Emphysema, which tends to be upper-lobe predominant in a smoker.

Pulmonary Langerhans cell histiocytosis, which features irregular cysts and nodules predominantly in the upper lungs.

Diffuse cystic bronchiectasis. Bronchiectasis is dilation of the bronchioles.

Although cystic fibrosis is the most common cause of bronchiectasis and has an upper-lobe predominance, congenital or post-infectious causes can have a diffuse or lower-lobe distribution.

***Pneumocystis jiroveci* pneumonia**, which features cysts in late-stage disease.

Lymphoid interstitial pneumonia (LIP), an exceptionally rare disease usually associated with Sjögren syndrome and characterized by alveolar distortion from lymphocytic infiltrate and multiple cysts.

• The differential for a single cyst includes:

Bulla. A bulla is an air-filled cyst measuring >1 cm. A giant bulla occupies at least 30% of the volume of the thorax.

Bleb. A bleb is a air-filled cystic structure contiguous with the pleura measuring <1 cm. Rupture of a bleb is the most common cause of spontaneous pneumothorax.

Pneumatocele, which is an air-filled space caused by prior lung trauma or infection.

Fibrotic changes

Lower lobe fibrotic changes

The differential diagnosis of basal-predominant fibrotic change includes:

Upper lobe fibrotic changes

Coronal schematic shows fibrotic changes in the upper lobes.

Coronal CT shows upper-lobe predominant subpleural fibrosis and traction bronchiectasis.

A pathologic diagnosis was not established in this case.

• Although IPF is the most common cause of pulmonary fibrosis, fibrosis primarily affecting the upper lobes should raise concern for an alternative diagnoses, such as:

Idiopathic pulmonary fibrosis (IPF), which is a clinical syndrome of progressive pulmonary fibrosis of unknown etiology and is most common cause of basilar fibrosis. It almost always features basilar honeycombing.

End-stage asbestosis. Asbestosis is an asbestos-induced inflammatory process ultimately producing pulmonary fibrosis.

Usually other signs of asbestos exposure are present, such as pleural plaques.

Nonspecific interstitial pneumonia (NSIP), fibrotic form. NSIP is an idiopathic pneumonia. It is a lung response to injury

commonly associated with collagen vascular disease and drug reaction.

The two histologic subtypes are cellular and fibrotic forms, of which the latter may produce basalpredominant fibrosis. In contrast to IPF, honeycombing is usually absent.

Upper lobe fibrotic changes

• Although IPF is the most common cause of pulmonary fibrosis, fibrosis primarily affecting the upper lobes should raise concern for an alternative diagnoses, such as:

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End-stage sarcoidosis. Sarcoidosis is a disease that primarily affects the upper lobes. The late stage of sarcoidosis leads to upper-lobe predominant fibrosis.

Chronic hypersensitivity pneumonitis may cause upper-lobe fibrosis in long-standing disease.

Pulmonary infection

Clinical classification of pneumonia

Community-acquired pneumonia (CAP)

• *S. pneumoniae* is the most common cause of community-acquired pneumonia (CAP).

• Atypical pneumonia, including *Mycoplasma*, viral, and *Chlamydia*, typically infects young and otherwise healthy patients.

Mycoplasma has a varied appearance and can produce consolidation, areas of ground glass attenuation, centrilobular nodules, and tree-in-bud nodules.

• *Legionella* most commonly occurs in elderly smokers. Infections tend to be severe.

Peripheral consolidation often progresses to lobar and multifocal pneumonia.

• Infection by *Klebsiella* and other gram-negatives occurs in alcoholics and aspirators.

Klebsiella classically leads to voluminous inflammatory exudates causing the *bulging fissure* sign.

End-stage silicosis. The late stage of silicosis may lead to fibrosis with an upper lobe predominance.

Hospital acquired pneumonia (HAP)

• Hospital acquired pneumonia (HAP) occurs in hospitalized patients and is due to aspiration of colonized secretions. HAP is caused by a wide variety of organisms, but the most important pathogens include MRSA and resistant gram-negatives including *Pseudomonas*.

Health care associated pneumonia (HCAP)

• Health care associated pneumonia is defined as pneumonia in a nursing home resident or in a patient with a >2 day hospitalization over the past 90 days.

Pathogens

are similar to HAP.

Ventilator associated pneumonia (VAP)

• Ventilator associated pneumonia is caused by infectious agents not present at the time mechanical ventilation was started. Most infections are polymicrobial and primarily involve gram-negative rods such as *Pseudomonas* and *Acinetobacter*.

Pneumonia in the immunocompromised patient

• Any of the above pathogens, plus opportunistic infections including *Pneumocystis*, fungi such as *Aspergillus*, *Nocardia*, CMV, etc., can be seen in immunocompromised patients.

Radiographic patterns of infection

Lobar pneumonia

• Lobar pneumonia is consolidation of a single lobe. It is usually bacterial in origin and is the most common presentation of community acquired pneumonia.

• The larger bronchi remain patent, causing air bronchograms.

Lobular pneumonia (bronchopneumonia)

• Lobular pneumonia manifests as patchy consolidation with poorly defined airspace opacities, usually involving several lobes, and most commonly due to *S. aureus*.

Interstitial pneumonia

• Interstitial pneumonia is caused by inflammatory cells located predominantly in the interstitial tissue of the alveolar septa causing diffuse or patchy ground glass opacification.

It can be caused by viral pneumonia, *Mycoplasma*, *Chlamydia*, or *Pneumocystis*.

Round pneumonia

• Round pneumonia is an infectious mass-like opacity seen only in children, most commonly due to *Streptococcus pneumoniae*.

• Infection remains somewhat confined due to incomplete formation of pores of Kohn.

Complications of pneumonia

Pulmonary abscess

• Pulmonary abscess is necrosis of the lung parenchyma typically due to *Staphylococcus aureus*, *Pseudomonas*, or anaerobic bacteria.

• An air–fluid level is almost always present.

• An abscess is usually spherical, with equal dimensions on frontal and lateral views.

Pulmonary gangrene

• Pulmonary gangrene is a very rare complication of pneumonia where there is extensive necrosis or sloughing of a pulmonary segment or lobe.

Pulmonary gangrene is a severe manifestation of pulmonary abscess.

Empyema

• Empyema is infection within the pleural space.

• There are three stages in the development of an empyema:

1) **Free-flowing exudative effusion**: Can be treated with needle aspiration or simple drain.

2) **Development of fibrous strands**: Requires large-bore chest tube and fibrinolytic therapy.

3) **Fluid becomes solid and jelly-like**: Usually requires surgery.

• Although pneumonia is often associated with a parapneumonic effusion, most pleural effusions associated with pneumonia are *not* empyema, but are instead a sterile effusion caused by increased capillary permeability.

• An empyema conforms to the shape of the pleural space, causing a longer air–fluid level on the lateral radiograph. This is in contrast to an abscess, discussed above, which typically is spherical and has the same dimensions on the frontal and lateral radiographs.

• The *split pleura* sign describes enhancing parietal and visceral pleura of an empyema seen on contrast-enhanced study.

Pneumatocele

• A pneumatocele is a thin-walled, gas-filled cyst that may be post-traumatic or develop as a sequela of pneumonia, typically from *Staphylococcus aureus* or *Pneumocystis*.

• Pneumatoceles almost always resolve.

Bronchopleural fistula (BPF)

• Bronchopleural fistula (BPF) is an abnormal communication between the airway and the pleural space.

It is caused by rupture of the visceral pleura. By far the most common cause of BPF is surgery; however, other etiologies include lung abscess, empyema, and trauma.

• **On imaging**, new or increasing gas is present in a pleural effusion. A connection between the bronchial tree and the pleura is not always apparent, but is helpful when seen.

• The treatment of BPF is controversial and highly individualized.

Empyema necessitans

• Empyema necessitans is extension of an empyema to the chest wall, most commonly secondary to tuberculosis.

Other causative organisms include *Nocardia* and *Actinomyces*.

Tuberculosis (TB)

• Tuberculosis (TB), caused by *Mycobacterium tuberculosis*, remains an important disease despite remarkable progress in public health and antituberculous therapy over the past century. Tuberculosis remains a significant problem in developing

countries.

In the United States, TB is seen primarily in the immigrant population and immunocompromised individuals.

• Initial exposure to TB can lead to two clinical outcomes:

1) **Contained disease (90%)** results in calcified granulomas and/or calcified hilar lymph nodes.

In a patient with normal immunity, the tuberculous bacilli are sequestered with a caseating granulomatous response.

2) **Primary tuberculosis** results when the host cannot contain the organism. Primary tuberculosis is seen more commonly in children and immunocompromised patients.

• **Reactivation (post-primary)** TB is reactivation of a previously latent infection.

Primary tuberculosis

Primary tuberculosis represents infection from the first exposure to TB. Primary TB may involve the pulmonary parenchyma, the airways, and the pleura.

Primary TB often causes adenopathy.

• As many as 15% of patients infected with primary TB have no radiographic changes and the imaging appearance of primary tuberculosis is nonspecific.

• The four imaging manifestations of primary TB (of which any, none, or all may be present) are ill-defined consolidation, pleural effusion, lymphadenopathy, and miliary disease.

Primary TB may occur in any lobe, but the most typical locations are the lower lobes or right middle lobe. It can be difficult to distinguish between primary and postprimary

TB, and in clinical practice, the treatment (antituberculous therapy) is the same.

• Classic imaging findings are not always seen, but include:

Ghon focus: Initial focus of parenchymal infection, usually located in the upper part of the lower lobe or the lower part of the upper lobe.

Ranke complex: *Ghon focus* and lymphadenopathy.

• Cavitation is rare in primary TB, in contrast to reactivation TB.

Adenopathy is common in primary TB, typically featuring central low-attenuation and peripheral enhancement, especially in children.

In contrast, post-primary TB does not feature prominent adenopathy.

Reactivation (post-primary) tuberculosis

Reactivation TB, also called post-primary TB, usually occurs in adolescents and adults and is caused by reactivation of a dormant infection acquired earlier in life.

• Reactivation TB most commonly occurs in the upper lobe apical and posterior segments.

• In an immunocompetent patient, the imaging hallmarks of reactivation TB are upper lobe predominant disease with cavitation and lack of adenopathy. Focal upper lobe

consolidation and endobronchial spread are common. Although not specific to TB, tree-in-bud nodules suggest active **endobronchial spread**.

• In an immunosuppressed patient (such as HIV), low-attenuation adenopathy is a typical additional finding, similar to

the adenopathy seen in primary TB. Low density lymph nodes may mimic immune reconstitution syndrome in HIV patients.

• A tuberculoma is a well-defined rounded opacity usually in the upper lobes.

Healed tuberculosis

• Healed TB is evident on radiography as apical scarring, usually with upper lobe volume loss and superior hilar retraction.

• Calcified granulomas may be present as well, which indicate containment of the initialinfection by a delayed hypersensitivity response.

Miliary tuberculosis

Miliary tuberculosis is a diffuse random distribution of tiny nodules seen in hematogenously disseminated TB.

• Miliary TB can occur in primary or reactivation TB.

Atypical mycobacteria

Atypical mycobacteria infection

• The classic presentation of atypical mycobacteria is an elderly woman with cough, low-grade fever, and weight loss, called *Lady Windermere* syndrome.

Mycobacterium avium intracellulare and *M. kansasii* are the two most common organisms.

• Classic radiographic findings are bronchiectasis and tree-in-bud nodules, most common in the right middle lobe or lingula.

“Hot-tub” lung

• “Hot-tub” lung is a hypersensitivity pneumonitis in response to atypical mycobacteria, which are often found in hot tubs. There is no active infection and the typical patient is otherwise healthy. Imaging is similar to other causes of hypersensitivity pneumonitis, featuring centrilobular nodules.

Endemic fungi

• Endemic fungi can cause community acquired pneumonia in normal individuals, with each subtype having a specific geographic distribution.

Most infected patients are asymptomatic.

Histoplasma capsulatum

• *Histoplasma capsulatum* is localized to the Ohio and Mississippi river valleys, in soil contaminated with bat or bird guano.

• The most common sequela of infection is a calcified granuloma.

A less common radiologic manifestation is a pulmonary nodule (histoplasmoma), which can mimic a neoplasm.

• Chronic infection can mimic reactivation TB with upper lobe fibrocavitary consolidation.

• Fibrosing mediastinitis is a rare complication of *Histoplasma* infection of mediastinal lymph nodes leading to pulmonary

venous obstruction, bronchial stenosis, and pulmonary artery stenosis. Affected lymph nodes tend to calcify.

Coccidioides immitis and Blastomyces dermatitidis

• *Coccidioides immitis* is found in the southwestern US and has a variety of radiologic appearances, including multifocal consolidation, multiple pulmonary nodules, and

miliary nodules.

• *Blastomyces dermatitidis* is found in central and southeastern US. Infection is usually asymptomatic, but may present as flu-like illness that can progress to multifocal consolidation, ARDS, or miliary disease.

Infections in the immunocompromised

• Immunosuppressed patients are susceptible to the same organisms that infect immunocompetent patients; however, one must be aware of several additional opportunistic organisms that may present in the immunocompromised.

• An immunocompromised patient with a focal air space opacity is most likely to have a bacterial pneumonia (most commonly pneumococcus), but TB should also be considered if the CD4 count is low.

• In contrast, multifocal opacities have a wider differential diagnosis including *Pneumocystis* pneumonia and opportunistic fungal infection such as *cryptococcus* or *aspergillus*.

Pneumocystis jiroveci pneumonia

Pneumocystis jiroveci (previously called Pneumocystis carinii) is an opportunistic fungus that may cause pneumonia in individuals with CD4 counts <200 cells/cc.

The incidence of Pneumocystis pneumonia is decreasing due to routine antibiotic prophylaxis.

• Chest radiograph findings can be normal but a classic finding of Pneumocystis pneumonia is **bilateral perihilar (central) airspace opacities** with peripheral sparing.

• The classic CT appearance is geometric **perihilar ground glass opacification**, sometimes with crazy paving (ground glass and thickening of the interlobular septa).

• A normal CT rules out Pneumocystis pneumonia; however, the disease can hide in a normal chest radiograph.

• Pneumocystis pneumonia has a propensity to cause upper lobe pneumatoceles, which may predispose to pneumothorax or pneumomediastinum.

Cryptococcus neoformans

• *Cryptococcus* is an opportunistic organism and is the most common fungal infection in AIDS patients.

Pulmonary infection usually coexists with cryptococcal

meningitis.

Typically CD4 count is less than 100 cells/cc in affected individuals.

• In the immunosuppressed, *Cryptococcus* can have a wide range of appearances ranging from ground glass attenuation to focal consolidation to cavitating nodules.

Cryptococcus can also present as miliary disease, often associated with lymphadenopathy or effusions.

Aspergillus

Overview of Aspergillus

Aspergillus is a ubiquitous soil fungus that manifests as five distinct categories of pulmonary disease.

Aspergillus only affects individuals with abnormal immunity or

preexisting pulmonary disease. Depending on the manifestation, the predisposing abnormality may include asthma, immunocompromised state, prior infection, or structural/congenital abnormality.

Allergic bronchopulmonary aspergillosis (ABPA)

• Allergic bronchopulmonary aspergillosis (ABPA) is a hypersensitivity reaction to aspergillus seen most commonly in patients with long-standing asthma.

• Patients present clinically with recurrent wheezing, low-grade fever, cough, and sputum production.

The sputum contains fragments of aspergillus hyphae.

• The key finding on CT is upper lobe bronchiectasis and mucoid impaction, which can be high attenuation or even calcified. This combination of mucoid impaction within

bronchiectatic airways represents the *finger-in-glove* sign.

The *finger-in-glove* sign is not specific to ABPA and can also been seen in segmental bronchial atresia and many other diseases including cystic fibrosis (less commonly seen on a case-by-case basis).

Saprophytic aspergillosis (aspergilloma)

• An aspergilloma is a conglomeration of intertwined aspergillus fungal hyphae and cellular debris (a mycetoma or “fungus ball”)

in a *preexisting pulmonary cavity.*

The aspergilloma is mobile and will change position when the patient is imaged in a different position.

• The most common causes of a preexisting cavity are prior tuberculosis and sarcoidosis.

Less common causes include congenital anomalies such as bronchogenic cyst or sequestration, and post-infectious/post-traumatic pneumatocele.

• If an aspergilloma is symptomatic, hemoptysis

is the most common symptom.

• When a crescent of air is seen outlining the mycetoma against the wall of the cavity, the correct term is *Monod* sign.

The *air crescent* sign is reserved for angioinvasive aspergillus.

Semi-invasive (chronic necrotizing) aspergillosis

• Semi-invasive aspergillosis is a necrotizing granulomatous inflammation (analogous in pathology to reactivation TB) in response to chronic aspergillus infection. Semiinvasive

aspergillosis is seen in debilitated, diabetic, alcoholic, and COPD patients.

• Clinical symptoms include cough, chronic fever, and less commonly hemoptysis.

• On CT, there are segmental areas of consolidation, often with cavitation and pleural thickening, which progress slowly over months or years.

Airway-invasive aspergillosis

• Airway-invasive aspergillosis is aspergillus infection deep to the airway epithelial cells.

It is seen only in the immunocompromised, including neutropenic and AIDS patients.

• The spectrum of clinical disease ranges from bronchiolitis to bronchopneumonia.

• The main CT findings of airway-invasive aspergillosis are centrilobular and tree-inbud nodules.

When bronchopneumonia is present, radiograph and CT findings are indistinguishable from other causes of bronchopneumonia, such as *Staph. aureus*.

Angioinvasive aspergillosis

• Angioinvasive aspergillosis is an aggressive infection characterized by invasion and occlusion of arterioles and smaller pulmonary arteries by fungal hyphae.

Angioinvasive aspergillosis is seen almost exclusively in the severely immunocompromised, including patients on chemotherapy, stem cell or solid organ transplant recipients, and in AIDS.

• The CT *halo* sign represents a halo of ground glass attenuation surrounding a consolidation.

The ground glass is thought to correspond to hemorrhagic infarction of the lung.

• The *air crescent* sign, visually similar to the Monod sign surrounding a mycetoma, represents a crescent of air from retraction of infarcted lung. It a good prognostic sign as it indicates that the patient is in the recovery phase.

Pulmonary edema and ICU Imaging

Pulmonary edema

Overview of pulmonary edema

• The radiographic severity of pulmonary edema typically progresses through three stages, corresponding to progressively increased pulmonary venous pressures.

• Vascular redistribution is the first radiographic sign of increased pulmonary venous pressure.

Imaging shows increased caliber of the upper lobe vessels compared to the lower lobe vessels.

• Interstitial edema is caused by increased fluid within the pulmonary veins, which surround the periphery of each hexagonal secondary pulmonary lobule.

On radiography, there are increased interstitial markings, indistinctness of the pulmonary vasculature, peribronchial cuffing, and Kerley B and A lines.

Kerley B lines are seen at the peripheral lung and represent thickened interlobular septa.

Kerley A lines radiate outward from the hila and may represent dilation of lymphatic channels.

They are not thought to be clinically relevant.

• Alveolar edema is caused by filling of the alveoli with fluid. On imaging, perihilar (central) opacifications are present. Pleural effusions and cardiomegaly are often

present.

• CT findings of pulmonary edema include dependent ground glass opacification and interlobular septal thickening. Intrathoracic causes of pulmonary edema, such as heart failure, generally cause patchy ground glass, while systemic cause of pulmonary edema (e.g., sepsis or low protein states) often cause diffuse ground glass.

• Pulmonary edema is usually symmetric and dependent. A classic cause of asymmetric pulmonary edema is isolated right

upper lobe pulmonary edema, caused by acute mitral regurgitation secondary to myocardial infarction and

papillary muscle rupture.

• A complication of aggressive thoracentesis is **reexpansion pulmonary edema**, caused by rapid reexpansion of a lung in a state of collapse for more than three days.

Vascular pedicle

• The vascular pedicle is the transverse width of the upper mediastinum.

The right border of the vascular pedicle is the interface of the superior vena cava (SVC) and the right mainstem bronchus.

The left border of the vascular pedicle is the lateral border of the takeoff of the subclavian from the aorta.

• The vascular pedicle width is normally <58 mm.

• An interval increase in vascular pedicle width (both >63 and >70 mm have been proposed as cutoffs) on sequential supine AP ICU-type chest radiographs generally correlates with increased pulmonary capillary wedge pressure (>18 mm Hg) and fluid overload.

Support devices

Endotracheal tube

• The endotracheal tube tip should be approximately 4–6 cm above the carina with the neck in neutral alignment.

However, in situations with low pulmonary compliance (e.g.,

ARDS), a tip position closer to the carina may reduce barotrauma.

• Direct intubation of either the

right or left mainstem bronchus (right mainstem bronchus far more common) is an emergent finding that can cause complete atelectasis of the un-intubated lung.

Central venous catheters

• The tip of a central venous catheter, including a PICC, should be in lower SVC or the cavoatrial junction.

Azygos malposition is seen in approximately 1% of bedside-placed PICCs.

Azygos malposition is associated with increased risk of venous perforation and catheter-associated thrombosis, and repositioning is recommended.

• A dialysis catheter should be located in the right atrium.

• The tip of a Swan–Ganz pulmonary artery catheter should be in either the main, right, or left pulmonary artery.

• If the tip is distal to the proximal interlobar pulmonary artery, there is a risk of pulmonary artery rupture or pseudoaneurysm. Other complications of pulmonary artery catheter placement include intracardiac catheter knot and arrhythmia.

Lung cancer

Clinical overview of lung cancer

Epidemiology

• Lung cancer is the leading cause of cancer death in the USA.

• Including all stages and subtypes, the 5-year survival is 15%.

Risk factors for lung cancer

• Tobacco smoking is thought to cause 80–90% of lung cancers.

Almost all cases of squamous cell and small cell carcinoma are seen in smokers.

Adenocarcinoma is also associated with smoking, but primary bronchogenic carcinoma arising in a lifelong nonsmoker with no history of secondhand exposure is almost always adenocarcinoma.

• Occupational and environmental exposures, including beryllium, radon, arsenic, etc., remain an important risk factor for lung cancer.

Asbestos exposure increases the risk of lung cancer by a factor of five, synergistic with smoking.

• Pulmonary fibrosis increases the risk of lung cancer by a factor often.

• Pulmonary scarring, such as from prior TB, also increases the risk of lung cancer.

Solitary pulmonary nodule

Overview of the solitary pulmonary nodule

• Pulmonary nodules are very common and the vast majority are benign; however, nodules are often followed with serial CT scans to screen for development of lung cancer.

• Calcified nodules are usually benign.

• Although less commonly seen, ground glass nodules (or mixed attenuation nodules containing both solid and ground glass) are more likely to be malignant than a solid nodule.

Nodule morphology essentially diagnostic for a benign etiology

• Central, laminar, and diffuse calcification are almost always benign.

• Popcorn calcification, suggestive of a pulmonary hamartoma, is benign.

• Intra-lesional fat, suggestive of hamartoma or lipoid granuloma, is benign.

Nodule morphology suggesting, but not diagnostic for, a benign etiology

• Small nodules are usually benign. A nodule <3 mm has a 0.2% chance of being cancer and a 4–7 mm nodule is malignant in 2.7% of cases.

• Any calcification in a small nodule is usually benign.

• Non-round shape, including oblong, polygonal, triangular, or geometric is probably benign.

• Subpleural location is often benign.

• Clustering of nodules suggests an infectious process.

Nodule morphology suggesting malignancy

• Large size is the single most important risk factor for malignancy, regardless of morphology: 0.8 to 3 cm nodules have 18% risk of being lung cancer and masses >3 cm have a very high chance of being malignant.

• Irregular edge or spiculated margin is concerning.

• Round shape (as opposed to oblong) is suggestive of malignancy.

• A cavitary nodule or nodule containing small cystic spaces is suspicious for malignancy.

• Follow-up is not recommended for a solitary pulmonary nodule if the nodule is small (<4 mm) and the patient doesn’t have a history of smoking or other risk factors.

• Any interval nodule growth is suspicious. A 26% increase in diameter (for instance, from 1.0 to 1.26 cm) is a doubling in volume.

Doubling time for lung cancers ranges from 42 days in very aggressive tumors to over 4 years in indolent lesions such as bronchioloalveolar carcinoma.

• A nodule that has not changed in size over 2 years is very likely, but not definitely, benign. Follow-up of ground glass nodules, which are often indolent bronchioloalveolar carcinoma, may be appropriate beyond 2 years.

• A decrease in size of a suspicious nodule on a single follow-up study is not sufficient to establish a benign etiology.

Transient decrease in size of a malignant lesion can occur with collapse of aerated alveoli or necrosis.

• The Fleischner Society (2005) suggests the following follow-up recommendations for noncalcified nodules in patients older than 35 without a history of malignancy.

A high risk patient is defined as a patient with a history of smoking or other risk factors for lung cancer.

Nodule ≤4 mm Low-risk: No follow-up needed. High-risk: At least one follow-up at 12 months. If unchanged, no further follow-up.

Nodule >4 and ≤6 mm Low-risk: At least one follow-up at 12 months. If unchanged, no further follow-up.

High-risk: At least two follow-ups at 6–12 months and 18–24 months if no change. Nodule >6 and ≤8 mm Low-risk: At least two follow-ups at 6–12 months and 18–24 months if no change.

High-risk: At least three follow-ups at 3–6 months, 9–12, and 24 months if no change.

Nodule >8 mm regardless of risk, either PET, biopsy, or at least three follow ups at 3, 9, and 24 months.

Histologic subtypes of lung cancer

• Lung cancer can be thought of as two types: “Small cell” and all other histologic types that are not small cell, such as adenocarcinoma, squamous cell carcinoma, etc.

However, although previously small cell and non-small cell used separate staging systems, the 2009 staging revision unifies the staging of all types.

• Small cell is usually disseminated at diagnosis and has a much worse prognosis.

Adenocarcinoma

• Adenocarcinoma is the most common subtype of lung cancer. It is related to smoking, but less strongly than squamous cell.

• Adenocarcinoma tends to occur in the peripheral lung.

• The typical radiographic appearance of adenocarcinoma is of a pulmonary nodule, which often has a spiculated margin due to reactive fibrosis.

• Cavitation can occur but is less commonly seen compared to squamous cell.

• A useful pathologic marker is TTF-1 (thyroid transcription factor), which is positive in primary lung adenocarcinoma and negative in pulmonary metastases from an extrathoracic adenocarcinoma.

Squamous cell carcinoma (SCC)

• Squamous cell carcinoma (SCC) is slightly less

common today than adenocarcinoma. Prior to filtered cigarettes, SCC was more common.

• The majority of SCC arise centrally from main, lobar, or segmental bronchi, where the tumor tends to cause symptoms early due to bronchial obstruction.

SCC may also present as a hilar mass.

• Common radiographic findings are

lobar atelectasis, mucoid impaction, consolidation, and bronchiectasis. SCC has a propensity to cavitate.

SCC: Axial CT shows a spiculated cavitary lesion

(arrow) shown at pathology to be squamous

cell carcinoma.

Bronchioloalveolar carcinoma (BAC)

• Bronchioloalveolar carcinoma (BAC) refers to a spectrum of well-differentiated adenocarcinoma that demonstrates *lepidic* growth.

The hallmark of lepidic growth is a spreading of malignant cells using the alveolar walls as a scaffold.

The opposite of *lepidic* growth is *hilic* growth, demonstrated by most other forms of lung cancer, which describes cancer growth by invasion and destruction of lung parenchyma.

• A spectrum of lesions have been called BAC ranging from small peripheral tumors with 100% survival to lesions causing widespread advanced disease.

Despite this variability, BAC is most commonly indolent and is often negative on PET.

• To create more uniformity in the pathological, clinical, and research domains, a new classification for the spectrum of the adenocarcinoma subtypes formerly called BAC has recently been proposed.

This classification is primarily based on the pathology of the lesion and differentiation of these entities on imaging is difficult. In clinical radiologic practice, this spectrum of lesions is routinely still referred to as BAC.

This textbook will continue the common practice and refer to these lesions as BAC.

Adenomatous hyperplasia (AAH):

A precursor lesion.

Adenocarcinoma in situ:

A preinvasive lesion.

Minimally invasive adenocarcinoma.

Adenocarcinoma, predominantly invasive with some nonmucinous lepidic component:

Formerly nonmucinous BAC.

Invasive mucinous adenocarcinoma:

Formerly mucinous BAC. Nonmucinous and mucinous subtypes of BAC occur with approximately equal

prevalence.

• **Nonmucinous BAC** (*adenocarcinoma, predominantly invasive with some nonmucinous lepidic component*) classically presents as a ground glass or solid nodule with air bronchograms and has a better prognosis compared to the mucinous subtype.

• **Mucinous BAC** (invasive mucinous adenocarcinoma) tends to present with chronicconsolidation. It has a worse prognosis compared with non-mucinous BAC.

Mucinous BAC is an important differential consideration for chronic ground glass or consolidation, often with air bronchograms.

The *CT angiogram* sign (not imaged above) describes the especially prominent appearance of enhancing pulmonary vessels seen in a low attenuation, mucin-rich consolidation of mucinous BAC.

Small cell carcinoma

• Small cell carcinoma is the third most common lung cancer cell type (after adenocarcinoma and squamous cell).

Neoplastic cells are of neuroendocrine origin and are associated with various paraneoplastic syndromes.

• Small cell carcinoma is strongly associated with smoking.

• Small cell tends to occur in central bronchi with invasion through the bronchial wall, typically presenting as a large hilar or parahilar mass.

Involvement of the SVC may cause SVC syndrome.

Small cell rarely presents as a solitary pulmonary nodule.

• Small cell is considered a disseminated disease and is generally not amenable to surgery.

Large cell carcinoma

• Large cell carcinoma is a wastebasket pathologic diagnosis for tumors that are not squamous, adenocarcinoma, or small cell. Large cell carcinoma is strongly associated with smoking and has a poor prognosis.

• Large cell carcinoma often occurs in the lung periphery, where it presents as a large mass.

Carcinoid tumor

• Neoplastic carcinoid cells originate from neuroendocrine cells in the bronchial walls.

• A common presentation of carcinoid is an endobronchial mass distal to the carina, which may cause obstructive atelectasis. Up to 20% of cases present as a pulmonary nodule.

• Carcinoid may be typical (low-grade) or the more aggressive atypical variant.

Typical carcinoids without nodal or distant metastases have an excellent prognosis (92% 5-year survival).

Atypical carcinoids tend to arise peripherally and have a worse prognosis.

• Diffuse idiopathic pulmonary neuroendocrine cell hyperplasia (DIPNECH) is an extremely uncommon precursor lesion to typical carcinoid tumor, characterized by multiple foci of neuroendocrine hyperplasia or tumorlets (carcinoid foci <5 mm in size) and bronchiolitis obliterans.

Radiologic presentation of lung cancer

Solitary pulmonary nodule or lung mass

• A nodule is defined as <3 cm and a mass is >3 cm.

• Adenocarcinoma, including subtypes formerly called BAC, comprises approximately 50% of cancers presenting as a solitary pulmonary nodule.

Segmental or lobar atelectasis

• Atelectasis due to bronchial obstruction is a common presentation of lung cancer.

• Despite the presence of atelectasis, volume loss is variable, secondary to the volume displacing effects of desquamated cells, mucus, and fluid.

• Obstructive pneumonia is a common presentation of lung cancer, caused by bronchial obstruction and parenchymal consolidation by inflammatory cells and lipid-laden

macrophages.

• If two foci of atelectasis are present simultaneously that cannot be explained by a single endobronchial lesion, a benign process is much more likely.

However, CT and bronchoscopy are still needed for workup.

Consolidation

• Consolidation that is indistinguishable from pneumonia can be seen with BAC (mucinous subtype). Although BAC and pneumonia may appear similar, BAC is usually a non-resolving consolidation with (near) normal white blood cell count.

Hilar mass

• A hilar mass is a common presentation of squamous cell and small cell carcinoma.

• Hilar enlargement may be due to a primary central tumor or nodal metastasis from a parenchymal neoplasm.

• Tumor may compress and narrow the bronchus. A tapered bronchus is highly specific for lung cancer.

Superior sulcus tumor

• A superior sulcus tumor is a lung cancer occurring in the lung apex.

• A Pancoast tumor is a type of superior sulcus tumor with involvement of the sympathetic ganglia causing a Horner syndrome, with ipsilateral ptosis, miosis, and anhidrosis.

• A superior sulcus tumor is a stage T3 tumor. The staging of lung cancer is subsequently discussed.

Lymphangitic carcinomatosis

Lymphangitic carcinomatosis represents diffuse spread of neoplasm through the pulmonary lymphatics, typically seen in late-stage disease.

• On imaging, carcinomatosis manifests as nodular interlobular septal thickening, which is usually asymmetric.

Pleural effusion

• Pleural effusions are relatively common in lung cancer, and may be due to lymphatic obstruction or pleural metastases.

• A *malignant* effusion is the presence of malignant cells within the effusion.

A malignant effusion is an M1a lesion, which precludes curative resection.

Not all effusions associated with lung cancer are malignant effusions, so cytologic evaluation is necessary.

Pneumothorax

• Pneumothorax is not a common presentation of lung cancer, but can be seen with peripheral tumors that cavitate or invade into the pleura.

Staging of lung cancer

Overview of lung cancer staging

• The 7th edition of the TNM system, published in 2009, is based on data collected on over 100,000 patients with lung cancer from 1990 to 2000.

• Staging of small cell lung cancer was previously simplified to “limited-stage” or “extensive-stage.”

The new 7th TNM system, however, recommends that the same

TNM staging system be used for both small cell and non-small cell lung cancer.

Stage T N M

IA

IB

IIA

IIB

IIIA

IIIB

IV

Up to T1b

T2a

Up to T2a (or T2b if N0)

T2b (or T3 if N0)

Up to T3 (or T4 if N0–1)

Any T (or T4 if N2)

Any T

N0

N0

N1

N1

N2

N3

Any N

M0

M0

M0

M0

M0

M0

M1a or **M1b**

Treatment based on staging

• For early stages up to IIB and sometimes IIIA, surgery is usually performed.

Neoadjuvant or adjuvant chemotherapy and radiotherapy can be used.

• Stage IIIB (N3 - contralateral or supraclavicular nodes; or T4/N2) is unresectable.

• Stage IV disease is generally not treated surgically unless there is a solitary adrenal or brain metastasis.

T (tumor)

• T1: Tumor ≤3 cm surrounded by lung or visceral pleura.

T1a: Tumor ≤2 cm; 5-year survival rate 77%; T1b: >2 and ≤3 cm; 5-year survival rate 71%.

• T2: Tumor >3 cm and ≤7 cm, or local invasion of the visceral pleura, or endobronchial

lesions >2 cm from the carina.

T2a: > 3 and ≤5 cm; 5-year survival rate 58%; T2b: >5 and ≤7 cm; 5-year survival rate 49%.

• T3: Tumor >7 cm, or local invasion of chest wall, diaphragm, pleura, or superior sulcus tumor, or endobronchial lesion <2 cm from carina.

T3: 5-year survival rate 35%.

T3: Separate tumor nodule in the same lobe; 5-year survival rate 28%.

• T4: Separate tumor nodule in a different lobe in the ipsilateral lung, or tumor of any size with invasion of mediastinal structures including carina, heart, great vessels, or vertebral bodies.

T4: 5-year survival rate 22%.

N (nodes)

• The color coding of the lymph nodes on the diagram below represents the nodal staging of lung cancer for an example left-sided mass:

N1: ipsilateral hilar nodes and intrapulmonary nodes

10 - hilar

11 - interlobar (adjacent to interlobar bronchi)

12 - lobar (adjacent to lobar bronchi)

13 - segmental

14 - subsegmental

N3: 1 - supraclavicular nodes (any side)

any contralateral node

N2: ipsilateral mediastinal nodes

2 - upper paratracheal

3 - prevascular (anterior to aorta, not shown)

4 - paratracheal

5 - subaortic (AP window)

6 - paraaortic

7 - subcarinal

8 - paraesophageal

9 - pulmonary ligament

• the boundary between right and left for level 2 and 4 lymph nodes is set as the left lateral border of the trachea due to _ow of lymphatic drainage

• in this example of a left-sided primary, the right-sided nodes are drawn in orange to demonstrate N3 due to contralaterality

AP window (between aorta and

pulmonary artery)

• N0: No lymph node metastases.

• N1: Ipsilateral hilar or intrapulmonary lymph nodes.

• N2: Ipsilateral mediastinal nodes.

• N3: Contralateral lymph nodes or supraclavicular nodes on either side.

M (metastasis)

• M0: No metastatic disease.

• M1a: Local thoracic metastatic disease.

Separate tumor nodule in contralateral lung; 5-year survival rate 3%.

Malignant pleural or pericardial effusion; 5-year survival rate 2%.

• M1b: Distant or extrathoracic metastatic disease.

Median survival 6 months. *1-year* survival rate 22%.

Pulmonary vascular disease

Pulmonary hypertension

Definition of pulmonary hypertension

• Clinically, the term *pulmonary hypertension* is used to encompass both pulmonary arterial and pulmonary venous hypertension.

The term *pulmonary arterial hypertension* (PAH) is generally reserved for the WHO class 1 entities, discussed below.

• It may be clinically difficult to distinguish pulmonary arterial from pulmonary venous hypertension.

Further complicating the distinction, venous hypertension may be a cause of arterial hypertension.

• Pulmonary arterial hypertension is defined as pulmonary arterial systolic pressure ≥25 mm Hg at rest or ≥30 mm Hg during exercise.

• Elevated pulmonary venous pressures are present when pulmonary capillary wedge pressure (an approximation of pulmonary venous pressure) is ≥18 mm Hg.

Overview of pulmonary hypertension classification

• There are a number of causes of pulmonary hypertension including chronic thromboembolic disease, chronic respiratory disease, chronic heart disease, and

idiopathic causes.

• The classification in widest use in the radiology literature is the hemodynamic division of precapillary and postcapillary etiologies.

In **precapillary causes** of pulmonary hypertension, the primary abnormality is either the pulmonary arterial system or pulmonary arterial blood flow.

Abnormalities of the pulmonary parenchyma leading to chronic alveolar hypoxia are also included in this category.

In **postcapillary causes** of pulmonary hypertension, an abnormality of the pulmonary veins or elevation of pulmonary venous pressure leads to pulmonary arterial hypertension.

• In contrast, the World Health Organization (WHO) clinical classification, based on the 2003 World Symposium on Pulmonary Hypertension, describes five groups of pulmonary hypertension based on etiology.

There is no correlation between the pre/postcapillary classification and the WHO classification, and in fact there can be both pre- and postcapillary etiologies within a single WHO group.

Group 1: **Pulmonary arterial hypertension** (PAH).

Primary pulmonary hypertension (PPH) may be idiopathic or familial.

Congenital left-to-right shunts, such as atrial septal defect (ASD) and ventricular septal defect (VSD), may cause PAH and shunt reversal (Eisenmenger syndrome).

PAH may be caused by pulmonary venous or capillary involvement, such as pulmonary venoocclusive disease and pulmonary capillary hemangiomatosis.

Group 2: **Pulmonary venous hypertension**.

Left-sided heart disease (left atrial, left ventricular, or mitral/aortic valve disease) may cause elevated pulmonary venous pressure in chronic disease.

Group 3: **Pulmonary hypertension associated with chronic hypoxemia**.

COPD, interstitial lung disease, and sleep apnea can cause pulmonary hypertension in chronic

disease.

Group 4: **Pulmonary hypertension due to chronic thromboembolic disease**.

Group 5: **Pulmonary hypertension due to miscellaneous disorders**.

Sarcoidosis is a rare cause of pulmonary hypertension.

Compression of pulmonary vessels, which can be due to neoplasm, fibrosing mediastinitis, etc., may cause pulmonary hypertension.

General imaging of pulmonary hypertension

• A main pulmonary artery diameter ≥2.9 cm suggests the presence of pulmonary hypertension, although pulmonary hypertension may be present in a normal caliber pulmonary artery.

A main pulmonary artery diameter larger than the aortic root

diameter is also suggestive of pulmonary hypertension.

• Pulmonary artery calcifications are pathognomonic for chronic pulmonary artery hypertension.

• Pulmonary hypertension may cause mosaic attenuation due to perfusion abnormalities, most commonly seen in chronic thromboembolic pulmonary hypertension (CTEPH).

• Pulmonary hypertension may be associated with ground glass centrilobular nodules, especially in pulmonary veno-occlusive disease.

• An enlarged pulmonary artery can mimic a mediastinal mass. The *hilum convergence* sign is helpful to confirm that the apparent “mass” in fact represents the pulmonary artery.

The *hilum convergence* sign describes the appearance of hilar pulmonary artery branches converging into an enlarged pulmonary artery.

• In contrast, the *hilum overlay* sign describes the visualization of hilar vessels through a mass.

It indicates that a mediastinal mass is present, which cannot be in the middle mediastinum. Usually this means the mass is in the anterior mediastinum.

Primary pulmonary hypertension (PPH) – WHO group 1, precapillary

• The pathologic hallmark of primary pulmonary hypertension (PPH) is the *plexiform lesion* in the wall of the muscular arteries, which is a focal disruption of the elastic lamina by an obstructing plexus of endothelial channels. There is a relative

paucity of prostacyclins and nitric oxide expressed by endothelial cells.

• PPH may be idiopathic (females > males) or familial (approximately 10% of cases).

• On imaging, there is typically enlargement of the main pulmonary arteries with rapidly tapering peripheral vessels.

Pulmonary hypertension due to left-to-right cardiac shunts – WHO group 1, precapillary

• Congenital left-to-right cardiac shunts, such as ventricular septal defect (VSD), atrial septal defect (ASD), and partial anomalous pulmonary venous return, cause increased

flow through the pulmonary arterial bed. This chronically increased flow may eventually lead to irreversible vasculopathy characterized by pulmonary hypertension and reversal of the congenital shunt, known as Eisenmenger syndrome.

• Imaging of PAH secondary to a congenital shunt is similar to that of PPH.

There is enlargement of the central and main pulmonary arteries, with peripheral tapering.

Pulmonary veno-occlusive disease – WHO group 1, postcapillary

• PAH secondary to pulmonary veno-occlusive disease is caused by fibrotic obliteration of the pulmonary veins and venules. Pulmonary veno-occlusive disease may be idiopathic

but is associated with pregnancy, drugs (especially bleomycin), and bone marrow

transplant.

• Imaging features pulmonary arterial enlargement. Pulmonary edema and ground glass centrilobular nodules are often present.

Pulmonary venous hypertension – WHO group 2, postcapillary

• Left-sided cardiovascular disease leads to elevated pulmonary venous pressure, which is a cause of pulmonary hypertension.

• Any left-sided lesion may cause pulmonary venous hypertension, including left ventricular outflow tract lesions, mitral stenosis, and obstructing intra-atrial tumor/

thrombus.

Pulmonary hypertension associated with hypoxemic lung disease – WHO group 3, precapillary

• COPD, sleep apnea, and interstitial lung disease can all lead to pulmonary hypertension.

• Chronic hypoxic vasoconstriction is thought to invoke vascular remodeling leading to hypertrophy of pulmonary arterial vascular smooth muscle and intimal thickening.

• Chronic lung disease can further contribute to obliteration of pulmonary microvasculature through emphysema and the perivascular fibrotic changes of pulmonary fibrosis.

Chronic thromboembolic pulmonary hypertension (CTEPH) – WHO group 4, precapillary

• Chronic occlusion of the pulmonary arterial bed can lead to pulmonary arterial hypertension, which is a complication affecting 1–5% of patients who develop acute pulmonary embolism (PE).

Due to the high prevalence of PE, a PE-protocol CT is typically the first step in workup of newly diagnosed pulmonary hypertension.

• Characteristic imaging features are peripheral, eccentric filling defects (in contrast to be central) in the pulmonary arterial tree. Fibrous strands are sometimes visible on cross-sectional imaging.

Mosaic perfusion may be present.

• CTEPH may cause secondary corkscrew bronchial arteries that are tortuous and dilated.

• Treatment of CTEPH is surgical thromboendarterectomy (similar to carotid endarterectomy).

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• Treatment of CTEPH is surgical thromboendarterectomy (similar to carotid endarterectomy).

Fibrosing mediastinitis – WHO group 5, postcapillary

• Progressive proliferation of fibrous tissue within the mediastinum may lead to encasement and compression of mediastinal structures. The most common causes of

fibrosing mediastinitis are histoplasmosis and tuberculosis.

• Fibrous encasement of the pulmonary veins leads to permanent histological changes within the endothelial cells.

• Fibrosing mediastinitis may also encase the pulmonary arteries, creating a precapillary pulmonary hypertension.

• Imaging features of fibrosing mediastinitis include increased mediastinal soft tissue, often with calcified lymph nodes due to prior granulomatous infection.

Pulmonary embolism (PE)

Clinical diagnosis

• Diagnosis of pulmonary embolism (PE) can be challenging because the presenting symptoms are both common and nonspecific, including dyspnea, tachycardia, and

pleuritic chest pain.

• Most pulmonary emboli originate in the deep veins of the thighs and pelvis.

The risk factors for deep venous thrombosis are widely prevalent in a hospital environment, including:

Immobilization, malignancy, catheter use, obesity, oral contraceptive use, and thrombophilia.

Approximately 25% of patients with PE don’t have any identifiable risk factor.

• The Wells score assigns point values to clinical suspicion and various symptoms suggestive of pulmonary embolism.

• D-dimer is sensitive for thromboembolic disease and has a high negative predictive

value, but is of little value in the typical inpatient population as there are many false positives.

Imaging of pulmonary embolism

• CT pulmonary angiogram is the most common method to image for PE, where an embolism is typically seen as a central intraluminal pulmonary artery filling defect.

Pulmonary emboli tend to lodge at vessel bifurcations.

• An eccentric, circumferential filling defect suggests chronic thromboembolic disease.

• Associated pulmonary abnormalities are commonly seen in patients with PE, including wedge-shaped consolidation, pleural effusion, and linear bands of subsegmental atelectasis.

These findings, however, are nonspecific. In particular, pleural effusions and consolidation are seen approximately equally in patients with or without pulmonary embolism.

Plain film evaluation of pulmonary embolism

• While a CT pulmonary angiogram is the standard tool to evaluation for pulmonary embolism, it is important to be aware

of plain film findings that could suggest pulmonary embolism in case the diagnosis is not clinically suspected.

• The *Fleischner* sign describes widening of the pulmonary arteries due to clot.

• *Hampton's hump* is a peripheral wedge-shaped opacity representing pulmonary

infarct.

• *Westermark* sign is regional oligemia in the lung distal to the pulmonary artery thrombus.

Cardiac evaluation

• Pulmonary embolism may cause acute right heart strain. After evaluation of the pulmonary arterial tree, one should always examine the heart for imaging findings of right heart dysfunction.

• Massive PE may cause acute right ventricular dilation with bowing of the intraventricular septum to the left. An elevated RV:LV ratio (caused by RV enlargement) is linearly correlated with increased mortality.

Pitfalls of CT pulmonary angiogram

• Hilar lymph nodes may simulate large PE.

• Cardiac motion causes blurring of the left lower lobe pulmonary arteries, which may simulate small peripheral emboli.

• Respiratory motion overall decreases accuracy in evaluation of small pulmonary arteries.

• Mucus-impacted bronchi may simulate PE.

• Transient disruption of contrast bolus occurs when unopacified blood from the IVC enters the right atrium and is pumped into the lungs.

• Unopacified pulmonary veins may simulate PE on a single CT slice; however, one may distinguish between a pulmonary artery and vein by tracing the vessel back to the heart.

Diffuse lung disease

Idiopathic interstitial pneumonias

Overview of idiopathic interstitial pneumonias

• The lung has a limited repertoire of responses to injury. Common responses to injury range from recruiting lymphocytes or macrophages, increasing inflammatory debris, and instigating a fibrotic reaction.

• The idiopathic interstitial pneumonias (IIPs) are seven different patterns of injury response.

These patterns can be associated with collagen vascular disease, drug reaction, occupational exposure, or they may be idiopathic.

• A disease can only be classified as an IIP if there is no other explanation for the pathologic changes.

• Each of the seven entities has both a clinical syndrome and a separate pathologic diagnosis. Sometimes the clinical syndrome and the pathologic terms have different names and sometimes they share the same name, but in every case there is an

acronym!

• In order to understand the histopathological responses to injury, it is important to

review the normal alveolar anatomy.

Idiopathic pulmonary fibrosis (IPF)

• Idiopathic pulmonary fibrosis (IPF) is the most common idiopathic interstitial pneumonia and has the second-worse prognosis of all, with a mean survival of 2–4 years. The mean

survival of IPF is not much different compared to lung cancer. Of all the interstitial pneumonias, only acute interstitial pneumonitis (AIP) has a worse prognosis.

• Clinical symptoms of IPF include dry cough and dyspnea.

• IPF usually affects patients >50 years old.

• The pathologic diagnosis corresponding to the clinical syndrome of IPF is usual interstitial pneumonia (UIP), which features interstitial fibroblastic foci and chronic alveolar inflammation.

• IPF is the clinical syndrome of UIP with unknown cause and is the most common cause of UIP.

IPF has a much worse clinical outcome compared to secondary causes of UIP.

• Other triggers of lung injury that may result in a UIP pattern include:

Collagen vascular disease (rheumatoid arthritis much more commonly than scleroderma).

Drug injury.

Asbestosis. An imaging clue to the presence of asbestosis is calcified plaques indicative of prior asbestos exposure.

• On imaging, early UIP features irregular reticulation in the posterior subpleural lung

bases.

• In later stages of UIP, reticulation becomes fibrosis, traction bronchiectasis develops, and posterior subpleural honeycombing becomes prominent.

The lung bases are most severely affected. The **CT** diagnosis of late UIP is very specific based on these findings, in particular the presence and posterior basal location of honeycombing. Surgical biopsy can often be avoided if these characteristic imaging features are present.

Nonspecific interstitial pneumonitis (NSIP)

• Nonspecific interstitial pneumonitis (NSIP) is both the name of the clinical syndrome and the corresponding pathologic diagnosis.

Affected patients are typically younger (40–50 years old) compared to IPF.

Symptoms are similar to IPF with chronic dry cough

and dyspnea.

• NSIP has a better 5-year survival compared to IPF. Unlike IPF, NSIP does respond to steroids.

• NSIP may be idiopathic or associated with other diseases. NSIP is the most common pulmonary manifestation in patients with collagen vascular disease.

NSIP may also be caused by drug reaction or occupational exposure.

NSIP may be associated with dermatomyositis.

• There are two forms of NSIP. An important imaging feature of NSIP, regardless of the form, is the presence of ground glass opacities (GGO), which are nearly always bilateral.

A key feature differentiating NSIP from IPF is the presence of ground glass in NSIP.

• **Fibrotic NSIP** predominantly features GGO with fine reticulation and traction bronchiectasis.

Honeycombing is usually absent in NSIP. If honeycombing is present, consider UIP.

Fibrotic NSIP has a worse prognosis compared to cellular NSIP, but a better prognosis compared to UIP.

• **Cellular NSIP** also features GGO, but without significant fibrotic changes.

Cellular NSIP is much less common than fibrotic NSIP and has a better prognosis compared to fibrotic NSIP.

• A key imaging finding (not always seen but very specific) is sparing of immediate subpleural lung.

This feature is NOT seen in UIP, and can be seen in both cellular and fibrotic NSIP.

• Like UIP, NSIP tends to affect the posterior peripheral lower lobes. Other diagnoses

should be considered (e.g., chronic hypersensitivity pneumonitis or sarcoidosis) if there is primarily upper lobe disease.

Cryptogenic organizing pneumonia (COP)

• Cryptogenic organizing pneumonia (COP) is the clinical syndrome of organizing pneumonia (OP) without known cause. The clinical syndrome of COP was previously called bronchiolitis obliterans organizing pneumonia (BOOP), which is a term still in general use.

• COP clinically responds to steroids with a good prognosis and may resolve completely, although recurrences are common.

• Organizing pneumonia (OP) is the pathologic pattern of granulation tissue polyps that fill the distal airways and alveoli. Organizing pneumonia may be a response to infection, drug reaction, or inhalation.

CT of OP shows mixed consolidation and ground glass opacities in a peripheral and peribronchovascular distribution.

The *reverse halo* sign (also known as the *atoll* sign) is

relatively specific for OP and features a central lucency surrounded by a ground glass halo.

The reverse halo sign should not be confused with the halo sign that is typical of invasive aspergillus, which shows a central opacity with peripheral ground glass.

Respiratory bronchiolitis–interstitial lung disease (RB-ILD)

• Respiratory bronchiolitis–interstitial lung disease (RB-ILD) is both the clinical syndrome and the pathologic diagnosis of this smoking-related interstitial lung disease.

• Respiratory bronchiolitis (RB, without the ILD) is very common in smokers, where pigmented macrophages are found in respiratory bronchioles.

RB is usually asymptomatic, but if symptoms are present (usually cough and shortness of breath),the clinical syndrome is called RB-ILD.

• Histologically, RB-ILD is characterized by sheets of macrophages filling the terminal airways, with relative sparing of the alveoli.

• **The key imaging features** of RB-ILD are centrilobular nodules and patchy ground glass opacities.

In contrast to NSIP, the distribution of ground glass in RB-ILD is more random than the peripherally predominant pattern of NSIP.

Desquamative interstitial pneumonia (DIP)

• Like RB-ILD, desquamative interstitial pneumonia (DIP) is both the clinical syndrome and the pathologic diagnosis. RB, RB-ILD, and DIP represent a continuous spectrum of smoking-related lung disease.

• Like RB, brown-pigmented macrophages are involved in DIP; however, sheets of these abnormal macrophages also extend into the alveoli in DIP.

• Imaging of DIP shows diffuse basal-predominant patchy or subpleural ground glass opacification, more extensive than RB-ILD.

Although the predominant abnormality is ground glass, a few cysts may also be present.

Lymphoid interstitial pneumonia (LIP)

• Lymphoid interstitial pneumonia (LIP) is both a clinical syndrome and the pathologic diagnosis.

LIP is exceptionally rare as an isolated idiopathic disease and is more commonly associated with Sjögren syndrome or HIV.

• The histologic hallmark of LIP is diffuse infiltration of the interstitium by lymphocytes and other immune cells, with resultant distortion of the alveoli.

• **Imaging findings** of LIP include diffuse or lower-lobe predominant ground glass.

Scattered thin-walled perivascular cysts are often present, which are thought to be due to air trapping from peribronchiolar

cellular debris. LIP may be complicated by pneumothorax in advanced disease.

Acute interstitial pneumonia (AIP)

• Acute interstitial pneumonia (AIP), synonymous with diffuse alveolar damage (DAD), is the pathologic diagnosis seen in the clinical syndrome of acute respiratory distress syndrome (ARDS). Unlike the other IIPs, AIP is the only syndrome with an acute onset and has the worst prognosis.

• The primary cause of AIP is surfactant destruction.

• Two phases of AIP are recognized: Early (exudative) and chronic (organizing).

• **The early (exudative) phase** features hyaline membranes, diffuse alveolar infiltration by immune cells, and noncardiogenic pulmonary edema.

• The **chronic (organizing) phase** features alveolar wall thickening due to granulation tissue.

The chronic phase usually begins one week after the initial injury.

Antigen and exposure-related lung disease

• Most inhalational lung diseases predominantly affect the upper lobes because the lower lobes have more robust blood flow and lymphatic drainage.

Hypersensitivity pneumonitis (HSP)

• Hypersensitivity pneumonitis (HSP) is a common lung disease caused by a The abnormalities of subacute HSP involve the entire axial cross-section of lung.

The *head-cheese* sign describes the combination of patchy ground glass and areas of lucency due to mosaic perfusion or air trapping.

• **Chronic** HSP, from long-term exposure to the offending antigen, leads to upper-lobe predominant pulmonary fibrosis. Often the findings of subacute disease, including centrilobular nodules, ground glass, and mosaic attenuation, may be superimposed.

Unlike IPF, honeycombing is not common in HSP, but when present may involve the upper lobes. If there is relative sparing of the bases, chronic HSP is much more likely than IPF.

Pneumoconioses

• A pneumoconiosis is a lung disease secondary to *inorganic* dust inhalation. In contrast, hypersensitivity pneumonitis is caused by *organic* dust inhalation.

• **Silicosis** and **coal workers pneumoconiosis (CWP)** are the two most common pneumoconioses.

They often have indistinguishable imaging findings even though

they are due to different inhaled dusts and have different histologic findings.

Silicosis is due to inhalation of silica dust, which miners may be exposed to.

CWP is caused by inhalation of coal dust, which does not contain any silica.

The most characteristic finding of *uncomplicated* disease is multiple upper lobe predominant centrilobular and subpleural nodules.

Eggshell lymph node calcifications are commonly seen in silicosis, less commonly in CWP.

Silicosis or CWP can become *complicated* with large conglomerate masses or progressive massive fibrosis.

Both silicosis and CWP confer an increased risk of TB.

Caplan syndrome is seen in patients with rheumatoid arthritis *and* either CWP or silicosis (more common in CWP) and represents necrobiotic rheumatoid nodules superimposed on the smaller centrilobular and subpleural nodules of the pneumoconiosis.

• **Asbestosis** is lung disease caused by inhalation of asbestos fibers. End-stage asbestosis can lead to pulmonary fibrosis with a UIP pathology.

Unlike the other inhalational lung diseases, asbestosis predominantly affects the lower lobes because the asbestos

particles are too large to be removed by the alveolar macrophages and lymphatic system.

The radiographic/CT appearance and distribution of advanced asbestosis may be indistinguishable from IPF; however, an important clue seen in asbestosis is evidence of asbestos exposure, such as pleural thickening and plaques.

Even though pleural plaques (which may or may not be calcified) are due to asbestos exposure, they are not a component of asbestosis, do not lead to fibrosis, and are usually asymptomatic.

Hypersensitivity reaction to inhaled organic antigens, such as bird proteins or thermophilic actinomycetes, although a history of antigen exposure is not always elicited.

• HSP is characterized by three distinct phases, from acute to subacute to chronic.

• **Acute** HSP is characterized by inflammatory exudate filling the alveoli, which manifests **on imaging** as nonspecific groundglass or consolidation. Small, ill-defined centrilobular

nodules may also be present.

• The imaging hallmark of **subacute** HSP is centrilobular ground glass nodules. Mosaic attenuation (geographic areas of relative lucency) and ground glass can also be seen.

Mosaic attenuation can be secondary to mosaic perfusion on inspiration and air trapping on expiration.

Eosinophilic lung disease

• Eosinophilic lung disease is a spectrum of diseases that feature accumulation of eosinophils in the pulmonary airspaces and interstitium.

Simple pulmonary eosinophilia (Löffler syndrome)

• Simple pulmonary eosinophilia (also known as Löffler syndrome) is characterized by transient and *migratory* areas of focal consolidation, with an elevated eosinophil count in the peripheral smear.

• An identical appearance can be seen as a response to injury, especially with parasitic disease and drug reactions.

The term *simple pulmonary eosinophilia* is reserved for idiopathic cases.

Chronic eosinophilic pneumonia

• Chronic eosinophilic pneumonia is an important consideration in the differential diagnosis of chronic consolidation.

Chronic eosinophilic pneumonia causes extensive alveolar filling and interstitial infiltration with inflammatory eosinophils.

• Consolidation is patchy and *peripheral*, with an *upper lobe* predominance. Unlike simple pulmonary eosinophilia, the pattern of consolidation can remain unchanged for months.

• Chronic eosinophilic pneumonia responds rapidly to steroids.

Pulmonary vasculitis

Churg–Strauss

• Also called allergic angiitis and granulomatosis, Churg–Strauss is a systemic small vessel vasculitis associated with asthma and peripheral eosinophilia.

• P-ANCA is positive, which is not very specific. P-ANCA can also be positive in collagen vascular disease and microscopic polyangiitis (discussed below).

• Imaging findings of Churg–Strauss are varied. The most common appearance is peripheral consolidation or ground glass.

Microscopic polyangiitis

• Microscopic polyangiitis is the most common cause of pulmonary hemorrhage with renal failure.

P-ANCA is positive.

• Imaging shows diffuse central-predominant ground glass representing hemorrhage.

Wegener granulomatosis (WG)

• Wegener granulomatosis (WG) is a systemic small-vessel vasculitis with a classic clinical triad of sinusitis, lung involvement, and renal insufficiency.

• C-ANCA is positive, which is very specific for Wegener granulomatosis.

• In the upper airways, WG may cause nasopharyngeal and eustachian tube obstruction.

Involvement of the larynx and bronchi is common, leading to airway stenosis.

• In the lungs, WG may cause **multiple cavitary nodules** that don’t respond to antibiotic therapy.

An intra-cavitary fluid level suggests superimposed infection.

Iatrogenic lung disease

Drug toxicity

• The lung has a diverse but finite repertoire of responses to injury, including pulmonary edema (due to increased capillary permeability), ARDS, organizing pneumonia, eosinophilic pneumonia, bronchiolitis obliterans, pulmonary hemorrhage, NSIP, and UIP.

• Pulmonary drug reaction, most commonly to cytotoxic drugs, may elicit any of these injury responses.

Radiation lung injury

• Up to 40% of patients develop radiographic abnormalities after external radiotherapy, although most patients are asymptomatic. The radiographic abnormality is largely

confined to the radiation port, usually with non-anatomic linear margins.

• Radiation pneumonitis is the early stage of radiation injury, which can occur within 1 month of radiotherapy and is most severe 3–4 months after treatment.

Radiation pneumonitis features ground glass centered on the radiation port, although extension out of the port is relatively common.

• Radiation fibrosis is the late stage of radiation injury. Fibrosis becomes apparent approximately 6–12 months after therapy. The key imaging finding is the distribution of fibrosis and traction bronchiectasis *within the radiation port*, although fibrosis may extend outside the port in 20%.

Idiopathic systemic diseases affecting the lungs

Sarcoidosis

• Sarcoidosis is an idiopathic systemic disorder of noncaseating granulomas that become coalescent to form nodules and masses throughout the body.

• Pulmonary sarcoidosis may progress to pulmonary fibrosis with honeycombing.

Unlike IPF (the most common cause of pulmonary fibrosis), the fibrotic changes of sarcoid have a mid and upper-lung predominance, similar to end-stage hypersensitivity pneumonitis.

• A historical staging system has been used for radiographic findings (not CT) in sarcoidosis; however, there is not always stepwise progression through the stages.

Stage 0: Normal radiograph.

Stage 1: Hilar or mediastinal adenopathy only, *without* lung changes.

Stage 2: Adenopathy *with* lung changes.

Stage 3: Diffuse lung disease *without* adenopathy.

Stage 4: End-stage fibrosis.

• The **most common radiographic finding** in sarcoidosis is *symmetric adenopathy.*

Lymph nodes may contain stippled or eggshell calcification in up to 50%.

• **The most common CT finding** in sarcoidosis, in addition to adenopathy, is upper lobe predominant *perilymphatic* nodules of variable sizes, representing sarcoid granulomas.

• Bronchial involvement may cause mosaic perfusion due to air trapping.

• Sarcoidosis may involve other organs, including the spleen, brain, and rarely bone.

Pulmonary Langerhans cell histiocytosis (PLCH)

• Pulmonary Langerhans cell histiocytosis (PLCH) is a smoking-related lung disease –nearly 100% of adults with PLCH are smokers.

The other smoking-related interstitial lung diseases (aside from emphysema) are RB-ILD and DIP.

Multiple smoking-related lung diseases may be present simultaneously.

• PLCH may present as a spontaneous pneumothorax.

• Disease is most often isolated to the lungs; however, lucent bone lesions, diabetes insipidus from inflammation of the pituitary stalk (hypophysitis), and skin involvement can occasionally be seen.

In addition to LCH, the differential diagnosis for diseases affecting the lungs and bones includes **malignancy**, **tuberculosis**, **fungal disease** (including blastomycosis, histoplasmosis, and coccidiomycosis), **sarcoidosis**, and **Gaucher disease** (pulmonary involvement is rare and may resemble DIP).

• The first detectable abnormality is nodules associated with airways. As the disease progresses, the nodules cavitate and resultant irregular cysts predominate.

• The natural progression is from nodules ☐ cavitary nodules ☐ irregular cysts.

• Radiographic and CT findings include upper lobe predominant cysts and irregular peribronchovascular nodules, both sparing the costophrenic sulci.

• PLCH is generally steroid responsive and smoking cessation is critical.

Miscellaneous diffuse pulmonary disease

Pulmonary alveolar proteinosis (PAP)

• Pulmonary alveolar proteinosis (PAP) is an idiopathic disease causing filling of the alveoli with a proteinaceous lipid-rich material.

• On **chest radiography**, PAP may resemble pulmonary edema with perihilar opacification; however, the heart is normal in size in PAP and pleural effusions are not typically seen.

• The **CT** hallmark of PAP is the *crazy paving* pattern of smooth interlobular septal thickening in areas of patchy or geometric ground glass.

Although initially described for PAP, the *crazy paving* pattern is not specific for PAP, and can also be seen in *Pneumocystis* pneumonia, cryptogenic organizing pneumonia, bronchoalveolar carcinoma, and lipoid pneumonia, among others.

• Patients with PCP are susceptible to superimposed infection, classically with *Nocardia*,

which typically presents as a consolidation.

• Treatment of PAP is bronchoalveolar lavage.

Lymphangioleiomyomatosis (LAM)

• Lymphangioleiomyomatosis (LAM) is a diffuse cystic lung disease caused by bronchiolar obstruction and lung destruction due to proliferation of immature smooth muscle cells in small vessels, lymphatics, and bronchioles.

• Approximately 1% of patients with tuberous sclerosis (triad of seizures, mental retardation, and adenoma sebaceum) have a lung disease that is nearly identical to

LAM.

• Almost all cases of sporadic LAM are in women of childbearing age.

• Some cases respond to anti-estrogen therapy.

• LAM is associated with pneumothorax and chylous pleural effusion.

• CT of LAM shows numerous thin-walled lung cysts. In contrast to LCH, the cysts tend to be round and regular. Also, LAM affects all five lobes while LCH is upper-lobe

predominant.

Mediastinum

Anatomy of the mediastinum

• The mediastinum is divided into three arbitrary compartments to aid in the differential diagnosis of a mediastinal mass. However, there are no anatomic planes separating these divisions and disease can spread from one “compartment” to another.

Several methods have been proposed to divide the mediastinum, including the anatomical method, the Felson method, and others. This text follows the method proposed by Whitten et al. in Radiographics 2007.

• The three divisions are primarily used to aid in the differential diagnosis of a mass seen on radiography. For CT diagnosis of a mediastinal mass, it is often possible to be more precise and state exactly where a lesion is arising from.

Anterior mediastinum

• The anterior mediastinum is the space between the sternum and the pericardium inferiorly and ascending aorta and brachiocephalic vessels superiorly.

• The anterior mediastinum can be thought of as two compartments – the prevascular compartment superiorly and the precardiac compartment inferiorly.

• The contents of the **prevascular anterior mediastinum** include:

Thymus.

Lymph nodes.

Enlarged thyroid gland, if it extends inferiorly into the mediastinum.

• The **precardiac anterior mediastinum** is a potential space.

Middle mediastinum

• The anterior border of the middle mediastinum is the anterior pericardium and the posterior borders are the posterior pericardium and posterior tracheal wall.

• The contents of the **middle mediastinum** include:

Heart and pericardium. Some authors place the heart in the anterior mediastinum.

However, for the purpose of differential diagnosis, diseases of the heart and pericardium have more in common with the other vascular structures of the middle mediastinum.

Ascending aorta and aortic arch.

Great vessels including SVC, IVC, pulmonary arteries and veins, and brachiocephalic vessels.

Trachea and bronchi.

Lymph nodes.

Phrenic, vagus, and recurrent laryngeal nerves (all of which pass through the AP window).

Posterior mediastinum

• The anterior border of the posterior mediastinum is the posterior trachea and posterior pericardium. The posterior border is somewhat loosely defined as the anterior aspect of the vertebral bodies; however, paraspinal masses are generally

included in the differential of a posterior mediastinal mass.

• The contents of the **posterior mediastinum** include:

Esophagus.

Descending thoracic aorta.

Azygos and hemiazygos veins.

Thoracic duct.

Vagus nerves.

Lymph nodes.

Lines, stripes, and interfaces

• Interfaces between anatomic structures in the lungs, mediastinum, and pleura may be displaced or thickened in the presence of a mediastinal mass or abnormality.

With the exception of the right paratracheal stripe, it is generally uncommon to see these interfaces in the absence of pathology.

A “line” is a thin interface formed by tissue (typically <1 mm in thickness) with air on both sides.

A “stripe” is a thicker interface formed between air and adjacent soft tissue.

An “interface” is formed by the contact of two soft tissue structures of different densities.

• The **anterior junction line** is formed by four layers of pleura (parietal and visceral pleura of each lung) at the anterior junction of the right and left lungs.

On a frontal radiograph, the anterior junction line is a vertical line projecting over the superior twothirds of the sternum.

Abnormal convexity or displacement of this line suggests an anterior mediastinal mass.

• The **posterior junction line** is also formed by four layers of pleura (parietal and visceral pleura of each lung), but at the posterior junction of the right and left lungs.

On a frontal radiograph, the posterior junction line is a vertical line projecting through the trachea on the frontal view, more superior than the anterior junction line. Unlike the anterior

junction line, the posterior junction line is seen above the clavicles because the posterior lungs extend more superiorly than the anterior lungs.

Abnormal convexity or displacement of this line suggests a posterior mediastinal mass or aortic aneurysm.

• The **right and left paratracheal stripes** are formed by two layers of pleura where the medial aspect of each lung abuts the lateral wall of the trachea and intervening mediastinal fat.

The right paratracheal stripe is the most commonly seen of these landmarks, seen in up to 97% of normal PA chest radiographs.

Thickening of the right paratracheal stripe is most commonly due to pleural thickening, although a paratracheal or tracheal mass (including adenopathy or thyroid or tracheal neoplasm) can also be a cause.

Thickening of the left paratracheal stripe has a similar differential. In addition, however, a mediastinal hematoma should also be considered, especially in trauma.

• The **posterior tracheal stripe** is the only interface seen on the *lateral radiograph*, representing the interface of the posterior wall of the trachea with the two pleural layers of the medial right lung.

The **right and left paraspinal lines** are actually interfaces but appear as lines due to Mach effect and are formed by 2 layers of pleura abutting the posterior mediastinum.

In contrast to the posterior junction line, the paraspinal lines are located inferiorly in the thorax, typically from the 8th through 12th ribs.

A paraspinal line abnormality suggests a posterior mediastinal mass lesion, including hematoma, neurogenic tumor, aortic aneurysm, extramedullary hematopoiesis, esophageal mass, and osteophyte.

• The **azygoesophageal recess** is an interface formed by the contact of the posteromedial right lower lobe and the retrocardiac mediastinum.

The azygoesophageal recess extends from the subcarinal region to the diaphragm inferiorly.

Distortion of the azygoesophageal recess may be due to esophageal mass, hiatal hernia, left atrial enlargement, and adenopathy.

Aortopulmonary (AP) window

• The aortopulmonary (AP) window is a mediastinal space nestled underneath the aortic arch (which forms the superior, anterior, and posterior boundaries) and the top of the pulmonary artery.

The medial border of the AP window is formed by the esophagus, trachea, and left mainstem bronchus.

• On a **normal frontal radiograph**, the AP window is a shallow concave contour below the aortic knob and above the pulmonary artery.

• Abnormal convexity (outwards bulging) of the AP window suggests a mass arising from or involving structures that normally live within the AP window, including:

Lymph nodes: Adenopathy is the most common cause of an AP window abnormality.

Left phrenic nerve: Injury may cause paralysis of the left hemidiaphragm.

Recurrent laryngeal nerve: The AP window should be carefully evaluated in new-onset

hoarseness, especially if associated with diaphragmatic paralysis.

Left vagus nerve.

Ligamentum arteriosum.

Left bronchial arteries.

• A thoracic aortic aneurysm may also cause convexity of the AP window.

Retrosternal clear space

• The retrosternal clear space is a normal area of lucency posterior to the sternum seen on the lateral radiograph only. It correlates to the prevascular space on CT.

• Obliteration of the retrosternal clear space suggests an anterior mediastinal mass, right ventricular dilation, or pulmonary artery enlargement.

• Increase in the retrosternal clear space can be seen in emphysema.

Left superior intercostal vein (LSIV)

• The left superior intercostal vein (LSIV) is a normal vein that is not often seen on radiography. When visible, the LSIV produces the *aortic nipple*, appearing as a small round shadow to the left of the aortic knob on the frontal radiograph.

• The LSIV may be dilated as a collateral pathway in SVC obstruction.

Radiographic localization of a mediastinal mass

Detection of an anterior mediastinal mass

• Deformation of the anterior junction line suggests an anterior mediastinal mass.

However, since the anterior junction line is not always seen, it is more common to infer the anterior location of a mass by preservation of the posterior lines in the presence of a mass.

• The *hilum overlay* sign is present on the frontal view if hilar vessels are visualized through the mass.

It indicates that the mass cannot be in the middle mediastinum.

The mass may be in the anterior (most likely) or posterior mediastinum.

Obliteration of the retrosternal clear space on the lateral radiograph is a direct sign of anterior mediastinal location.

Detection of a middle mediastinal mass

• Distortion of the paratracheal stripes or convexity of the AP window suggests a middle mediastinal mass.

Detection of a posterior mediastinal mass

• Distortion of the azygoesophageal recess, distortion of the posterior junction line, or displacement of the paraspinal lines suggest paravertebral/posterior mediastinal disease.

Anterior mediastinal mass - prevascular (superior anterior mediastinum)

Overview of prevascular anterior mediastinal masses

• A mnemonic for prevascular anterior mediastinal masses often learned in medical school is the *four T's*: Thymoma, teratoma, thyroid lesion, and terrible lymphoma. However, a more sophisticated approach is necessary when communicating with subspecialized clinicians.

• A better (but considerably less catchy) differential for an anterior mediastinal mass includes:

Thymic epithelial neoplasm, such as thymoma if the patient is middle aged or older, or has a

history of myasthenia gravis. Less common would be thymic carcinoma.

Germ cell tumor, including teratoma, if the patient is a young adult.

Thyroid lesion, if there is extension of the mass above the thoracic inlet.

Lymphoma.

Thymoma

• Thymoma is the most common primary tumor of the anterior mediastinum and typically occurs in middle-aged or older individuals, between 45 and 60 years.

• Thymoma is associated with myasthenia gravis (MG). Approximately 33% of patients with thymoma have MG, and 10% of patients with MG have a thymoma.

• In addition to MG, thymomas are often associated with other diseases including red cell aplasia, hypogammaglobulinemia, paraneoplastic syndromes, and malignancies such as lymphoma or thyroid cancer.

• Thymoma can be pathologically classified as low-risk or high-risk based on histology, and non-invasive or invasive based on whether the capsule is intact.

Approximately 30% of thymomas are invasive.

If invasive, the tumor may invade adjacent structures including the airways, chest wall, great vessels, and phrenic nerves.

Elevation of a hemidiaphragm is suggestive of phrenic nerve invasion.

Invasive thymoma may spread along pleural and pericardial surfaces, called *drop metastases*.

However, hematogenous metastases are exceedingly rare.

• Thymoma is histologically classified by the WHO system into A, AB, B1, B2, B3, and C subtypes, with progressively worse prognosis.

Type A tumors are relatively uncommon, but are usually encapsulated.

Type B tumors contain increasing number of epithelial cells, which represent the malignant component.

Type C represents thymic carcinoma, which may metastasize hematogenously.

Less common thymic lesions

• **Thymic carcinoma**

Unlike invasive thymoma, thymic carcinoma is histologically malignant, very aggressive, and often metastasizes hematogenously to lungs, liver, brain, and bone. Prognosis is poor.

Distinction between invasive thymoma and thymic carcinoma is difficult on CT, unless there is evidence of distant metastatic disease.

• **Thymic carcinoid**

Thymic carcinoid is of neural crest origin. 50% of thymic carcinoids are hormonally active, often secreting ACTH and causing Cushing syndrome.

Thymic carcinoid is associated with multiple endocrine neoplasia (MEN) I and II.

On imaging, thymic carcinoid is generally indistinguishable from thymoma and thymic carcinoma on CT.

If carcinoid is suspected, a preoperative Indium-111 Octreotide scan can be performed.

• **Thymic cyst**

A thymic cyst may be secondary to radiation therapy (e.g., administered to treat Hodgkin disease), may be associated with AIDS (especially when multilocular), or may be congenital. When congenital, thymic cysts arise from remnants of the thymopharyngeal duct.

A congenital thymic cyst may occur anywhere along the course of thymic descent from the neck, but most commonly in the anterior mediastinum.

Thymic cyst is typically evident on CT as a simple fluid-attenuation cyst in the anterior mediastinum.

• **Thymolipoma**

Thymolipoma is a benign fat-containing lesion with interspersed soft tissue.

It may become quite large and drape over the mediastinum.

Germ cell tumor (GCT)

• Several different types of germ cell tumors may arise in the anterior mediastinum from primitive germ cell elements, most of which are benign.

Malignant GCT occurs more commonly in males.

• **Teratoma** is the most common anterior mediastinal germ cell tumor, usually encapsulated and predominantly cystic in nature, but fat and calcification are common.

A fat/fluid level is specific for teratoma, but is not commonly seen.

Teratoma can rarely be malignant, especially if large in size and irregular in shape.

• **Seminoma** is the most common *malignant* anterior mediastinal germ cell tumor.

It occurs almost exclusively in men.

Thyroid lesion

• Benign and malignant thyroid masses may extend into the mediastinum, including goiter, thyroid neoplasm, and an enlarged gland due to thyroiditis.

• The key to diagnosis is to show continuity superiorly with the thyroid.

Lymphoma

• Both Hodgkin disease and non-Hodgkin lymphoma are important differential considerations for an anterior mediastinal mass.

• Hodgkin disease commonly involves the thorax, most often the superior mediastinal lymph nodes including prevascular, AP window, and paratracheal nodal stations.

• Non-Hodgkin lymphoma is a diverse group of diseases, which less commonly involve the thorax compared to Hodgkin disease.

• Calcification is rare in untreated lymphoma.

Non-lymphomatous adenopathy

• Lymph node calcification, low attenuation, and avid enhancement are unusual features for lymphoma.

An alternative diagnosis should be considered if these imaging findings are seen.

• **Eggshell calcification** of lymph nodes is often present in silicosis and coal workers pneumoconiosis, less commonly in sarcoidosis.

Given the greater prevalence of sarcoidosis, however, eggshell calcification may be seen more commonly in sarcoid overall.

• Dense calcification within a lymph node can be seen in sarcoidosis or as a sequela of prior granulomatous disease.

• Low attenuation lymph nodes, while nonspecific, should raise concern for active tuberculosis.

Low attenuation lymph nodes can also be seen in fungal infection, lymphoma, and metastatic disease.

• Avid lymph node enhancement can be seen in Castleman disease, sarcoidosis, tuberculosis, and vascular metastases. Avidly enhancing vascular metastases include:

Renal cell carcinoma.

Thyroid carcinoma.

Lung carcinoma.

Sarcoma.

Melanoma.

Castleman disease

• Castleman disease, also known as angiofollicular lymph node hyperplasia, is a cause of highly vascular thoracic lymph node enlargement, of uncertain etiology.

• Localized Castleman disease is seen in children or young adults. Surgical resection is usually curative.

• Multicentric Castleman disease manifests in older patients or in association with AIDS.

Multicentric disease often results in systemic illness including fever, anemia, and lymphoma. It is typically treated with chemotherapy.

• The key imaging finding of Castleman disease is avidly enhancing adenopathy.

Anterior mediastinal mass - precardiac (inferior anterior mediastinum)

Overview of precardiac anterior mediastinal masses

• A precardiac anterior mediastinal mass is in contact with the diaphragm.

Epicardial fat pad

• A prominent epicardial fat pad silhouettes the cardiac border on a frontal radiograph and may simulate cardiomegaly.

Pericardial cyst

• A pericardial cyst is a benign cystic lesion thought to be congenital.

Most are located at the right cardiophrenic angle (between the heart and the diaphragm).

• Imaging shows a cystic lesion abutting the pericardium, which may change in shape on subsequent studies.

Morgagni hernia

• Morgagni hernia is a diaphragmatic hernia through the foramen of Morgagni, containing omental fat and often bowel.

A Morgagni hernia usually occurs on the right.

• An anterior mediastinal mass in contact with the diaphragm containing bowel gas is diagnostic for a Morgagni hernia.

• If bowel gas is absent, a key to diagnosis on CT is the detection of omental vessels in the mass which can be traced into the upper abdomen.

Middle mediastinal mass

Lymphadenopathy

• Lymphadenopathy is an important cause of a middle mediastinal mass on radiography.

Ascending aortic or aortic arch aneurysm

• An abnormality of the ascending aorta or aortic arch may appear as a middle

mediastinal mass on radiography.

Enlarged pulmonary artery (PA)

• An enlarged pulmonary artery (PA) can simulate a mass on a chest radiograph. As previously discussed, the *hilum convergence* sign can help distinguish between an enlarged PA and a mediastinal mass.

The hilum convergence sign shows the peripheral

pulmonary arteries converging into the “mass” if the mass represents an enlarged pulmonary artery.

Foregut duplication cyst

• Foregut duplication cysts include bronchogenic cysts, esophageal duplication cysts, and neurenteric cysts. Foregut duplication cysts may occur in the middle or posterior mediastinum.

Posterior mediastinal mass

Neurogenic tumor

• A neurogenic tumor may arise from either a peripheral nerve or the sympathetic ganglia.

Most adult tumors are peripheral nerve sheath tumors and the vast majority of tumors in children are of sympathetic ganglionic origin.

Overall, neurogenic tumors are the most common posterior mediastinal masses.

• Peripheral nerve tumors (more common in adults) include:

Schwannoma (most common), **neurofibroma**, and **malignant peripheral nerve sheath tumor**.

• Sympathetic ganglion tumors (more common in children/young adults) include:

Ganglioneuroma (most common), a benign tumor of sympathetic ganglion cells.

Neuroblastoma, a malignant tumor of ganglion cells seen in early childhood.

Ganglioneuroblastoma, intermediate in histology between ganglioneuroma and neuroblastoma, and neurenteric cysts. Foregut duplication cysts may occur in the middle or posterior

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Hiatal hernia

• A hiatal hernia is protrusion of a portion of the stomach through the esophageal hiatus.

The esophageal hiatus is an elliptical opening in the diaphragm just to the left of midline.

• On radiography, air or an air–fluid level is present above the diaphragm.

Descending thoracic aortic aneurysm

• Thoracic aortic aneurysm may appear as a posterior mediastinal mass on radiography.

Extramedullary hematopoiesis

• Extramedullary hematopoiesis causes soft tissue paravertebral masses in patients with severe hereditary anemias including thalassemia and sickle cell anemia.

The masses are of uncertain origin but may represent herniation of vertebral marrow or may represent elements of the reticuloendothelial system.

• On imaging, lobulated soft tissue masses are typically bilateral and inferior to T6.

Lateral meningocele

• A lateral meningocele is lateral herniation of the spinal meninges through either an intervertebral foramen or a defect in the vertebral body.

Lateral meningocele is associated with neurofibromatosis.

Esophageal neoplasm

• Esophageal carcinoma can present on radiography as abnormal convexity of the azygoesophageal recess, mediastinal widening, or a retrotracheal mass.

Benign mesenchymal esophageal tumors, such as leiomyoma, fibroma, or lipoma may appear similar on radiography.

Foregut duplication cyst

• A foregut duplication cyst, a remnant of the fetal foregut, may present as a middle or posterior mediastinal mass.

Paraspinal abscess

• A clue to vertebral body pathology on radiography may be paraspinal line displacement.

Airways

Multifocal or diffuse non-neoplastic tracheal stenosis/wall thickening

Relapsing polychondritis

• Relapsing polychondritis is a multisystemic disease of unknown etiology characterized by recurrent inflammation of cartilaginous structures.

The nose, ear, joints, larynx, trachea, and bronchi can be affected, with airway involvement seen in 50% of patients.

The larynx and subglottic trachea are the most common sites of airway involvement.

• Relapsing polychondritis usually occurs in middle-aged women.

• On cross-sectional imaging, there is smooth tracheal/bronchial wall thickening, with *sparing of the posterior membranous* trachea.

The tracheal cartilage is an incomplete ring, with no cartilage in the posterior membranous portion.

• There is often increased attenuation of the airway wall, ranging from subtly increased attenuation to frank calcification.

Tracheobronchopathia osteochondroplastica (TPO)

• Tracheobronchopathia osteochondroplastica (TPO) is a benign condition of multiple submucosal calcified osteocartilaginous nodules along the tracheal walls.

Tuberculosis

• Endobronchial spread of tuberculosis occurs in a prominent minority of patients with pulmonary tuberculosis, most commonly involving the distal trachea and proximal

bronchi.

• Imaging findings are nonspecific. There is usually smooth concentric narrowing of a relatively long airway segment (typically >3cm).

• Similar to relapsing polychondritis, there is sparing of the posterior membranous trachea.

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Amyloidosis

• Amyloidosis causes irregular narrowing of the airways due to submucosal amyloid deposition, which may be calcified. Tracheal amyloidosis is very rare.

• The posterior membranous trachea is not spared.

Wegener granulomatosis

• Large airway involvement is seen in approximately 20% of patients with Wegener granulomatosis, most commonly manifesting as subglottic tracheal stenosis with circumferential mucosal thickening.

• The posterior membranous trachea is not spared in Wegener granulomatosis.

Sarcoidosis

• Tracheal involvement by sarcoid is rare and usually seen in advanced disease. Tracheal sarcoid has a variable appearance ranging from smooth stenosis to a nodular or masslike

appearance.

• The posterior membranous trachea is not spared in tracheal sarcoidosis.

Focal non-neoplastic tracheal stenosis/wall thickening

Intubation/tracheostomy

• There is approximately 1% risk of tracheal stenosis after intubation, but approximately 30% risk of stenosis after long-standing tracheostomy.

Rare causes of focal tracheal stenosis

• Extremely uncommon causes of focal tracheal stenosis include Behçet and Crohn disease.

Large airways

Bronchiectasis

• Bronchiectasis is progressive, irreversible dilation of cartilage-containing bronchi.

• Three etiologies of bronchiectasis have been described, with a final common pathway of mucus plugging, superimposed bacterial colonization, and inflammatory response.

Bronchial wall injury, typically from infection or inflammation.

Bronchial lumen obstruction.

Traction from adjacent fibrosis.

• Specific causes of bronchiectasis include:

Ineffective clearing of secretions – **cystic fibrosis** and **Kartagener** (primary ciliary dyskinesia).

Bronchiectasis from cystic fibrosis with superimposed pneumonia: Radiograph shows

upper-lobe bronchiectasis with a focal left upper lobe opacity (yellow arrow). CT confirms bronchiectasis (red arrows) and a left upper lobe consolidation, representing pneumonia.

Recurrent pneumonia.

Bronchocentric infections, such as **tuberculosis** and **atypical mycobacteria.**

Exaggerated immune response, as seen in **ABPA** and **vasculitis.**

Impaired immunity: **Congenital immunodeficiency**, **transplant recipients,** and **agammaglobulinemia.**

Congenital connective tissue disorders – **Mounier–Kuhn** (a connective tissue disorder causing tracheobronchomegaly leading to recurrent pneumonia), or **Williams–Campbell** (a rare disorder of the distal bronchial cartilage, which may be congenital or acquired as a sequela of viral infection).

CAPTAI*n* ***K****angaroo has* ***Mounier–Kuhn*** is a mnemonic to remember the common causes of bronchiectasis.

Cystic fibrosis is one of the most common causes of bronchiectasis.

Allergic bronchopulmonary aspergillosis (ABPA).

Post infectious.

TB/atypical mycobacteria.

Agammaglobulinemia.

Immunodeficiency.

Kartagener.

Mounier–Kuhn.

• Cystic fibrosis tends to be upper lobe predominant. In contrast, post-infectious bronchiectasis tends to be lower-lobe predominant.

• Morphologic classification of bronchiectasis is most useful as a rough gauge of severity.

Cylindrical bronchiectasis (least severe): Mild bronchial dilation.

Varicose bronchiectasis (moderately severe): Bronchi may become beaded and irregular.

Cystic bronchiectasis (most severe): Bronchi are markedly enlarged and ballooned, with formation of multiple cysts that may not connect to the airways.

• Radiographic findings depend on severity. In mild cases only *tram tracks* may be visible, representing thickened bronchial walls causing parallel radiopaque lines resembling tram tracks. In more severe cases there can be extensive cystic change.

• CT findings include the *signet ring* sign, which describes a dilated bronchus adjacent to a normal pulmonary artery branch.

Normally each bronchus should be approximately the same size as the adjacent pulmonary artery branch.

Other CT findings of bronchiectasis include lack of bronchial tapering, bronchial wall thickening, and mucus-filled bronchi. Often, adjacent tree-in-bud nodules are present, likely representing associated small-airways infection.

Broncholithiasis

• Broncholithiasis is a rare disorder of calcified/ossified material within the bronchial lumen, most commonly caused by erosion of an adjacent calcified granulomatous lymph node.

• Broncholithiasis clinically presents with nonproductive cough, hemoptysis, and air trapping.

Emphysema

• Emphysema is the destruction of alveolar walls resulting in irreversible enlargement of the distal airspaces.

Although emphysema initially does not involve fibrosis, end-stage emphysema may lead to a reparative response that ultimately leads to pulmonary fibrosis.

• Elastase is produced by alveolar macrophages and neutrophils, both of which are increased in smokers.

Elastase is a powerful destructive enzyme which functions in

the host defense mechanism, but excess elastase can be highly harmful to the native tissues.

Alpha-1-antitrypsin normally neutralizes elastase. Either a surplus of elastase (in smoking-related emphysema) or a deficiency of neutralizing enzyme (in alpha-1-

antitrypsin) can cause lung destruction and resultant emphysema.

Centrilobular emphysema

• Centrilobular emphysema is a smoking-related lung disease.

• Centrilobular emphysema predominantly affects the upper lobes. Like RB-ILD, another smoking-related lung disease, centrilobular emphysema primarily affects the center of

the secondary pulmonary lobule.

All smoking-related lung disease (RB, RB-ILD, DIP, PLCH, and emphysema) may be within the same spectrum of disease caused by macrophage-mediated inflammation in reaction to inhaled particles and toxins.

Paraseptal emphysema

• Paraseptal emphysema is usually seen in combination with other forms of emphysema.

It is also usually smoking related.

• Paraseptal emphysema is subpleural in location and may predispose to pneumothorax.

Panacinar (panlobular) emphysema

• Panacinar (also called panlobular) emphysema affects the entire acinus diffusely throughout the lung.

The emphysematous changes are usually more severe at the

lung bases.

• Alpha-1-antitrypsin deficiency is an important cause of panacinar emphysema.

Airway tumors

• Primary tumors of the trachea and central bronchi are rare. In adults, the vast majority of tumors are malignant, while in children most are benign.

• Squamous cell carcinoma and adenoid cystic carcinoma are by far the two most common primary central airway tumors in adults.

Squamous cell carcinoma (SCC)

• Squamous cell carcinoma is the most common primary tracheal malignancy.

It is strongly associated with cigarette smoking.

• The **typical CT appearance** of tracheal squamous cell carcinoma is a polypoid intraluminal mass. The contours of the mass can be irregular, smooth, or lobulated.

The tumor can occasionally invade into the esophagus, causing tracheoesophageal fistula.

Adenoid cystic carcinoma (ACC)

• Adenoid cystic carcinoma (ACC) is a relatively low grade malignancy that usually affects patients in their forties, a decade or two younger than the typical SCC patient.

It is not associated with cigarette smoking.

• ACC has a propensity for perineural and submucosal spread. Despite indolent growth, metastatic disease tends to have intense FDG uptake on PET imaging.

• The typical CT appearance of ACC is a submucosal mass that infiltrates the tracheal wall and surrounding mediastinal fat. ACC may also present as circumferential tracheal or bronchial thickening causing airway stenosis.

Carcinoid

Carcinoid is a spectrum of neuroendocrine neoplasms including low-grade carcinoid, aggressive carcinoid, and small cell carcinoma, featuring a range of biologic behaviors.

Carcinoid tumors may rarely secrete hormones such as ACTH.

While endobronchial carcinoid is rare in adults, it is the most common bronchial tumor in children.

• Carcinoid almost always occurs distal to the carina.

• CT shows an endoluminal bronchial mass with homogeneous arterial enhancement.

In addition to carcinoid, the differential diagnosis of an enhancing endobronchial mass includes:

mucoepidermoid carcinoma and very rare entities such as **hemangioma** and **glomus tumor**.

Mucoepidermoid carcinoma

• Mucoepidermoid carcinoma is a rare tumor that originates from tiny salivary glands lining the tracheobronchial tree.

• Mucoepidermoid carcinoma tends to affect younger patients than adenoid cystic carcinoma.

• CT appearance is a round or oval endobronchial mass, indistinguishable from carcinoid.

Tracheal lymphoma

• Tracheal lymphoma is rare. It is usually associated with mucosa-associated lymphoid tissue (MALT), a low-grade malignancy.

Endobronchial metastasis

Breast cancer, renal cell carcinoma, thyroid cancer, lung cancer, melanoma, and sarcoma are the most common malignancies to metastasize to the central airways.

• The mnemonic **BR**e**T**h **Lung** may be helpful to remember the four most common airway metastases (breast, renal cell, thyroid, and lung).

Direct invasion of the central airways by adjacent malignancy

• Direct central airway invasion occurs more commonly than endobronchial metastases.

Aggressive laryngeal, thyroid, esophageal, and lung cancer may cause direct airway invasion.

Benign endobronchial lesions

• Papilloma is a benign but potentially pre-malignant lesion that may transform into carcinoma.

Suspected papillomas are typically closely followed.

A single papilloma is usually caused by chronic irritation.

Multiple papillomas (laryngotracheal papillomatosis) are caused by HPV, which may be acquired at birth.

Rarely (<1% of cases), papillomas spread to the lungs, where they may form multiple cavitary nodules.

• Chondroma is a benign cartilaginous tumor that rarely may occur in the airways.

• Other benign endobronchial lesions include schwannoma, adenoma, hamartoma, hemangioma, lipoma, and leiomyoma.

Pleura

Pleural malignancy

Mesothelioma

• Mesothelioma is a highly aggressive neoplasm arising from the pleura.

Most cases are due to prior asbestos exposure, with a latency of >20 years.

• The epithelial subtype is more common and has a slightly better prognosis.

Sarcomatoid and mixed subtypes are more aggressive.

• CT of mesothelioma typically shows nodular concentric pleural thickening, often with an associated pleural effusion.

• The role of surgery is evolving, with the goal to resect all visible tumor.

• **Extrapleural pneumonectomy** is performed for patients with locally invasive disease.

• **Pleurectomy and decortication** is preferred for less advanced disease, where the lung and fissures are spared.

Prognosis is equivalent to pleurectomy or pneumonectomy

for the less aggressive epithelial subtype.

• Trimodality therapy involving surgery, intraoperative heated chemotherapy, and radiation has been shown to provide benefit for a subset of patients.

Metastases

• Lung cancer, gastrointestinal and genitourinary adenocarcinoma, and invasive thymoma can metastasize to the pleura.

Multiple myeloma/plasmacytoma

• Rib-based lesions may appear to be pleural-based, although the epicenter of a mass arising from a rib will be centered on the rib.

Fibrous tumor of the pleura (FTP)

• Fibrous tumor of the pleura (FTP) is a focal pleural mass not related to asbestosis or mesothelioma.

It is not mesothelial in origin.

• Approximately 20–30% of FTP are malignant, so all are excised. Malignant potential is determined by number of mitoses seen at pathology.

• FTP may be associated with hypoglycemia or hypertrophic pulmonary osteoarthropathy, although these associated conditions are uncommon (5%).

• FTP may be pedunculated. A pleural-based mass that changes position is suggestive of FTP.

FTP tends to have low FDG uptake on PET.

Pleural effusion

Transudate

• A transudative effusion is caused by systemic or local imbalances in hydrostatic and oncotic forces.

Common causes include systemic low-protein states, heart failure, and nephrotic syndrome.

Exudate

• An exudative effusion is distinguished from a transudate by thoracentesis.

There are no reliable imaging features to distinguish between transudative and exudative effusions.

• Analysis of the thoracentesis fluid by Light's criteria is the most reliable method to distinguish transudate from exudate.

If **any** one of the following is present, the effusion is classified as an exudate:

Ratio of pleural fluid protein to serum protein >0.5.

Ratio of pleural fluid LDH to serum LDH >0.6.

Pleural fluid LDH >2/3 upper limits of normal for serum.

• The presence of an exudate implies pleural disease causing increased permeability of pleural capillaries, which may be due to:

Pneumonia with parapneumonic effusion, **empyema**, or **tuberculous** pleuritis.

Mesothelioma or pleural **metastasis**.

Rheumatoid arthritis or other collagen vascular diseases.

Chylothorax

• A chylothorax is a pleural effusion consisting of intestinal lymph, most commonly caused by neoplastic obstruction of the thoracic duct.

Chylothorax is also associated with lymphangioleiomyomatosis (LAM).

• The thoracic duct originates at the cisterna chyli in the upper abdomen and drains into the left brachiocephalic or subclavian vein.

Gastrointestinal imaging

Liver

Liver anatomy

• The Couinaud classification divides the liver into eight segments.

Because each segment is self-contained, an individual segment can be completely resected without disturbing the other segments.

• Numbering of hepatic segments is clockwise when looking at a frontal/coronal view.

• **Each hepatic segment features its own:**

Central portal triad including branches of the portal vein, hepatic artery, and bile duct.

Peripheral venous drainage to the hepatic veins and ultimately the IVC.

• Mnemonic for remembering the segments:

Superior segments, from left to right: 2, 4, 8, 7, 1 (caudate). 2 doubled is 4; 4 doubled is 8; 8 minus 1 is 7.

Inferior segments, from left to right: 3, 4, 5, 6.

• Segments 2, 3, and 4 are in the left lobe of the liver.

• Segments 5, 6, 7, 8 are in the right lobe of the liver.

• The left and right main portal veins divide the superior from the inferior segments and continue to branch superiorly and inferiorly before terminating in the center of each segment.

• The branching of the portal veins is variable. The most common pattern is a bifurcation into right and left main portal veins, with the right main portal vein then branching into anterior and posterior branches.

• Small hepatic vein tributaries mark the peripheral margins of each segment.

• The caudate lobe drains directly to the IVC, not into the hepatic veins.

The caudate lobe is spared in early cirrhosis since the direct drainage to the IVC spares the caudate from increased venous pressures due to portal hypertension.

This leads to compensatory hypertrophy of the caudate lobe, which is a typical morphologic change of early cirrhosis.

Similarly, direct venous drainage to the IVC allows the caudate lobe to bypass the increased hepatic venous pressures seen in Budd–Chiari syndrome.

Compensatory hypertrophy of the caudate lobe

may preserve liver function in these patients.

CT and MRI of the liver

Liver CT

• A “routine” contrast-enhanced abdominal CT is acquired in the portal venous phase of enhancement, typically obtained 70 seconds following intravenous contrast administration.

• A portal venous phase CT reveals characteristic attenuation alterations and/or morphologic changes of diffuse liver disease, such as hepatic steatosis and cirrhosis.

Most metastatic tumors are *not* hypervascular (although a few notable exceptions will be subsequently discussed) and can also generally be detected on the portal venous phase.

Of note, rarely some breast cancers may be isoattenuating on the portal venous phase and more conspicuous on unenhanced CT.

• Most benign and malignant primary liver masses are hypervascular and thus most conspicuous in the arterial phase of enhancement.

The arterial phase begins approximately 20–25 seconds after intravenous contrast injection.

Many authors advocate that optimal conspicuity of a hypervascular liver lesion is obtained in the late arterial phase, which is between 9 and 16 seconds after abdominal aortic

enhancement, or approximately 35 seconds after intravenous injection.

Liver MRI

• Compared to CT, MRI of the liver displays the same patterns of contrast enhancement but has superior lesion-to-liver contrast. MRI also does not impart ionizing radiation.

• MRI is able to obtain dynamic post-contrast images in multiple phases without any penalty in radiation exposure.

In- and out-of-phase gradient imaging allows detection

of intracytoplasmic lipid, which is seen in hepatic steatosis. Additionally, advanced techniques such as diffusion-weighted imaging may show potential for clinical use.

Hepatic metabolic disorders (diffuse liver disease)

Fatty liver (hepatic steatosis)

• Nonalcoholic fatty liver disease can be divided into steatosis and steatosis with associated inflammatory activity (steatohepatitis).

Overall, greater than 15% of the population is afflicted with nonalcoholic fatty liver disease (NAFLD), which is a

component of the metabolic syndrome of obesity, insulin resistance, and dyslipidemia.

Ultimately, steatohepatitis may progress to cirrhosis.

• CT can determine if steatosis is present and can provide a rough gauge as to its severity.

In- and out-of-phase MRI imaging can more accurately quantify the degree of steatosis, although liver biopsy is the gold standard and best evaluates for the presence of inflammatory and early fibrotic change.

By the time imaging can detect morphologic changes of cirrhosis, the changes may be irreversible.

• On unenhanced CT, the liver should be slightly hyperattenuating relative to the spleen.

The traditional teaching is that steatosis is present if the liver attenuates at least 10 Hounsfield units (HU) less than the spleen, although new work suggests that even a single HU of relative hypoattenuation compared to the spleen may represent hepatic steatosis.

• On contrast-enhanced CT, evaluation of hepatic steatosis is much less reliable compared to unenhanced CT due to different contrast uptake rates of the liver and the spleen.

However, the liver is considered diffusely hypoattenuating if it attenuates atleast 25 HU less than the spleen in the portal venous phase.

• In- and out-of-phase GRE MRI is a sensitive imaging technique to evaluate for the presence of (and to quantify the degree of) hepatic steatosis.

Variations in portal venous supply may cause geographic regions that are more or less affected by fatty change.

Focal fat does not have any mass effect, vessels characteristically run through it, and it tends to occur in the following typical locations and distributions:

Gallbladder fossa (drained by gallbladder vein).

Subcapsular (along the falciform ligament).

signal intensity on out-of-phase dual-phase GRE, consistent with nodular focal fat.

Periportal.

Focal fat may also be nodular throughout the liver. Ultrasound would demonstrate multiple hyperechoic lesions which would be hypoattenuating on CT. MRI shows pseudo-lesions drop in signal intensity on out-of-phase dual-phase GRE, consistent with nodular focal fat.

Amyloid

• Abnormal extracellular deposition of amyloid protein in the liver can cause focal or diffuse areas of decreased attenuation on CT imaging.

Wilson disease

• Wilson disease causes high levels of copper to accumulate in the basal ganglia, cornea, and liver due an autosomal recessive genetic defect.

The liver may be hyperattenuating on CT with multiple nodules, eventually leading to hepatomegaly and cirrhosis.

Hepatic iron overload

• There are two pathways to excess hepatic iron accumulation. Accumulation within hepatocytes is seen in hemochromatosis. Uptake within the reticuloendothelial system (RES) causes hepatic Kupffer cell iron overload, as seen in hemosiderosis.

• Regardless of the etiology, the iron-overloaded liver is hypointense on all MRI sequences, relative to the paraspinal muscles as an internal control.

• Hemochromatosis is the most common cause of iron overload, due to a genetic defect causing increased iron absorption.

Excess iron is *unable* to be stored in the RES, so the spleen and bone marrow are not affected. Treatment of hemochromatosis is phlebotomy.

Excess iron is deposited in **hepatocytes** (not the Kupffer cells that make up the intrahepatic RES), pancreas, myocardium, skin, and joints. Excess iron in hepatocytes can cause cirrhosis.

The spleen and bone marrow are normal since the RES is not involved.

• Hemosiderosis is excess iron stored within the reticuloendothelial system, which may be due to frequent blood transfusions or defective erythrocytosis.

Treatment of hemosiderosis is with iron chelators, not phlebotomy.

The RES has a large capacity for iron. Iron stored in the RES is generally not harmful and the liver is normal in morphology, without cirrhosis.

MRI imaging of hemosiderosis demonstrates hypointense liver on conventional MRI sequences, similar to hemochromatosis. Additionally, the spleen and bone marrow will also appear

hypointense due to increased iron stores throughout the entire reticuloendothelial system.

• Hemosiderosis is a precursor to secondary hemochromatosis. Secondary hemochromatosis is hepatic damage from iron overload after the RES system becomes saturated from prolonged hemosiderosis.

When the RES becomes overwhelmed with iron, the hepatocytes begin to store the excess.

Similar to hemochromatosis, hepatocyte iron uptake may lead to cirrhosis.

• In clinical practice, the distinction between hemosiderosis and secondary hemochromatosis often overlaps. Many authors recommend against the term "secondary hemochromatosis" in favor of describing the primary disease (e.g., thalassemia) with secondary iron overload.

Differential based on CT attenuation

• **Hypoattenuating** liver: The liver is considered hypoattenuating if it attenuates less than the spleen on an unenhanced CT.

Fatty liver (hepatic steatosis) is by far the most common cause of a diffusely hypoattenuating liver.

Hepatic amyloid is rare and may cause either focal or diffuse hepatic hypoattenuation.

• **Hyperattenuating** liver: The normal unenhanced attenuation of the liver is 30 to 60 HU.

An absolute attenuation greater than 75 HU is considered hyperattenuating.

Iron overload is by far the most common cause of a hyperattenuating liver.

Medications (e.g., amiodarone, gold, and methotrexate).

Copper overload (Wilson disease).

Glycogen excess.

Hepatic infection

Viral hepatitis

• Patients with viral hepatitis often have a normal CT scan. Viral hepatitis may cause nonspecific CT findings, such as gallbladder wall thickening or periportal edema (fluid on both sides of the portal veins).

Candidiasis

• Systemic fungal infection may seed the liver (and commonly the spleen as well) due to portal venous drainage of infected bowel.

• CT shows multiple tiny hypoattenuating microabscesses in the liver and the spleen, which may be rim-enhancing.

• Candidiasis is almost always seen in immunocompromised patients.

• The differential diagnosis for multiple tiny hypoattenuating hepatic lesions includes metastatic disease, lymphoma, biliary

hamartomas, and Caroli disease.

Candidiasis:

Noncontrast CT shows innumerable tiny hypoattenuating lesions scattered throughout the liver and spleen, representing

candidal microabscesses in a bone marrow transplant patient with fungemia.

Abscess

• Hepatic abscess is most commonly caused by a bowel process and resultant infectious nidus carried through the portal system to the liver.

Common causes include diverticulitis, appendicitis, Crohn disease, and bowel surgery. *E. coli* is the most common organism.

A primary hepatobiliary infection, such as ascending cholangitis, may be a less common cause.

• Imaging features of hepatic abscess may mimic metastasis, appearing as a ring-enhancing mass on CT.

On MRI, there is typically central hyperintensity on T2-weighted images with an irregular wall that enhances late. Perilesional enhancement may be present.

Echinococcal disease

• Hepatic echinococcosis is caused by ingestion of the eggs of *Echinococcus granulosus*, which is endemic in the Mediterranean basin and associated with sheep-raising.

Echinococcal eggs can develop into hydatid cysts.

• **On CT**, a hydatid cyst is a well-defined hypoattenuating mass featuring a characteristic floating membrane or an associated daughter cyst.

Peripheral calcification may be present.

Cirrhosis

Etiology and pathology

• Cirrhosis is caused by repeated cycles of injury and repair, which can be due to metabolic (alcohol, steatohepatitis, hemochromatosis, or Wilson disease), infectious (chronic hepatitis B or C), or inflammatory (primary biliary cirrhosis or primary sclerosing cholangitis) etiologies.

The hallmarks of cirrhosis are fibrosis and attempted, disorganized regeneration.

• The micronodular form of cirrhosis is most often due to metabolic causes.

• The macronodular form of cirrhosis is most often post-viral (hepatitis B or C).

Early signs of cirrhosis

• One of the earliest signs of cirrhosis is expansion of the pre-portal space.

Atrophy of the medial segment of the left hepatic lobe in early cirrhosis causes increased fat anterior to the right main portal vein.

• Enlargement of the caudate lobe is a specific sign of cirrhosis. Specifically, a caudate to right lobe size ratio of >0.65 highly suggests cirrhosis.

As previously discussed, the caudate drains directly to the IVC, not via the hepatic veins, which results in compensatory caudate hypertrophy.

• The *empty gallbladder fossa* sign results when hepatic parenchyma surrounding the gallbladder is replaced with periportal fat.

Secondary manifestations of cirrhosis

• Portal hypertension causes splenomegaly, portosystemic collaterals, and varices.

• Gallbladder wall thickening is due to hypoalbuminemia and resultant edema.

• **Gamna–Gandy bodies** are splenic microhemorrhages, which appear hypointense on GRE.

Malignant hepatic masses

Pathway to hepatocellular carcinoma

• In the setting of cirrhosis, hepatocellular carcinoma (HCC) is thought to develop in a sequence from regenerative nodule to dysplastic nodule to HCC.

Regenerative and dysplastic nodules cannot be reliably differentiated on imaging; and high-grade dysplastic nodules cannot be reliably differentiated from low-grade HCC.

• **Regenerative nodule**: A regenerative nodule is completely supplied by the portal vein and is not premalignant.

A regenerative nodule should *not* enhance in the arterial phase.

Most regenerative nodules show low signal intensity on T2-weighted images, with variable signal intensity on T1-weighted images.

Rarely, a regenerative nodule may be hyperintense on T1-weighted images due to glycogen deposition.

On contrast-enhanced MRI, most regenerative nodules enhance to the same (or slightly less) degree as the adjacent hepatic parenchyma.

• **Dysplastic nodule**: Unlike a regenerative nodule, a dysplastic nodule is premalignant.

However, most dysplastic nodules do not demonstrate arterial phase enhancement (unless high grade), since blood supply is still from the portal vein.

Dysplastic nodules are variable in signal intensity on T1-weighted images.

Most dysplastic nodules are hypointense on T2-weighted images, although high-grade dysplastic nodules may be T2 hyperintense.

Contrast-enhanced MRI shows low-grade dysplastic nodules to be iso-enhancing relative to liver and thus indistinguishable from regenerative nodules. High grade dysplastic nodules may demonstrate arterial enhancement and be indistinguishable from well-differentiated hepatocellular carcinoma.

• A siderotic nodule is an iron-rich regenerative or dysplastic nodule. A siderotic nodule is hypointense on T1 and T2*-weighted images and hyperattenuating on CT.

A siderotic nodule is rarely, if ever, malignant.

Hepatocellular carcinoma (HCC)

• Hepatocellular carcinoma (HCC) is the most common primary liver tumor.

Cirrhosis is the major risk factor for development of HCC. A hypervascular liver mass in a patient with cirrhosis or chronic hepatitis is an HCC until proven otherwise.

• Alpha-feto protein (AFP) is elevated in approximately 75% of cases of HCC.

• Arterial phase enhancement is the characteristic imaging feature of HCC.

However, between 10 and 20% of HCCs are hypovascular and thus slightly hypoenhancing relative to surrounding liver on arterial phase imaging.

• **The classic CT or MRI appearance** of HCC is an encapsulated mass that enhances on arterial phase and washes out on portal venous phase.

HCC may be difficult to detect on non-contrast or portal venous phase CT.

On unenhanced MRI, HCC is characteristically slightly hyperintense on T2-weighted images relative to surrounding liver.

The nodule in a nodule appearance describes an enhancing nodule within a dysplastic nodule and represents an early HCC.

• HCC is locally invasive and tends to invade into the portal and portal veins, IVC, and bile ducts.

In contrast, metastases to the liver are much less likely to be locally invasive.

• Treatment options for HCC include partial hepatectomy, orthotopic liver transplantation, percutaneous ablation, and transcatheter embolization.

Fibrolamellar HCC

• Fibrolamellar carcinoma is a subtype of HCC that occurs in young patients without cirrhosis.

• The tumor tends to be large when diagnosed, but has a better prognosis than typical HCC.

Unlike in HCC, AFP is not elevated.

• On MRI, fibrolamellar HCC is a large, heterogeneous mass. A fibrotic central scar is classic, which is hypointense on T1- and T2-weighted images (in contrast, focal nodular hyperplasia features a T2 hyperintense scar that enhances late). Capsular

retraction may be seen in 10%.

• Unlike HCC, the fibrolamellar subtype does not have a capsule, although there may be a pseudocapsule of peripherally compressed normal hepatic tissue.

Hepatic metastases

• Although metastases are supplied by branches of the hepatic artery induced by tumoral angiogenesis, most metastases are hypovascular and best appreciated on portal venous phase (in contrast to HCC, which is hypervascular and best visualized on late arterial phase).

• Hypervascular metastases (best seen on arterial phase) classically include:

Neuroendocrine tumors, including pancreatic neuroendocrine tumors and carcinoid.

Renal cell carcinoma.

Thyroid carcinoma.

Melanoma.

Sarcoma.

• Colorectal and pancreatic adenocarcinoma metastases are typically hypovascular and can usually be diagnosed on portal venous imaging.

• Calcifications can be seen in mucinous colorectal tumors or ovarian serous tumors.

Calcification within a metastatic lesion may imply a better prognosis.

• On MRI, metastatic lesions tend to be hypointense on T1-weighted images and hyperintense on T2-weighted images. Blood products and melanin (as in melanoma) are T1 hyperintense.

• *Pseudocirrhosis* describes the macronodular liver contour resulting from multiple scirrhous hepatic metastases, which may mimic cirrhosis.

Treated breast cancer is the most common cause of this appearance.

Capsular retraction, although not always seen, is characteristic

of pseudocirrhosis, and when present suggests pseudocirrhosis over cirrhosis.

Hepatic lymphoma

• Primary hepatic lymphoma is very rare. Lymphomatous involvement of the liver tends to be secondary to systemic disease, with associated splenomegaly and lymphadenopathy.

Epithelioid hemangioendothelioma

• Epithelioid hemangioendothelioma is a rare vascular malignancy that characteristically causes multiple spherical subcapsular masses than can become confluent.

The individual masses may have a *halo* or *target* appearance.

• Epithelioid hemangioendothelioma is one cause of capsular retraction.

Hepatic capsular retraction

• Capsular retraction is a focal concavity of the normally convex external liver contour.

differential diagnosis of capsular retraction

• **Metastatic tumor** (more commonly post-treatment).

• **Fibrolamellar hepatocellular carcinoma** (10% of cases of fibrolamellar HCC).

• **Hepatocellular carcinoma** (capsular retraction has been reported but is uncommon).

• **Epithelioid hemangioendothelioma.**

•. **Intrahepatic cholangiocarcinoma.**

• **Confluent hepatic fibrosis** (wedge-shaped fibrosis seen in cirrhosis, most commonly the medial segment of the left hepatic lobe or the anterior segment of the right hepatic lobe).

Benign liver masses

Focal nodular hyperplasia (FNH)

• Focal nodular hyperplasia (FNH) is disorganized liver tissue with no malignant potential.

It is primarily seen in asymptomatic women and is not associated with oral contraceptives.

• FNH has a characteristic central “scar” which does not contain fibrotic tissue and is therefore not a true scar.

Instead, the central area consists of T2-hyperintense ductules

and venules, and demonstrates delayed enhancement. FNH does not have a capsule.

• FNH can be difficult to see without contrast on CT and T1- and T2-weighted MRI sequences.

FNH avidly enhances during the arterial phase, then washes out very quickly.

The portal venous phase will often show just the unenhanced scar, which enhances late.

• Kupffer cells and bile duct epithelium are both present. Kupffer cells may be confirmed by a sulfur colloid study (1/3 of the time) and bile duct cells can be seen on a HIDA scan.

Hemangioma

• A hepatic hemangioma is a benign mass composed of disorganized endotheliallined pockets of blood vessels, supplied by a branch of the hepatic artery at the periphery.

• Hemangioma is more common in females and uncommon in cirrhosis.

When a known hemangioma is sequentially followed in a patient with early cirrhosis, the hemangioma involutes as the liver becomes more cirrhotic.

• Hemangiomas may range in size from <1 cm to >10 cm. Giant hemangiomas tend to have a nonenhancing central area representing cystic degeneration.

• A virtually pathognomonic imaging feature is peripheral, discontinuous, progressive, nodular enhancement.

The attenuation (or signal intensity on MR) of the

enhancement is identical to the aorta and features gradual centripetal fill-in on later

phases.

• The unenhanced CT appearance of a hemangioma is a nonspecific hypoattenuating liver mass.

Hepatic adenoma

• Hepatic adenoma is a benign hepatic neoplasm containing hepatocytes, scattered Kupffer cells, and no bile ducts.

The absence of bile ducts makes a nuclear medicine HIDA scan a useful test to distinguish between focal nodular hyperplasia (which contains bile ducts and would be positive on HIDA) and a hepatic adenoma, which does not contain bile ducts.

• Adenomas are much more common in females, especially with prolonged oral contraceptive use.

When seen in males, adenoma may be associated with anabolic

steroids.

• Adenomas have a relatively high risk of hemorrhage, which is often the presenting symptom.

For this reason, incidentally discovered adenomas are usually resected.

• Multiple hepatic adenomas are seen in von Gierke disease (type I glycogen storage disease).

• A pseudocapsule may be present, which tends to enhance late.

• Adenomas lack portal venous drainage and thus are hypervascular on arterial phase.

The presence of microscopic fat, when present, is best seen on in- and out-of-phase MRI.

Intralesional hemorrhage may cause T1 hyperintensity. Adenomas may be difficult to differentiate from other hypervascular liver lesions in the absence of fat or hemorrhage.

Vascular liver disease

Budd-Chiari

• Budd–Chiari is hepatic venous outflow obstruction, which can be thrombotic or nonthrombotic.

Budd–Chiari may be due to hypercoagulative states including hematological disorders, pregnancy, oral contraceptives, malignancy, infection, and trauma.

It is very rare to have primary Budd–Chiari due to congenital hepatic vein anomaly.

• Acute Budd–Chiari presents with a clinical triad of hepatomegaly, ascites, and abdominal pain.

• Direct vascular findings include lack of flow within hepatic veins, thrombus in the hepatic veins/IVC,

and the formation of collateral vessels.

• Acute intraparenchymal findings include an edematous peripheral liver with sparing of the caudate

lobe.

The caudate is spared as it drains directly into the IVC.

• Progressive liver failure may result in chronic disease, producing caudate lobe hypertrophy and atrophy of peripheral liver with prominent regenerative nodules.

Veno-occlusive disease

• Veno-occlusive disease (VOD) is destruction of post-sinusoidal venules, with patent hepatic veins. VOD is seen in bone marrow transplant patients, possibly due to chemotherapy.

• Imaging findings are nonspecific. Periportal edema, narrowing of the hepatic veins, hepatomegaly, and heterogeneous hepatic enhancement have been reported.

In contrast to Budd–Chiari, the caudate lobe is not spared.

Cardiac hepatopathy

• Cardiac hepatopathy is passive hepatic congestion from heart failure, constrictive pericarditis, or right-sided valvular disease, which ultimately may lead to cirrhosis.

• Imaging clues are enlarged hepatic veins and IVC, with reflux of intravenous contrast from the right atrium into the IVC and hepatic veins.

The liver is typically enlarged and demonstrates mottled enhancement.

Ascites is usually present.

Congenital cystic liver disease

Biliary hamartomas (von Meyenburg complexes)

• Biliary hamartomas are incidental small cystic hepatic lesions that do not communicate with the biliary tree, caused by embryologic failure of normal bile duct formation.

• Biliary hamartomas tend to be smaller and more irregularly shaped than simple cysts.

Autosomal dominant polycystic liver disease (ADPLD)

• 40% of patients with autosomal dominant polycystic kidney disease (ADPKD) have a similar disease process in the liver, called ADPLD. Even in severe disease, hepatic failure is rare.

• On imaging, there are innumerable nonenhancing simple cysts throughout the liver.

Liver trauma

Overview of liver trauma

• The liver is the second most commonly injured solid organ due to blunt trauma, second to the spleen.

The CT description and grading of liver injury is similar to splenic injury.

• The American Association for the Surgery of Trauma (AAST) classification describes hepatic injury based on findings at laparotomy.

• The MDCT hepatic injury grading scale is based on CT findings and is more commonly used by radiologists. It is similar to the MDCT grading scale for splenic trauma.

MDCT hepatic trauma

MDCT grading of hepatic injury

• Grade I: Superficial laceration or subcapsular hematoma <1 cm in size.

• Grade II: Laceration or subcapsular/intraparenchymal hematoma >1 and <3 cm in size.

• Grade III: Laceration or subcapsular/intraparenchymal hematoma >3 cm in diameter.

• Grade IV: Massive hematoma >10 cm, or destruction/devascularization of one hepatic lobe.

• Grade V: Destruction or devascularization of both hepatic lobes.

Biliary imaging

Introduction to MRCP

Magnetic resonance cholangiopancreatography (MRCP) overview

• Magnetic resonance cholangiopancreatography (MRCP) is an abdominal MRI acquired with heavily T2-weighted sequences that increase the contrast between T2 hyperintense stationary fluid in the biliary tract and surrounding structures.

• Fast spin echo sequences are most commonly used for MRCP acquisition.

Various techniques can be employed to optimize imaging including breath-hold sequences and respiratory-triggered sequences.

• Heavily T2-weighted sequences primarily image the biliary tree.

• Sequences with intermediate T2 (TE 80–100 ms) are best suited for visualization of the biliary ductal system *and* surrounding tissue, in particular to evaluate extraluminal

structures.

• Advantages of MRCP over ERCP include:

MRCP has the ability to see extra-luminal findings.

MRCP can visualize excluded (obstructed) ducts.

MRCP is non-invasive.

• Disadvantages of MRCP compared to ERCP include:

MRCP does not allow for concurrent therapeutic intervention.

MRCP does not actively distend the biliary ductal system with contrast.

MRCP has worse spatial resolution compared to ERCP.

• Contrast-enhanced MRCP can also be performed with fat-saturated T1-weighted imaging after injection of gadolinium contrast agents that have biliary excretion, such as gadoxetic acid disodium (Eovist, Bayer Healthcare, Germany) and gadobenate dimeglumine (Multihance, Bracco Diagnostics). These agents shorten T1 relaxation, resulting in T1 hyperintense biliary fluid, but require a 20–45 minute delay prior to

imaging to allow time for biliary excretion.

Choledochal cysts

Overview and Todani classification of choledochal cysts

• Choledochal cysts are thought to represent a heterogeneous group of diseases with a common end pathway of intrahepatic or extrahepatic biliary ductal dilation.

• The Todani system divides the cysts into types I–V based on their number, distribution, and morphology.

• Most choledochal cysts are diagnosed in childhood, but less commonly may be a new diagnosis for an adult.

Clinically, choledochal cysts can present with nonspecific

abdominal pain or may be found incidentally.

• Choledochal cysts are often resected due to increased cholangiocarcinoma risk, which

can be as high as 25%.

• In contrast to biliary hamartomas, choledochal cysts *do* communicate with the biliary tree.

Type I choledochal cyst

• A type I choledochal cyst, representing extrahepatic dilation of the common bile duct, is the most common type of extrahepatic cyst.

Caroli disease (Type V choledochal cysts)

• Caroli disease represents saccular dilation of the intrahepatic bile ducts, which may be segmental or diffuse.

Caroli disease may be associated with polycystic kidneys.

• Caroli *syndrome* is Caroli disease plus hepatic fibrosis.

• The *central-dot* sign describes the small branches of the portal vein and hepatic artery bridging the dilated bile ducts, which look like a central dot on contrast-enhanced CT.

Biliary anatomical variants

Low insertion of cystic duct

• With a low insertion of the cystic duct, the surgeon may misidentify the common duct as the cystic duct if the patient undergoes cholecystectomy, possibly leading to inadvertent common duct ligation.

Aberrant right posterior duct

• An aberrant right posterior duct is only important if the patient is a right hepatic lobe liver donor, as the two right hepatic ducts need to be anastomosed separately in the recipient.

Gallbladder infection and inflammation

• Gallbladder pathology is further discussed in the ultrasound section.

Acute cholecystitis

• Cholecystitis is inflammation and localized infection secondary to obstruction of the gallbladder neck or cystic duct.

• Calculous cholecystitis is caused by a gallstone which blocks the cystic duct.

• Acalculous cholecystitis is a functional obstruction of the cystic duct without a culprit stone.

It is typically seen in ICU patients.

• Acute cholecystitis is typically diagnosed by ultrasound, but similar criteria can be applied to CT:

Gallbladder wall thickening >3 mm (a nonspecific finding).

Pericholecystic fluid or inflammatory changes in the pericholecystic fat.

Gallbladder hyperemia.

Gallbladder calculi (although not all gallstones are radiopaque; ultrasound is more sensitive).

• Complications of acute cholecystitis include gangrenous cholecystitis, gallbladder perforation, and emphysematous cholecystitis.

• **Gangrenous cholecystitis** is due to increased intraluminal pressure, leading to gallbladder wall ischemia.

On imaging, the gallbladder wall thickening may be notably

asymmetric and intraluminal membranes may be present. Due to the increased risk of perforation, treatment is emergent cholecystectomy or cholecystostomy.

• **Acute gallbladder perforation** has a very high mortality due to generalized bile peritonitis.

Subacute perforation may lead to a pericholecystic abscess and chronic perforation may cause a cholecystoenteric fistula.

• **Emphysematous cholecystitis** is a severe complication of acute cholecystitis caused by gas-forming bacteria. Gas may be present either within the lumen or the wall of the gallbladder. The typical patient susceptible to emphysematous cholecystitis is an elderly diabetic.

Treatment of emphysematous cholecystitis is most often emergent cholecystectomy or cholecystostomy, although

treatment can be conservative in patients with a very high surgical risk.

Porcelain gallbladder

• Porcelain gallbladder describes a peripherally calcified gallbladder wall, thought to be a sequela of chronic cholecystitis.

• Porcelain gallbladder is associated with a (somewhat controversial) increased risk of gallbladder carcinoma. Typically, a porcelain gallbladder is an indication for

non-emergent cholecystectomy.

Bile duct infection and inflammation

Ascending cholangitis

• Obstruction of the biliary tree, most commonly due to choledocholithiasis, may cause ascending cholangitis, which presents with the clinical triad of fever, abdominal pain,

and jaundice (Charcot's triad).

• **On imaging**, the key finding is hyperenhancement and thickening of the walls of the bile ducts, often with a common bile duct stone present.

On ultrasound, debris within the biliary system may be apparent.

• Initial treatment is antibiotics and fluid resuscitation. Endoscopic biliary intervention may be necessary if the patient does not respond to conservative management.

Primary sclerosing cholangitis (PSC)

• Primary sclerosing cholangitis (PSC) is idiopathic inflammation and destruction of bile ducts.

• PSC is associated with ulcerative colitis (UC) and is more common in males.

Most (75%) patients with PSC have UC, while only a few (4–5%) of patients with UC have PSC.

• Biliary imaging shows a characteristic beaded, irregular appearance of the common bile duct and intrahepatic bile ducts.

• PSC appears similar to HIV-cholangiopathy, although cholangitis in HIV patients is more commonly associated with papillary stenosis.

• Long-term complications of PSC include cirrhosis, cholangiocarcinoma, and recurrent biliary infections. Cross-sectional imaging is better at evaluating for these complications compared to ERCP.

Primary biliary cirrhosis (PBC)

• Primary biliary cirrhosis (PBC) is inflammation and destruction of smaller bile ducts compared to PSC.

PBC affects middle-aged women and often initially presents with pruritus.

• Similar to PSC, chronic PBC can lead to hepatic cirrhosis.

AIDS cholangitis (AIDS cholangiopathy)

• Patients with acquired immunodeficiency syndrome are susceptible to biliary infection with *Cryptosporidium* and CMV, which clinically present with right upper quadrant

pain, fever, and elevated LFTs.

• The imaging of AIDS cholangitis appears nearly identical to primary sclerosing cholangitis, with multiple strictures and a beaded appearance of the bile ducts.

A distinguishing feature of AIDS cholangitis is papillary stenosis, which is not typically seen in PSC.

Recurrent pyogenic cholangitis (oriental cholangiohepatitis)

• Recurrent pyogenic cholangitis, also known as oriental cholangiohepatitis, is thought to be caused by the parasite *Clonorchis sinensis*, which leads to pigment stone formation, biliary stasis, and cholangitis. Nutritional deficiency may also play a role.

The disease typically affects patients indigenous to Southeast Asia.

Clinically, patients present with recurrent jaundice and fevers.

• Recurrent pyogenic cholangitis features an imaging triad of:

1) Pneumobilia.

2) Lamellated bile duct filling defects.

3) Intrahepatic and extrahepatic bile duct dilation and strictures.

• Patients with recurrent pyogenic cholangitis have an increased risk of cholangiocarcinoma.

Biliary neoplasia

Biliary cystadenoma

• Biliary cystadenoma is a benign cystic neoplasm, occurring predominantly in middle-aged women.

Biliary cystadenoma may be quite large at presentation and cause nonspecific symptoms such as abdominal pain, nausea, vomiting, and obstructive jaundice.

• Biliary cystadenoma does not communicate with the biliary system.

• On imaging, biliary cystadenoma appears as a large, multiloculated, cystic mass.

The presence of septations distinguishes cystadenoma from a simple cyst.

The septations may mimic an echinococcal cyst.

In contrast to hepatic abscess or necrotic metastasis,

a thick enhancing wall is not a feature of cystadenoma.

• Although benign, cystadenoma may uncommonly recur after resection.

• Malignant degeneration to biliary cystadenocarcinoma has been reported but is rare.

The presence of a large solid component or thick calcification should raise concern for cystadenocarcinoma.

Cholangiocarcinoma

• Cholangiocarcinoma is a highly malignant tumor of the biliary ductal epithelium.

• A hilar tumor (at the confluence of the right and left intrahepatic biliary ducts), known as a **Klatskin tumor**, is the most common form of cholangiocarcinoma. In contrast,

peripheral cholangiocarcinoma is rare.

• Cholangiocarcinoma tends to obstruct bile ducts and cause intrahepatic ductal dilation.

Eventually, the obstruction may lead to lobar atrophy.

• Risk factors for development of cholangiocarcinoma include:

Choledochal cyst(s).

Primary sclerosing cholangitis.

Familial adenomatous polyposis syndrome.

Clonorchis sinensis infection.

Thorium dioxide (alpha-emitter contrast agent), not used since the 1950s. Thorium dioxide is also

associated with angiosarcoma and HCC.

• On cross-sectional imaging, cholangiocarcinoma typically presents as an intrahepatic mass at the confluence of the central bile ducts (Klatskin tumor), with resultant bile

duct dilation and capsular retraction. Tumor fingers often extend into the bile ducts.

Gallbladder carcinoma

• Gallbladder carcinoma is rare and is usually due to chronic gallbladder inflammation.

• Gallstones and concomitant chronic cholecystitis are typically present.

Porcelain gallbladder, a result of chronic cholecystitis, is thought to be a risk factor for gallbladder cancer, although this is controversial.

• Gallbladder carcinoma most commonly presents as a scirrhous infiltrating mass that invades through the gallbladder wall into the liver.

Less commonly, gallbladder carcinoma may appear as a polypoid mass.

Very rarely it can present as mural

thickening.

• Tumor spread is via direct extension into the liver, although lymphatic and hematogenous metastases are also common.

• Prognosis is generally poor, although small polypoid lesions may undergo curative resection.

Gallbladder metastasis

• Melanoma has a propensity to metastasize to the gallbladder.

Pancreas

Solid pancreatic epithelial neoplasms

Adenocarcinoma (ductal adenocarcinoma)

Pancreatic ductal adenocarcinoma makes up 80–90% of all pancreatic tumors.

It is typically seen in patients over age 60, with a slight male predominance.

Risk factors include smoking, alcohol, and chronic pancreatitis.

• A pancreatic-mass CT includes unenhanced, late arterial phase, and portal venous phase images.

The late arterial phase (pancreatic parenchymal phase) has the greatest conspicuity for detecting the hypoenhancing tumor against the background enhancing pancreas.

• The most common location of ductal adenocarcinoma is the pancreatic head.

• The classic appearance is a hypodense (CT), T1 hypointense (MR), ill-defined, hypovascular mass causing ductal obstruction and atrophy of the pancreatic tail. The *double duct* sign describes dilation of both the pancreatic duct and the common bile duct.

• Since pancreatic adenocarcinoma is almost always associated with a dilated pancreatic duct, an alternative diagnosis should be strongly considered if there is a pancreatic mass with no ductal dilation, such as:

Autoimmune pancreatitis.

Groove pancreatitis.

Cystic pancreatic tumor.

Neuroendocrine tumor.

Duodenal gastrointestinal stromal tumor (GIST).

Peripancreatic lymph node.

Pancreatic metastasis (e.g., renal cell, thyroid, or melanoma).

Lymphoma.

• Conversely, if the *double duct* sign is present but no mass is visible, one should still be suspicious for pancreatic adenocarcinoma. Approximately 10% of cases will be

isoattenuating relative to pancreas in the pancreatic parenchymal (late arterial) phase and thus extremely difficult to directly detect.

• Most cancers present at an advanced, unresectable stage. Unresectable tumors show encasement (>180° circumference) of the SMA, extensive venous invasion, or evidence of metastasis.

• For lower-stage tumors, complete surgical resection is the only chance for cure.

A resectable tumor features no evidence of celiac, SMA, or portal venous invasion.

Limited extension to the duodenum, distal stomach, or CBD does not preclude resection, as these structures are resected during the Whipple procedure.

Limited venous extension may be resectable.

Acinar cell carcinoma

• Acinar cell carcinoma is a rare, aggressive variant of pancreatic adenocarcinoma, exclusively seen in elderly males.

• The malignant cells produce a large amount of lipase to cause the clinical triad of lipase hypersecretion syndrome: Subcutaneous fat necrosis; bone infarcts causing polyarthralgias; and eosinophilia.

Cystic pancreatic epithelial neoplasms

Serous cystadenoma

• Serous cystadenoma is a benign tumor that occurs in elderly women and has been nicknamed the *grandmother* tumor.

• It consists of many small cysts (>6 cysts that are <2 cm) that may have a solid appearance on CT due to apposition of

many cyst walls.

MRI is useful to show the cystic nature of the lesion.

• Serous cystadenoma is hypervascular, unique among cystic pancreatic tumors.

• Unlike adenocarcinoma, serous

cystadenoma does not cause pancreatic duct dilation or tail atrophy.

• A classic imaging feature is central stellate calcification.

Mucinous cystic neoplasm

• Mucinous cystic neoplasm affects middle-aged women and has

therefore been nicknamed the *mother* tumor.

• Mucinous cystic neoplasm is benign, but does have malignant potential.

Treatment is typically resection due to

malignant potential.

• The tumor consists of a single or a few large cysts (<6 cysts that are >2 cm) and typically occurs in the pancreatic body and tail.

• Mucinous cystic neoplasm has a capsule.

The only other pancreatic tumor with a capsule is SPEN (below).

Solid and papillary epithelial neoplasm (SPEN)

• Solid and papillary epithelial neoplasm (SPEN) occurs in young women and children and is nicknamed the *daughter* tumor. It may be a rare cause of abdominal pain.

• It has a low malignant potential and is typically resected.

• On imaging, SPEN appears as a large mass with heterogeneous solid and cystic areas.

Hemorrhage is typical. SPEN features a capsule, as does mucinous cystic neoplasm.

Intraductal papillary mucinous neoplasm (IPMN)

• Intraductal papillary mucinous neoplasm (IPMN) occurs most commonly in elderly males and is nicknamed the *grandfather* tumor, although these tumors exhibit the greatest age and sex variability of the cystic pancreatic neoplasms.

• IPMN features a spectrum of biological behavior from benign to indolent to aggressive carcinoma.

IPMNs may arise from the main pancreatic duct or a sidebranch. The main duct IPMNs have greater malignant potential.

• The classic appearance on endoscopy is a *fish-mouth* papilla pouring out mucin.

Crosssectional imaging shows a cystic intrapancreatic lesion in contiguity with the duct or sidebranch.

Any nodular or enhancing component should raise concern for malignancy.

• The recommended imaging follow-up and criteria for resectability are controversial.

Current guidelines published in 2006 recommend following simple pancreatic cysts <1 cm annually by imaging (typically MR).

However, up to 40% of elderly males have a pancreatic cyst, suggesting that they may be an acquired condition of aging rather than a premalignancy.

• In general, a suspected IPMN is resected if it is >3 cm in size, if there is a mural nodule, or if there is associated dilation of the pancreatic duct to >10 mm.

Pancreatic endocrine neoplasms

Overview

• Pancreatic neuroendocrine tumors may be hyperfunctioning or non-hyperfunctioning.

• Hyperfunctioning tumors come to clinical attention due to symptoms of endocrine excess.

• Non-hyperfunctioning tumors tend to be larger at diagnosis. These tumors may undergo cystic change and should be considered in the differential of a cystic pancreatic neoplasm. There is often central necrosis and calcification in these large tumors as well.

• Pancreatic endocrine tumors tend to be hypervascular and are best seen in the late arterial phase.

Most are solid unless large. A hypervascular liver mass with an

associated pancreatic mass is most likely a metastatic pancreatic endocrine neoplasm.

Insulinoma

• Insulinoma is the most common pancreatic endocrine tumor. Due to symptoms of hypoglycemia, insulinomas tend to present early and have the best prognosis of all

neuroendocrine tumors, with only 10% demonstrating malignant behavior.

• The Whipple triad describes the clinical symptoms of insulinoma: Hypoglycemia, clinical symptoms of hypoglycemia, and alleviation of symptoms after administration of glucose.

Gastrinoma

• Gastrinoma causes hypersecretion of gastric acid resulting in Zollinger–Ellison syndrome.

Gastrinoma is the second most common pancreatic endocrine tumor.

Gastrinoma is associated with multiple endocrine neoplasia (MEN) type 1.

When associated with MEN-1, gastrinomas tend to be multiple and located in the duodenum rather than the pancreas.

• The gastrinoma triangle describes the typical location of gastrinomas, in an area bounded by the junction of the cystic duct and CBD, the duodenum inferiorly, and the neck/body of the pancreas medially.

• High gastrin levels may cause formation of carcinoid tumors in the stomach, which may regress after the gastrinoma is resected.

Other pancreatic endocrine tumors

• Glucagonoma is the third most common pancreatic endocrine tumor.

Prognosis is poor.

VIPoma and somatostatinoma are very rare and also have poor prognoses.

Congenital pancreatic anomalies

Normal ductal anatomy

• Normally , the main pancreatic duct drains to the major papilla (the ampulla of Vater) through the duct of Wirsung, while the duct of Santorini drains to the minor papilla.

The sphincter of Oddi is a circular band of muscle encircling the ampulla of Vater.

• Mnemonic for normal anatomy: Santorini is superior and drains to small (minor) papilla.

• The following anatomy is *always* constant, regardless of whether an anomaly is present:

1) The common bile duct always drains to the major papilla where it meets the duct of Wirsung.

2) The main pancreatic duct always drains the pancreatic tail.

3) The duct of Santorini always drains to the minor papilla.

Pancreas divisum

• Pancreas divisum is the most common congenital pancreatic anomaly. It is caused by failure of fusion of ventral and dorsal pancreatic ducts.

The ventral duct (Wirsung) only drains a portion of the pancreas while the majority of the pancreatic exocrine gland output is drained through the smaller duct of Santorini into the minor papilla.

• Pancreas divisum may be a cause of pancreatitis due to obstruction at the minor papilla from a Santorinicele.

A Santorinicele is a focal dilation of the terminal duct of Santorini.

• The *crossing* sign describes the CBD crossing over the main duct to join the duct of

Wirsung.

Annular pancreas

• Annular pancreas is a rare congenital anomaly where a portion of the pancreas wraps completely around the duodenum, secondary to incomplete rotation of the ventral pancreatic bud.

• In an adult, annular pancreas can cause pancreatitis, peptic ulcer disease, and duodenal obstruction. In a neonate, it can cause duodenal obstruction and is in the differential for the *double bubble* sign.

Common channel syndrome/pancreaticobiliary maljunction

• Normally the common bile duct and duct of Wirsung both drain to the major papilla, where there is usually a thin septum separating these two systems.

• In common channel syndrome, also known as pancreaticobiliary maljunction, the distal CBD and pancreatic duct are missing the septum, allowing reflux between the

two systems.

• Common channel syndrome may be in the spectrum of choledochal cyst disease with the common channel representing a very mild form of choledochocele.

Common channel syndrome may predispose to cholangiocarcinoma, but this is rare and controversial.

Systemic diseases that affect the pancreas

Pancreatic manifestations of von Hippel–Lindau

• von Hippel–Lindau is an inherited multisystemic disease with increased risk of multiple malignancies and formation of cysts in various organs including the pancreas.

• Pancreatic neoplasms seen in von Hippel–Lindau include serous cystadenoma and pancreatic neuroendocrine tumors.

Cystic fibrosis (CF)

• Cystic fibrosis (CF) is the most common cause of childhood pancreatic atrophy.

• CF can cause either fatty atrophy of the pancreas or pancreatic cystosis (diffuse replacement of the pancreas with innumerable cysts).

Schwachman–Diamond

• Schwachman–Diamond is a rare inherited disorder characterized by diffuse fatty replacement of the pancreas, resultant pancreatic exocrine insufficiency, neutropenia,

and bone dysplasia.

• Schwachman–Diamond is the second-most common cause of childhood pancreatic atrophy.

Obesity and exogenous steroid use

• Both obesity and steroids can cause fatty atrophy of the pancreas.

Miscellaneous pancreatic lesions

Intrapancreatic accessory spleen

• Intrapancreatic accessory spleen is a benign mimic of a hypervascular pancreatic neoplasm.

• On imaging, an intrapancreatic spleen typically is a small (1–3 cm), well-defined mass usually found in the pancreatic tail. It follows the density, signal intensity, and enhancement of the spleen on all CT and MRI sequences.

• MRI is usually diagnostic. Either technetium-99m sulfur colloid or technetium-99m

RBC scintigraphy can confirm the diagnosis in ambiguous cases.

Pancreatitis

• Pancreatitis is inflammation of the pancreas, which may be due to a wide variety of etiologies that share a final common pathway of premature activation of pancreatic enzymes and resultant autodigestion of pancreatic parenchyma.

• Pancreatitis may range in severity from mild self-limited disease to necrotizing pancreatitis resulting in multi-organ failure and death.

CT protocol and role of imaging

• Imaging of pancreatitis is optimally performed in the pancreatic parenchymal phase (late arterial; ~40 seconds after contrast injection), which is the most sensitive timing to detect subtle areas of decreased enhancement suggestive of necrosis.

• CT is key for pancreatitis imaging. In addition to often identifying an etiology of the pancreatitis, CT can grade severity, detect complications, and guide possible percutaneous interventions.

• CT imaging is *not* indicated in patients with clinical diagnosis of mild acute pancreatitis, especially if they are improving. CT imaging may be negative or show a mildly edematous pancreas in these cases.

Acute pancreatitis

• Acute pancreatitis is most commonly caused by alcohol or an

obstructing gallstone.

• Acute pancreatitis can be classified

either with the Balthazar grading system or by the CT severity index.

• **Balthazar** grading system:

A: Normal-appearing pancreas

B: Focal or diffuse pancreatic enlargement

C: Mild peripancreatic inflammatory changes

D: Single fluid collection

E: Two or more fluid collections 0% mortality, 4% morbidity for grades A, B, and C.

14% mortality, 54% morbidity for grades D and E (a fluid collection is a poor prognostic indicator).

• **CT severity index (CTSI)** integrates the Balthazar grading system with the degree of

necrosis:

Assigns 0–4 points for Balthazar A–E, with 0 points for Balthazar A and 4 points for Balthazar E.

Adds 0–6 points for necrosis to create a total score from 0-10.

0 points: 0% necrosis

2 points: <30% necrosis

4 points: 30–50% necrosis

6 points: >50% necrosis

CTSI 0–3: 3% mortality, 8% morbidity

CTSI 7–10: 17% mortality, 92% morbidity

• Pancreatic and peripancreatic complications:

Pancreatic necrosis: On imaging, pancreatic necrosis appears as a focal or diffuse area of nonenhancing pancreatic parenchyma.

Evaluation of necrosis is best performed 48–72 hours after onset of acute pancreatitis.

Late arterial phase imaging has the highest sensitivity for detecting pancreatic necrosis.

Patients with pancreatic necrosis are at increased risk for infection and severe morbidity.

Fluid collections: Peripancreatic fluid may resolve or may evolve either into peripancreatic abscess or pseudocyst.

Pseudocyst: A pancreatic pseudocyst is a collection of pancreatic enzymes and fluid enclosed by a fibrous wall lacking an epithelial lining.

The fibrous wall usually takes about 4–6 weeks to mature.

Pancreatic abscess: Pancreatic abscess is a purulent collection featuring thicker, more irregular walls compared to a pseudocyst. Gas locules may be present within the abscess.

• Extrapancreatic complications:

Extrapancreatic pseudocyst may occur nearly anywhere below the diaphragm and should always be considered in the differential of a cystic structure in a patient with history of pancreatitis.

In particular, an intrasplenic pseudocyst may lead to intrasplenic hemorrhage.

Perihilar renal inflammation, which may lead to venous compression or thrombosis.

Bowel involvement, especially of the transverse colon.

• Secondary inflammation of adjacent vessels can cause vascular complications:

Arterial bleeding, most commonly due to erosion into the splenic artery.

Pseudoaneurysm, most commonly of the splenic artery.

Venous thrombosis, most commonly splenic vein thrombosis, which may lead to portal hypertension.

Chronic pancreatitis

• Chronic pancreatitis, most commonly from long-term alcohol abuse, causes irreversible pancreatic damage.

A much less common cause of chronic pancreatitis is pancreas divisum.

• Calcifications in the distribution of the pancreatic duct are pathognomonic for chronic pancreatitis.

Autoimmune pancreatitis

Segmental autoimmune pancreatitis: Contrast-enhanced axial CT (left image) shows a segmental region of low attenuation enlargement of the pancreatic tail and body (arrows), with loss of the normal ductal architecture.

T1-weighted unenhanced MRI (right image) shows a corresponding segmental loss of the normal T1-hyperintense pancreatic signal, with effacement of the pancreatic duct in the affected body and tail.

The differential diagnosis for this appearance would include pancreatic lymphoma, less likely pancreatic adenocarcinoma as there is no ductal dilation.

Case courtesy Cheryl Sadow, MD, Brigham and Women's Hospital.

• Autoimmune pancreatitis is caused by an inflammatory lymphoplasmacytic infiltrate.

It is associated with Sjögren syndrome and causes elevated serum IgG-4 levels.

• The typical imaging appearance of autoimmune pancreatitis is diffuse “sausage shaped” enlargement of the entire pancreas; however, a focal or segmental form may

mimic a pancreatic mass.

• Treatment is with steroids, which can lead to a complete resolution.

Groove pancreatitis

• Groove pancreatitis is an uncommon form of focal pancreatitis of the groove between the head of the pancreas, duodenum, and common bile duct.

Groove pancreatitis usually affects young men who are heavy drinkers.

• The histopathologic hallmark is fibrosis in the pancreaticoduodenal groove.

Chronic inflammation of the duodenum can cause varying levels of duodenal stenosis or cystic change of the duodenal wall.

Imaging reflects these findings, with duodenal thickening and cystic change often apparent.

The cystic change is best appreciated on MR.

• The main differential consideration is adenocarcinoma of the head of the pancreas.

Spleen

Congenital splenic variations and anomalies

Splenule (accessory spleen)

• Also called an accessory spleen, a splenule is a focus of normal splenic tissue separate from the main body of the spleen, due to embryologic failure of fusion of the splenic anlage.

The most common location is the splenic hilum.

• Although usually an incidental finding, the presence of a splenule does have significance in certain clinical settings. For instance, splenectomy for consumptive thrombocytopenia may not be curative if there is sufficient unresected accessory

splenic tissue present.

A splenule may be mistaken for a lymph node or mass when

in an unusual location.

As previously discussed, an intrapancreatic splenule may be mistaken for a hypervascular pancreatic mass.

• A splenule should follow splenic tissue on all MRI sequences. If in doubt, a Tc-99m sulfur colloid scan or a heat-damaged Tc-99m RBC scan can be confirmatory.

Polysplenia syndrome

• Polysplenia syndrome is a spectrum of anatomic disorders characterized by some degree of visceral heterotaxia in addition to multiple discrete foci of splenic tissue.

Multiple spleens may be on the right or left, but are always on the same side as the stomach.

• Polysplenia is usually associated with severe congenital cardiac anomalies.

Most patients die in early childhood, but a few may have only minor cardiac defects and may be incidentally discovered as adults.

• Polysplenia is associated with venous anomalies including interruption of the IVC with azygos or hemiazygos continuation. A less common association is a preduodenal portal vein.

Wandering spleen

• A wandering spleen is a normal spleen with abnormal laxity or absence of its fixed ligamentous attachments.

• Wandering spleen may present clinically as an abdominal mass or may cause acute abdominal pain secondary to torsion.

Benign non-cystic splenic lesions

Hemangioma

• Hemangioma is the most common benign splenic neoplasm. Hemangioma may be solitary or multiple, and lesions tend to be small.

• Splenic hemangiomas are associated with **Kasabach–Merritt** syndrome (anemia, thrombocytopenia, and consumptive coagulopathy) and **Klippel–Trenaunay–Weber** syndrome (cutaneous hemangiomas, varicose veins, and extremity hypertrophy).

These visceral hemangiomatosis syndromes are usually associated with phleboliths.

• **On CT**, hemangiomas are typically iso- or hypoattenuating pre-contrast and hyperenhancing. **On MR**, hemangiomas are typically hyperintense on T2-weighted images and may enhance peripherally or homogeneously.

However, the classic pattern of discontinuous nodular enhancement seen in hepatic hemangiomas is uncommon.

• Nuclear medicine scintigraphy with Tc-99m labeled red blood cells would show increased activity within the lesion on delayed images.

In contrast, Tc-99m sulfur colloid scanning may show either increased or decreased activity.

Hamartoma

• Splenic hamartoma is a rare, benign lesion composed of malformed red pulp elements.

It may be associated with tuberous sclerosis.

• Splenic hamartoma is typically a well-circumscribed, iso- or hypoattenuating mass on unenhanced **CT** that enhances heterogeneously after contrast administration.

On **MR**, a hamartoma is iso- to slightly hyperintense on T2-weighted images, featuring heterogeneous early enhancement and relatively homogeneous delayed enhancement.

Benign cystic splenic lesions

Congenital true (epithelial) cyst

• A congenital true cyst is defined as having an epithelial lining. Interestingly, a splenic epithelial cyst may cause elevation of tumor markers including CA19-9, CA125, and CEA, despite its completely benign nature.

• Unlike a post-traumatic pseudocyst, a true cyst may have septations, but mural calcification is uncommon.

Post-traumatic pseudocyst

• A post-traumatic pseudocyst is the end result of evolution of a splenic hematoma.

• Unlike a true (epithelial) splenic cyst, the periphery of a pseudocyst is not cellular but made of fibrotic tissue.

• On imaging, a post-traumatic pseudocyst appears as a well-circumscribed, fluiddensity lesion, with no peripheral enhancement.

• In contrast to a true cyst, septations are uncommon but there may be mural calcification.

Intrasplenic pancreatic pseudocyst

• A post-pancreatitis pseudocyst involving the tail of the pancreas may extend into the spleen.

There is almost always a history of pancreatitis.

• Unlike a true congenital cyst, an epithelial lining is lacking and histology more closely resembles a post-traumatic pseudocyst.

• Splenic rupture has been reported in some cases of intrasplenic post-pancreatitis pseudocysts.

Lymphangioma

• Splenic lymphangioma is a rare, benign neoplasm usually diagnosed in childhood, which may be solitary or multiple.

• Lymphangioma features a classic imaging appearance of a multilocular cystic structure with thin septations. Post-contrast images may show septal enhancement.

Inflammatory splenic lesions

Sarcoidosis

• Sarcoidosis is a systemic disease of unknown etiology characterized histologically by multiple nodules composed of noncaseating granulomas.

• When sarcoidosis involves the spleen, splenomegaly is the most common presentation, often associated with hepatomegaly and lymphadenopathy.

• Less commonly, sarcoidosis may involve the spleen in a multinodular pattern with numerous hypoattenuating 1–3 cm lesions demonstrating essentially no enhancement.

These nodules are formed by coalescent sarcoid granulomas and have low signal on all MRI sequences.

Sarcoid nodules are most conspicuous on T2-weighted images and early-phase post-contrast T1-weighted images.

On the post-contrast images the nonenhancing nodules will stand out against the avidly enhancing splenic parenchyma.

• Imaging appearance is generally indistinguishable from splenic lymphoma.

Inflammatory pseudotumor

• Splenic inflammatory pseudotumor is a rare focal collection of immune cells and associated inflammatory exudate, of unclear etiology.

Patients often have constitutional symptoms including fever and malaise.

• Inflammatory pseudotumor has a variable and nonspecific imaging appearance, but a typical presentation is of a well-circumscribed, heterogeneously enhancing mass.

Splenic infection

Pyogenic abscess

• Splenic bacterial abscesses are uncommon and usually seen in immunocompromised patients.

A solitary abscess is much more likely to be bacterial.

In contrast, multifocal small abscesses are more likely to be fungal.

• **On CT**, a bacterial abscess usually has an irregular, enhancing wall.

Gas is not usually seen, but is highly specific for a bacterial abscess when present.

• A characteristic ultrasound finding is the *wheel within a wheel* or *bull's-eye* appearance, which describes concentric hyperechoic and hypoechoic rings surrounding the abscess.

• Treatment is CT- or ultrasound-guided percutaneous drainage in addition to antibiotics.

Fungal abscess

• Splenic fungal abscesses are typically multiple and small, usually <1 cm in size. Almost all patients with splenic fungal abscesses are immunocompromised.

• The most common causative agents include *Candida*, *Aspergillus*, and *Cryptococcus*, which all appear as multiple tiny hypoattenuating foci on CT.

• Splenic *Pneumocystis jiroveci* (formerly known as *Pneumocystis carinii*) infection is rare, almost always seen in advanced AIDS, and has a classic appearance of multiple

calcified splenic lesions.

Echinococcal cyst

• Splenic involvement of *Echinococcus granulosus* infection is unusual, seen in only 1 to 3% of echinococcal infections, and is almost always associated with infection of other organs as well.

• An echinococcal cyst is a true cyst with a cellular lining. As with echinococcal abscess elsewhere, characteristic imaging findings are a cystic lesion with internal undulating membrane and daughter cysts.

Malignant splenic lesions

Splenic lymphoma

• Splenic lymphoma is the most common splenic malignancy.

• Primary splenic lymphoma is rare, accounting for less than 1% of all cases of lymphoma, and usually presents as a solitary hypovascular mass.

In contrast to splenic hamartoma or metastatic disease, primary splenic lymphoma may extend beyond the splenic capsule and involve adjacent organs.

• Secondary splenic involvement of systemic lymphoma is much more common.

Four imaging presentations have been described, depending on the size of the lymphomatous masses:

Miliary masses, in which discrete tiny masses may be difficult to see.

Multiple small to moderate sized masses.

One large mass.

Splenomegaly without discrete mass.

• Conspicuity is highest on post-contrast T1-weighted images, where the involved portion of the spleen tends to be *hypoenhancing*.

• On ultrasound, lymphoma may appear cystic but color Doppler shows internal flow.

Splenic metastasis

• Splenic metastases may be solitary or multiple, cystic or solid. However, metastases to the spleen are rare, occurring in 2–9% of cancer patients.

Most patients with splenic metastases already have known widespread disease.

Isolated splenic metastases are seen in only 5% of patients with metastatic involvement of the spleen.

• Several theories for the low rate of splenic metastases have been proposed, which is thought to be due to the antineoplastic properties of lymphoid-rich splenic tissue and lack of afferent lymphatics to bring tumor cells into the spleen.

• The most common primary tumors known to metastasize to the spleen include breast, lung, ovarian, and melanoma. Ovarian cancer and melanoma typically cause cystic metastasis.

• Calcification is rare unless the primary is a mucinous adenocarcinoma.

Angiosarcoma

• Angiosarcoma is a rare, extremely aggressive malignancy, with 20% 6-month survival.

• Unlike hepatic angiosarcoma, the association with Thorotrast, vinyl chloride, and arsenic has not been well established.

• Angiosarcoma usually presents as enlarged, heterogeneous mass that may completely replace the normal spleen. Enhancement is variable and heterogeneous.

Miscellaneous splenic lesions

Splenic infarct

• Splenic infarcts are most commonly due to emboli (in older patients) and thrombosis (in younger patients with

hematologic disease).

• Splenic infarct classically manifests as a wedge-shaped peripheral region of nonenhancement, but a more heterogeneous, mass-like appearance can also be seen.

• On MR, the affected regions may

be T1 hyperintense if acute and hemorrhagic, while chronic infarcts are T1 hypointense and T2 hyperintense.

• Lack of enhancement of the entire

spleen should raise the concern for complete infarction, possibly due to a wandering spleen with torsion.

Gamna-Gandy bodies

• Gamna–Gandy bodies are multiple tiny foci of hemosiderin deposition resulting from portal hypertension.

• Usually, other signs of portal hypertension and cirrhosis are apparent on imaging, such as splenomegaly, varices, recanalized umbilical vein, ascites, and nodular liver contour.

• The hemosiderin deposits demonstrate low signal on all sequences.

On in- and out of-phase gradient-echo sequences, the longer TE of the in-phase images demonstrate blooming (relatively decreased signal on in-phase images), due to longer dephasing time and exaggeration of T2* effect.

Gaucher disease

• Gaucher disease is an autosomal recessive deficiency of glucocerebrosidase, leading to accumulation of glucocerebrosides in the reticuloendothelial system.

• Splenic manifestations of Gaucher disease include splenomegaly (almost always) and multiple splenic nodules (seen in one third of Gaucher patients).

• Associated bony findings seen in Gaucher disease include the characteristic Erlenmeyer flask deformity of the distal femurs, femoral head avascular necrosis, and H-shaped vertebral bodies from endplate avascular necrosis.

Splenic trauma

Overview of splenic trauma

• The spleen is the most commonly injured abdominal organ from blunt trauma.

• Splenectomy dramatically increases the risk of subsequent sepsis, which has driven the trend towards splenic preservation and conservative management of splenic trauma.

• The spleen must be evaluated for injury in the portal venous phase, as physiologic heterogeneous enhancement in the arterial phase can both mask and mimic injury.

• A splenic hematoma is a focal collection of blood (hypoattenuating relative to enhanced spleen and hyperattenuating relative to unenhanced spleen), most commonly subcapsular.

Less commonly, a hematoma may be intraparenchymal, where it may assume an irregular shape.

• A splenic laceration can only be well seen on a contrast-enhanced study, where it appears as a linear or branching area of decreased attenuation.

• Active contrast extravasation due to vessel injury appears as an area of increased attenuation (initially iso-enhancing to the arterial blood pool), which further increases in size on delayed scanning due to ongoing bleeding.

• In contrast to active extravasation, pseudoaneurysm and arteriovenous fistula are contained vascular injuries that also initially appear as a well-circumscribed area of increased attenuation, but do not increase on delayed scanning.

A pseudoaneurysm is caused by injury to the intima and media of the arterial wall, and is essentially a rupture contained only by the adventitia.

There is a high chance of rupture without

treatment.

A traumatic arteriovenous fistula is indistinguishable from pseudoaneurysm by CT and is due to injury of an artery and the adjacent vein.

Splenic arteriography is the only way to differentiate pseudoaneurysm from arteriovenous fistula.

American Association for the Surgery of Trauma (AAST) grading scale

• The American Association for the Surgery of Trauma (AAST) splenic injury grading scale is based on the extent of injury seen at laparotomy, and is therefore limited in practicality as many splenic injuries are now managed conservatively.

• An additional limitation of the AAST grading scale is the exclusion of vascular injury.

MDCT-based splenic injury grading system.

• The most widely used CT grading system is the MDCT-based splenic injury grading system, which is similar to the AAST grading scale but incorporates vascular injury.

• MDCT grade I: Small (<1 cm) subcapsular hematoma, laceration, or parenchymal hematoma.

• MDCT grade IVB: Active intraperitoneal bleeding.

Esophagus

Anatomy

Pharynx

• **Nasopharynx:** Extends from the base of the skull to the soft palate.

• **Oropharynx:** Located behind the mouth and extends from the uvula to the hyoid bone.

• **Hypopharynx:** Extends from the hyoid bone to the cricopharyngeus muscle, which is located at the lower end of the cricoid cartilage.

Esophagus

• The cricopharyngeus muscle, located at C5–6, is the upper esophageal sphincter and demarcates the transition between the pharynx superiorly and the cervical esophagus.

• The esophagus extends from the neck to the gastroesophageal junction.

The distal esophagus passes through the diaphragmatic hiatus at approximately T10.

• The three anatomic rings of the distal esophagus are the A (muscular), B (mucosal), and C (diaphragmatic impression) rings.

Esophageal rings and webs

Esophageal web

• An esophageal web is a thin anterior infolding/indentation of the upper esophagus, which is usually asymptomatic but may be a cause of dysphagia.

There is a controversial association with anemia (Plummer–Vinson syndrome) and upper esophageal carcinoma.

Schatzki ring

• A Schatzki ring is a focal narrowing of the B (mucosal) ring of the distal esophagus, causing intermittent dysphagia.

• A true Schatzki ring requires clinical symptoms of dysphagia in addition to esophageal narrowing seen on imaging.

Asymptomatic narrowing of the B ring is referred to as a lower esophageal ring.

• An upper GI study is more sensitive than

endoscopy.

The key imaging feature is focal circumferential constriction near the gastroesophageal junction, almost always associated with a hiatal hernia.

On an upper GI study, most symptomatic rings do not allow passage of a 12 mm tablet.

• The differential of circumferential esophageal constriction includes:

Focal stricture.

Muscular esophageal ring above the GE junction (also known as an *A ring*).

Esophageal cancer.

Esophageal web (rarely circumferential, usually in cervical esophagus).

Esophagitis

Reflux (peptic) esophagitis

• Reflux (peptic) esophagitis is caused by exposure of the esophageal mucosa to acidic gastric secretions, which leads to distal ulcerations and eventual stricture.

• Peptic esophagitis is most commonly caused by gastroesophageal reflux, but is also seen in:

Zollinger–Ellison, due to increased acid production.

Scleroderma, due to gastroesophageal sphincter fibrosis and resultant incompetence.

• Reflux esophagitis appears as thickened distal esophageal folds.

• Chronic esophagitis and scarring develops after prolonged exposure to acid, which causes a smoothly tapered stricture above the GE junction.

Barrett esophagus

• An important long-term sequela of peptic esophagitis is Barrett esophagus, which is metaplasia of normal squamous epithelium to gastric-type adenomatous mucosa.

Barrett esophagus is a precursor lesion to esophageal carcinoma.

• Nearly 10% of patients with reflux esophagitis may have some adenomatous metaplasia.

• On imaging, Barrett demonstrates a featureless distal esophagus, with signs of active reflux esophagitis (mucosal granularity and superficial erosions) more proximally.

• Barrett esophagus is often associated with esophageal stricture, which is abnormally high in location compared to a peptic stricture.

Infectious esophagitis

• Although radiographic distinction between types of infections has been described, endoscopy and biopsy are typically performed in clinical practice.

• **Esophageal candidiasis** can present as a spectrum from scattered plaque-like lesions in mild disease to very shaggy esophagus in severe cases.

• **Herpes esophagitis** typically causes discrete small ulcerations scattered randomly throughout esophagus.

• **CMV/HIV esophagitis**

characteristically causes a large, flat, ovoid ulcer.

Medication esophagitis

• Medication-induced esophagitis typically causes an ulcer at the level of the aortic arch or distal esophagus, which are areas of relative narrowing that may predispose to temporary hold-ups in passage.

Crohn esophagitis

• Crohn esophagitis is very rare and is usually seen in the setting of severe disease in the small bowel and colon.

• Aphthous ulcers (discrete ulcers surrounded by mounds of edema) may become confluent.

Esophageal strictures

Peptic stricture

• As previously discussed, a peptic stricture is secondary to chronic reflux.

• Peptic strictures are located distally, usually just above the GE junction.

• A peptic stricture may be focal or involve a longer segment of esophagus.

• Fibrosis can cause esophageal shortening, leading to a hiatal hernia as the stomach is pulled into the thorax.

Barrett esophagus stricture

• A Barrett stricture typically occurs in the mid-esophagus, above the metaplastic adenomatous transition. Barrett strictures occur higher than peptic strictures because adenomatous tissue is acid-resistant and therefore unaffected by gastric secretions.

Malignant stricture (due to esophageal carcinoma)

• Key imaging finding is shouldered margins, which suggests circumferential luminal narrowing by a mass.

Caustic stricture/nasogastric (NG) tube stricture

• Both caustic strictures and strictures secondary to nasogastric tube placement are typically long, smooth, and narrow.

• Strictures develop 1–3 months after the caustic ingestion or NG tube placement.

• Caustic strictures are associated with an increased risk of cancer, with a long lag time of up to 20 years after the initial insult.

Caustic strictures are usually longer than peptic strictures.

Radiation stricture

• Radiation strictures are long, smooth and narrow, similar to caustic strictures.

However, in contrast to strictures from an NG tube, caustic ingestion, and reflux, radiation strictures usually spare the GE junction.

• It generally requires more than 50 Gy of radiation to cause an esophageal stricture.

• Acute radiation esophagitis occurs 1–4 weeks after radiation therapy.

Radiation strictures develop later, occurring 4–8 months after radiation.

Extrinsic compression from mediastinal adenopathy

• Cross-sectional imaging would best evaluate if extrinsic compression is suspected.

Evaluation of esophageal masses

• Masses arising from the mucosa, submucosa, and extrinsic to the esophagus produce characteristic effects on the esophagus, which are usually able to be seen on imaging.

Benign esophageal masses

Mesenchymal tumor

• Benign mesenchymal tumors are the most common submucosal tumors and include gastrointestinal stromal tumor (GIST), leiomyoma, lipoma, hemangioma, and others.

The classification varies in the literature, with both GIST and leiomyoma described as the most common.

On a barium swallow, a mesenchymal tumor typically appears as smooth, round, submucosal filling defect.

Adenoma

• An esophageal adenoma is a benign mucosal lesion with malignant potential, usually

arising within Barrett esophagus. Most are <1.5 cm in size and resected at endoscopy.

Inflammatory polyp

• An inflammatory polyp is a non-neoplastic, enlarged gastric fold that protrudes up into the lower esophagus.

Inflammatory polyps are almost always associated with reflux

and always contiguous with a gastric fold. They are mucosal in location.

Fibrovascular polyp

• A fibrovascular polyp is a pedunculated mass composed of mesenchymal elements with a significant fatty component.

In contrast to an esophageal adenoma, there is no malignant potential.

Fibrovascular polyps usually occur in the cervical esophagus. The clinical presentation can be dramatic, with regurgitation of a fleshy mass.

• CT is usually diagnostic, demonstrating intra-lesional fatty component.

Varices

• Esophageal varices are most commonly due to portal hypertension.

Varices can usually be distinguished from a solid mass since varices change in size and shape with peristalsis.

However, thrombosed varices may mimic a tumor.

• *Uphill* varices, due to portal hypertension, affect the distal esophagus.

Blood flows “uphill” from the portal vein to left gastric (coronary vein) to periesophageal venous plexus to azygos/hemiazygos collaterals to SVC.

• *Downhill* varices are much less common, are caused by superior vena cava obstruction, and usually affect the proximal esophagus.

Enlarged collateral vessels include the supreme intercostal veins (drain the first intercostal space), bronchial veins, and inferior thyroidal veins.

Foregut duplication cysts

• **Esophageal duplication cyst** is lined with squamous epithelium, has a smooth muscle wall, and is usually in the posterior mediastinum.

It may be either extrinsic to the esophagus or submucosal; the latter is impossible to differentiate from a leiomyoma by esophagram.

• **Bronchogenic cyst** is lined by respiratory epithelium. It is generally indistinguishable from an esophageal duplication cyst on esophagram and CT.

• **Neurenteric cyst** is associated with vertebral body anomalies.

Esophageal foreign body

• A radiopaque esophageal foreign body is best visualized with a lateral radiograph or CT.

• Bony foreign objects usually get stuck in the cervical esophagus.

• Meat impaction usually occurs at the gastroesophageal junction.

There is a risk of esophageal perforation from transmural ischemia if the food bolus is impacted for >24 hours.

Most cases of food bolus impaction are treated with endoscopic removal of the impacted food.

Historically, meat impaction was treated with effervescent

granules and meat tenderizer, but this technique is no longer commonly performed.

There is a risk of esophageal perforation from transmural ischemia if the food bolus is impacted for >24 hours.

Most cases of food bolus impaction are treated with endoscopic removal of the impacted food.

Historically, meat impaction was treated with effervescent granules and meat tenderizer, but this technique is no longer commonly performed.

Malignant esophageal masses

Esophageal carcinoma

• Esophageal carcinoma has a broad range of appearances. Early esophageal cancer may be apparent on barium swallow as a plaque-like lesion, polypoid lesion, or focal irregularity of the esophageal wall.

A classic appearance of advanced esophageal

carcinoma is a mass causing a stricture with a “shouldered” edge and irregular contour.

The uncommon varicoid appearance can be initially confused with varices, but the tumor does not change shape with peristaltic waves as varices typically do.

• Esophageal carcinoma may be squamous cell carcinoma (SCC) or adenocarcinoma, which cannot be reliably differentiated on barium studies.

SCC tends to involve the upper or mid-esophagus and adenocarcinoma typically involves the distal esophagus

and may extend into the stomach.

• Squamous cell carcinoma (SCC) is most commonly due to smoking and alcohol.

Less common risk factors include celiac disease, Plummer–Vinson, achalasia, and human papilloma virus (which more commonly causes laryngeal squamous cell carcinoma).

• Adenocarcinoma is due to chronic reflux, arising from Barrett esophagus distally.

Its incidence has been rising in recent years.

Metastasis

• Direct invasion of the esophagus is most commonly from gastric, lung, or breast primaries.

Hematogenous spread is very rare.

• Most often, mediastinal lymph node metastases will be prominent.

The midesophagus is most commonly affected due to its proximity to mediastinal lymph nodes.

Lymphoma

• Esophageal lymphoma is often indistinguishable from primary esophageal cancer.

Malignant GIST

• Malignant GIST tends to be bulkier and more irregular than the benign variant.

Esophageal motility disorders

Contraction waves

• A primary contraction wave is a normal, physiologic wave initiated by a swallow.

• A secondary contraction wave is a normal, physiologic wave initiated by a bolus in the esophagus.

• A tertiary wave is a nonpropulsive contraction that does not result in esophageal clearing.

Tertiary contractions are seen more commonly in the elderly. They are not normal, but are also not thought to be clinically significant when seen.

Achalasia

• Achalasia is a motility disorder of the distal esophagus, which is unable to relax due to an abnormality of myenteric ganglia in the Auerbach plexus.

Vigorous achalasia is a less severe form of achalasia consisting of repetitive nonpropulsive contractions.

• Chagas disease causes a secondary achalasia that is indistinguishable radiographically from primary achalasia.

• Potential complications of chronic achalasia include esophageal cancer, which has a lag period of at least 20 years, and candidal infection from stasis.

• **The classic imaging appearance of achalasia** is a massively dilated esophagus with a *bird's beak* stricture near the gastroesophageal junction.

• Surgical treatment of achalasia is the Heller myotomy, which is an incision of the lower esophageal muscle fibers.

• **Pseudoachalasia** is caused by an obstructing gastroesophageal junction cancer.

In achalasia, there is transient relaxation of the stricture when the patient stands.

In pseudoachalasia, however, the fixed obstruction does not relax with standing.

Diffuse esophageal spasm (corkscrew esophagus; shish kebab esophagus)

• Diffuse esophageal spasm is a clinical syndrome of chest pain or dysphagia caused by repetitive, nonpropulsive esophageal contractions.

The nonpropulsive contractions have a characteristic appearance on barium swallow, leading to the descriptive names

of *corkscrew esophagus* and *shish kebab esophagus*.

• **Nutcracker esophagus** is a related disorder characterized by high-amplitude contractions on manometry in conjunction with chest pain, with normal radiological findings.

Esophageal diverticula

Types of diverticula

• **Pulsion diverticula** are caused by increased esophageal pressure and comprise nearly all diverticula seen in the USA.

• **Traction diverticula** are caused by traction of adjacent structures, typically resulting from tuberculous mediastinal adenopathy.

They are rarely seen.

Zenker diverticulum

• Zenker diverticulum is an esophageal diverticulum caused by failure of the cricopharyngeus muscle to relax, leading to elevated hypopharyngeal pressure.

Symptoms of a Zenker diverticulum include halitosis, aspiration, and regurgitation of undigested food.

• A Zenker diverticulum is posteriorly protruding.

As a secondary finding, the cricopharyngeus muscle is usually hypertrophied.

• Treatment is with cricopharyngeal myotomy and diverticulopexy or diverticulectomy.

• A **pseudo-Zenker** diverticulum is barium trapped in a pharyngeal contraction wave.

Killian–Jamieson (KJ) diverticulum

• A Killian–Jamieson (KJ) diverticulum is located at the Killian–Jamieson space, which is an area of weakness below the attachment of the cricopharyngeus muscle.

• In contrast to Zenker diverticulum, KJ diverticula are more often bilateral.

• KJ diverticula protrude anteriorly, best seen on the lateral view.

Pseudodiverticulosis

• Pseudodiverticulosis is the imaging finding of multiple tiny outpouchings into the esophageal lumen caused by dilated submucosal glands from chronic reflux esophagitis.

• These submucosal glands are analogous to the Rokitansky–Aschoff sinuses of the gallbladder.

• Pseudodiverticulosis is often associated with a smooth stricture in mid/upper esophagus, which may cause symptoms.

• Candida is frequently cultured, but infection is not believed to be a causal factor.

Miscellaneous esophageal disorders

Feline esophagus

• Feline esophagus is thought to be a normal variant

characterized by multiple transverse esophageal folds.

• There is a controversial association with esophagitis,

where the incidence of esophagitis may be increased in the presence of feline esophagus.

Aberrant right subclavian artery

• Aberrant right subclavian artery (with a normal left arch) is seen in approximately 1% of patients and is almost always asymptomatic.

The aberrant right subclavian artery travels posterior to the esophagus, where it may rarely produce dysphagia.

• On an upper GI study, the resultant posterior esophageal indentation is always smooth.

Scleroderma

• Scleroderma is a systemic disease involving excess collagen deposition in multiple tissues.

• The esophagus is involved in 80% of patients with scleroderma, producing lack of peristalsis of the distal 2/3 of the esophagus due to smooth muscle atrophy and fibrosis, which leads to marked esophageal dilation.

• Secondary candidiasis or aspiration pneumonia can result from prolonged esophageal stasis.

• The esophageal dilation is often apparent before the typical skin changes of scleroderma become evident.

Esophageal hernias

Hiatal hernia (HH)

• A hiatal hernia (HH) is present when gastric folds are seen above the diaphragm.

A hiatal hernia may be sliding (most common) or short (secondary to chronic reflux esophagitis).

Paraesophageal hernia

• With a paraesophageal hernia, the GE junction is located normally below the diaphragm, but a portion of the stomach herniates into the thorax through the esophageal hiatus.

• Paraesophageal hernia is more prone to strangulation than HH. Most are surgically repaired.

Stomach

Thickened gastric folds

• Thickened gastric folds are most commonly due to inflammatory gastritis, which characteristically produces smooth fold thickening.

• Nodular fold thickening is suggestive of neoplasm, such as gastric lymphoma or submucosal carcinoma.

Helicobacter pylori gastritis

• *Helicobacter pylori* is a major cause of gastritis, gastric ulcers and duodenal ulcers.

Zollinger–Ellison (ZE)

• Zollinger–Ellison (ZE) is gastrin over-production from a gastrinoma, which is a pancreatic islet cell tumor that has a 50% rate of malignancy.

ZE features elevated gastrin level and a paradoxical increase in gastrin after secretin administration.

• 25% of patients with gastrinoma have multiple endocrine neoplasia (MEN) type 1.

MEN-1 consists of parathyroid adenoma, pituitary adenoma, and pancreatic islet cell tumors.

Eosinophilic gastritis

• Eosinophilic gastritis is characterized by thickened folds in the stomach and small bowel in a patient with a history of allergy.

Menetrier disease

• Menetrier disease is a protein-losing enteropathy that is often a diagnosis of exclusion.

It usually affects the proximal stomach and is pathologically characterized by replacement of parietal cells by hyperplastic epithelial cells, leading to achlorhydria.

• Menetrier disease has a controversial association with gastric carcinoma.

Crohn disease

• Gastric Crohn disease is almost always associated with small bowel disease.

Usually the distal half of the stomach is affected.

• The earliest pathologic change is the formation of aphthous ulcers.

Other causes of thickened gastric folds

• Gastric varices (from portal hypertension), gastric lymphoma, and submucosal carcinoma are non-inflammatory causes of thickened gastric folds.

Gastric polyps

Hyperplastic polyp (inflammatory polyp)

• A hyperplastic polyp, also known as an inflammatory polyp, is cystic dilation of a gastric gland that develops in response to chronic inflammation.

Hyperplastic polyps are almost always benign, with very rare cases of malignant transformation having been reported.

• **Fundic gland polyposis syndrome** is a variant of familial adenomatous polyposis syndrome that also involves the stomach.

In the stomach, most polyps are hyperplastic, but elsewhere in the GI tract the polyps are adenomatous.

Adenomatous polyp

• An adenomatous polyp is a neoplastic polyp with malignant potential.

There is an elevated risk of malignant transforation to adenocarcinoma if >2 cm in size.

• Adenomatous polyps are usually treated with endoscopic biopsy and polypectomy.

Hamartomatous polyp

• Hamartomatous polyps are benign polyps usually associated with syndromes such as Peutz–Jeghers, juvenile polyposis, and Cronkhite–Canada syndromes.

Benign gastric masses

Lipoma (benign)

• A lipoma is a benign, submucosal, mesenchymal neoplasm. At fluoroscopy, a gastric lipoma is indistinguishable from a GIST. Fatty attenuation on CT is diagnostic of a lipoma.

Gastrointestinal stromal tumor (GIST)

• Gastrointestinal stromal tumor (GIST) is the most common submucosal gastric tumor.

The tumor arises from the interstitial cells of Cajal, which are pacemaker cells that drive peristalsis.

GIST may occur anywhere in the gastrointestinal tract.

• GIST may be benign or malignant, with risk for malignancy determined by size and number of mitoses.

Regardless of size and number of mitoses, gastric GIST is less likely to be malignant compared to similar-sized GISTs in the duodenum, jejunum/ileum, or rectum.

Gastric tumors ≤2 cm in size are essentially always benign. Larger tumors carry a risk of malignancy as high as 86% for a gastric GIST >10 cm with an elevated mitotic rate.

• Small gastric GISTs are usually asymptomatic, but may be a cause of melena.

• On imaging, a smooth endoluminal surface is characteristic due to its submucosal location.

Larger tumors have a tendency to become exophytic, or less commonly to

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Larger tumors have a tendency to become exophytic, or less commonly to invade the lumen.

• The differential diagnosis of a submucosal gastric mass includes mesenchymal tumors (GIST, fibroma, lipoma, neurofibromas, etc.), carcinoid, and ectopic pancreatic rest.

Ectopic pancreatic rest

• An ectopic pancreatic rest is a focus of heterotopic pancreas in the gastric submucosa.

The ectopic pancreatic tissue is susceptible to pancreatic diseases, including pancreatitis and carcinoma.

On imaging, the classic appearance is an umbilicated submucosal nodule,

with the umbilication representing a focus of normal epithelium. The ulceration is not always seen, in which case the imaging is of a nonspecific submucosal gastric mass.

Malignant gastric masses

Gastric cancer

• Gastric adenocarcinoma may present either as a mass or as a gastric ulcer.

• Gastric cancer is generally caused by chronic inflammation, with specific risk factors including:

Ingestion of polycyclic hydrocarbons and

nitrosamines (from processed meats).

Atrophic gastritis.

Pernicious anemia.

Post-subtotal gastrectomy.

• Gastric carcinoma may spread locally from the mucosal surface to the serosa, in which case 90% of patients will have omental involvement from trans-serosal spread.

• Lymphatic spread is along lesser curvature then to gastrohepatic ligament and greater curvature.

• A Krukenberg tumor is classically described as metastatic spread of gastric carcinoma to the ovary; however, the term has also been used to describe any mucinous metastasis to the ovary.

GIST (malignant)

• Malignant GIST tends to be larger than benign GIST, often reaching sizes of greater than 10 cm, with central necrosis. Although the tumor begins in the submucosa, it can be difficult to determine the site of origin of large tumors.

Lymphoma

• Gastric lymphoma can have a wide variety of presentations.

If solitary, lymphoma can mimic gastric carcinoma.

To differentiate between lymphoma and gastric carcinoma, the

pattern of adenopathy can be helpful. In gastric cancer, adenopathy at or below the level of the renal hila is unusual, but occurs more commonly in patients with lymphoma.

• The stomach is a common extranodal site for non-Hodgkin lymphoma.

Metastases

• Metastatic disease to the stomach is rare. Breast, lung, and melanoma are most common.

Gastric ulcers

Benign gastric ulcer

• Although less commonly encountered in the modern era of proton pump inhibitors and *Helicobacter pylori* treatment, benign gastric ulcers tend to have typical imaging findings:

Radiating gastric folds are smooth and symmetric.

Ulcer extends beyond the normal contour of the gastric lumen.

The *Hampton line* represents nonulcerated acid-resistant mucosa surrounding the ulcer crater.

Most benign ulcers occur along the lesser curvature of the stomach, although benign ulcers associated with aspirin ingestion can occur in the greater curvature and antrum, which are dependent locations.

Gastric carcinoma

• Gastric carcinoma may present with malignant ulceration, which can usually be distinguished from a benign ulcer by the following features:

Asymmetric ulcer crater, with surrounding nodular tissue.

Abrupt transition between normal gastric wall and surrounding tissue.

Ulcer crater does *not* project beyond the expected location of gastric wall.

The *Carman meniscus* sign is considered pathognomonic for tumor.

It describes the splaying open of a large, flat malignant ulcer when compression is applied.

Overview of gastric bypass surgery

Postoperative anatomy of Roux-en-Y gastric bypass (RYGB)

• In order to evaluate for and accurately describe complications of Roux-en-Y gastric bypass (RYGB) surgery, it is important to be familiar with the procedure and normal postsurgical anatomy.

• A small gastric pouch is created with a volume of approximately 15 to 30 cc by excluding the distal stomach from the path of food.

• The Roux limb is created by transecting the jejunum approximately 35–45 cm distal to the ligament of Treitz, then bringing it up to be anastomosed to the gastric pouch via a

narrow gastrojejunostomy stoma.

• The current favored approach for placement of the Roux limb is antecolic (in front of the transverse colon).

The Roux limb used to be placed retrocolic, which required

the creation of a surgical defect through the transverse mesocolon (mesentery of the transverse colon).

A retrocolic Roux limb has a higher risk of a transmesocolic hernia due to the defect in the transverse mesocolon.

Although the antecolic approach is now more commonly performed, there are many patients who have previously undergone a retrocolic approach.

• A distal side-to-side jejunojejunostomy is created to connect the pancreaticobiliary limb to the jejunum.

• The RYGB leads to weight loss both from early satiety (due to small size of the gastric pouch) and malabsorption (due to surgical bypass of the proximal jejunum).

Complications of Roux-en-Y surgery

Postoperative leak

• Postoperative leak is usually diagnosed by 10 days after surgery.

• An upper GI study with water-soluble contrast is the study of choice if a leak is suspected.

• Leaks may arise from the distal esophagus, gastric pouch, or blind-ending jejunal limb.

It is rare for a leak to arise from the distal jejunojejunostomy.

Gastrogastric fistula

• A gastrogastric fistula is a communication between the gastric pouch and the excluded stomach, which may be an early or late complication of RYGB.

• A gastrogastric fistula may be a cause of inadequate weight loss or recurrent weight gain.

Small bowel obstruction (SBO)

• Small bowel obstruction (SBO) in the acute postoperative period is most often due to edema or hematoma at the gastrojejunostomy or jejunojejunostomy.

With a retrocolic Roux limb, edema at the transverse mesocolon defect may also cause obstruction.

Treatment is usually conservative, with most cases resolving as the edema and/or hematoma resolves.

• A late presentation of small bowel obstruction may be due to internal hernia (more common with laparoscopic surgery) or adhesions (more common with open surgery).

Internal hernia

• Laparoscopic Roux-en-Y procedures are associated with a higher rate of internal hernias (seen in 2.5% of laparoscopic procedures) compared to open procedures (0.5%).

Internal hernias can be difficult to diagnose, both clinically and by imaging.

• Internal hernias usually present within 2 years of bypass and are the most common cause of SBO after a laparoscopic Roux-en-Y.

• Most RYGB-associated internal hernias occur in three characteristic locations.

• The surgically created defect in the mesentery of the transverse colon is the most common site (the **transmesocolic** hernia), associated with a retrocolic Roux limb.

• Less common sites of internal hernia include **Peterson's space** (located between the mesentery of the Roux limb and the transverse mesocolon) and the **mesenteric defect created by the jejunojejunostomy**.

• Imaging features of internal hernia include swirling of the mesentery, a mushroom shape of the mesentery, and/or the presence of small bowel loops posterior to the superior mesenteric artery.

Stomal stenosis

• Narrowing of the gastrojejunostomy stoma may occur in up to 10% of patients, leading to dilation of the pouch and distal esophagus.

Stomal stenosis is usually treated with endoscopic dilation.

• Narrowing of the distal jejunojejunostomy is much more rare and usually requires surgery.

Marginal ulcers

• The jejunal mucosa adjacent to the gastrojejunal anastomosis is susceptible to gastric secretions, which can cause marginal ulcers in up to 3% of patients.

• A marginal ulcer is diagnosed by upper GI as a thickening and small outpouching of a gastric fold.

• Treatment is conservative.

Small bowel

Small bowel anatomy

• The wall of the small intestine is made of four layers, from outside in:

Serosa.

Muscularis (thin longitudinal and thick circumferential smooth muscle).

Submucosa.

Mucosa (consists of intestinal villi, circular folds, glands, and lymphoid tissue).

• Valvulae conniventes create the characteristic small bowel fold pattern.

• The superior mesenteric artery (SMA) supplies both the jejunum and ileum.

A common small bowel mesentery anchors the jejunum and ileum to the posterior abdominal wall.

The jejunum features larger, more feature-full folds and larger villi compared to the ileum.

Small bowel obstruction (SBO)

• Small bowel obstruction (SBO) is common and most often due to adhesions from prior surgery or hernia.

Neoplasm, stricture, and intussusception are less common causes.

Radiographic evaluation of small bowel obstruction

• An abdominal radiograph is often the initial imaging evaluation for suspected obstruction.

• Radiographic findings of SBO include small bowel distention and multiple air–fluid levels at different heights seen on the upright view.

In addition, the lack of gas in the colon is especially suggestive of obstruction.

An upright or decubitus view is generally necessary to confidently diagnose obstruction.

• Potential false positives for diagnosing SBO on plain radiographs include:

Ileus with prior colectomy: Would not see gas in the colon.

Ileus with ascites: Ascites often compresses the ascending and descending colon and rectum as

these structures are not on a mesentery. However, gas in the transverse colon and sigmoid colon is still apparent.

CT imaging of small bowel obstruction

• CT is highly sensitive and specific for diagnosis of SBO. Small bowel distention ≥3 cm with a transition point to collapsed bowel is highly specific for a small bowel obstruction.

• In addition to diagnosing obstruction, CT can show the transition point, the cause of obstruction, and potential complications of obstruction such as ischemia or strangulation.

• It is important to approach the interpretation of an obstruction in a systematic way.

• **First**, look for the transition point to decompressed bowel to determine the cause.

• **Second**, always determine if the obstruction is *simple* or *closed-loop.* A closed-loop obstruction is a *never miss* lesion as there is very high risk for bowel ischemia and severe morbidity and mortality.

• **Third**, evaluate for signs of ischemia or impending ischemia, which include (in rough

order of severity):

Engorged mesenteric vessels.

Ascites surrounding the bowel, due to increased capillary permeability.

Wall thickening, due to submucosal edema.

Lack of bowel wall enhancement, due to vasoconstriction or under-perfusion. Note that the presence or absence of bowel wall enhancement can only be assessed if positive oral contrast was not given.

Pneumatosis intestinalis, which is gas in the bowel wall due to necrosis.

Pneumatosis produces multiple small locules of gas seen circumferentially in the bowel wall.

• In addition to small bowel distention >3 cm and a transition point to decompressed bowel, an additional helpful CT finding of SBO is the *small bowel feces sign,* which describes

particulate feculent material mixed with gas bubbles in the small bowel that resembles the CT appearance of stool.

The small bowel feces sign is often seen just proximal to the transition point and is helpful to localize the site of transition.

The small bowel feces sign may be especially helpful in subacute or partial obstruction, which can otherwise be difficult to diagnose.

The small bowel feces sign is thought to be due to bacterial overgrowth and undigested food.

Closed loop obstruction

• Closed loop obstruction is a surgical emergency that may lead to bowel ischemia.

Closed loop obstruction represents obstruction of both the efferent and afferent segments of a single loop of bowel.

• Closed loop obstruction may be secondary to adhesions or hernia.

The formation of a narrow pedicle can lead to volvulus, which predisposes to ischemia.

• CT imaging features include a U-shaped distribution of the bowel loop with radially oriented vessels.

If volvulus is present, the *whirl* sign may be seen, due to twisting of mesenteric vessels.

Obstruction due to adhesions

• Adhesions from prior surgery or intra-peritoneal inflammatory process are the most common cause of small bowel obstruction.

• Adhesions are an imaging diagnosis of exclusion. On CT, a transition point is seen, but no obvious cause for the transition (e.g., no mass or hernia, etc.) is identified.

• The vast majority of patients with SBO due to adhesions have had prior abdominal surgery.

Obstruction due to external hernia

• Protrusion of bowel through the abdominal wall is the second most common cause of small bowel obstruction.

Approximately 75% of external hernias occur in the groin,

with the majority being inguinal hernias.

• An **inguinal hernia** may be either indirect or direct, depending on the relation of the hernia to the inferior epigastric vessels.

Indirect: Indirect inguinal hernia is the most common type and is more common in males.

The neck of the hernia is lateral to the inferior epigastric vessels. Hernia contents travel with the spermatic cord, often *into the scrotum.*

Indirect inguinal hernias are considered a congenital lesion due to a patent processus vaginalis.

Direct: The neck of an indirect inguinal hernia is medial to the inferior epigastric vessels, protruding through a weak area in the

anterior abdominal wall. The hernia contents *do not go into scrotum.*

• In an **obturator hernia**, bowel herniates through the obturator canal.

Obturator hernias are almost always seen in elderly women due to pelvic floor laxity.

The key imaging finding is bowel located between the pectineus and obturator muscles.

It is important to correctly diagnose an obturator hernia preoperatively.

An obturator hernia requires a very different surgery from inguinal hernia, and has an especially high morbidity and mortality if incarcerated.

Ventral hernia is often due to prior laparotomy.

Obstruction due to internal hernia

• Protrusion of bowel through the peritoneum or mesentery into a compartment in the abdominal cavity is a relatively uncommon cause of small bowel obstruction.

• **Transmesenteric hernia** is a broad category of bowel herniation through defects in any of the three true mesenteries (small bowel mesentery, transverse mesocolon, and sigmoid mesentery).

The most common type of transmesenteric hernia is the

transmesocolic hernia, due to a defect in the transverse mesocolon (mesentery of the transverse colon). Transmesocolic hernia is seen most commonly post Roux-en-Y gastric bypass or biliary-enteric anastomosis from liver transplant.

The lack of confining hernia sac and variable imaging appearance make diagnosis difficult.

A clue on imaging may be posterior displacement of the colon, with small bowel located anterior to the colon.

The SMA and SMV may be displaced and engorged.

Internal hernias carry a high rate of volvulus. If volvulus is present, the *whirl* sign may be visible.

Transmesenteric hernias are also the most common type of hernia in children, not due to surgery but secondary to a congenital mesenteric defect thought to be from prenatal intestinal ischemia.

In children, the mesenteric defect has a variable position.

• **Paraduodenal hernia** was previously the most common internal hernia (older literature states 53% of internal hernias were paraduodenal), prior to the rise in gastric bypass surgery. Paraduodenal hernias are congenital anomalies, due to

embryologic failure of mesenteric fusion and resultant mesenteric defect.

They more commonly occur on the left.

Paraduodenal hernia is associated with abnormal rotation of the intestine.

A common clinical complaint described by patients with paraduodenal hernia is chronic postprandial pain often relieved by massaging, which reduces the hernia.

In the more common left paraduodenal hernia, the bowel can herniate through a mesenteric defect named Landzert's fossa, located behind the ascending (fourth) duodenum.

The key imaging finding is a cluster of small bowel loops between the pancreas and stomach.

• **Foramen of Winslow hernia**: The foramen of Winslow is the communication between the lesser sac and the greater peritoneal cavity.

The key imaging features of a foramen of Winslow hernia are dilated loops of bowel in the upper abdomen and presence of mesentery between the IVC and main portal vein.

Obstruction due to neoplasm

• A mass intrinsic to the bowel or external compression from an extrinsic mass may cause small bowel obstruction. An extrinsic mass is usually straightforward to diagnose by CT.

• Although the presence of an intraluminal mass may be more difficult to detect on CT, clues to the presence of an intrinsic mass include irregular bowel wall thickening and/or regional lymphadenopathy.

• Primary small bowel neoplasm causing intrinsic bowel obstruction may be due to adenocarcinoma, GIST, and carcinoid.

Metastatic causes of intrinsic bowel neoplasm include

melanoma, ovarian, and lung cancer. Melanoma is known to cause intussusception.

• Lymphoma is generally a “soft” tumor and rarely causes obstruction.

Aneurysmal expansion of the small bowel wall is a classic appearance, but presentation is highly variable.

Obstruction due to intussusception

• While transient intussusceptions are a common incidental finding, an intussusception causing obstruction should raise suspicion for an underlying lesion and prompt

surgery.

Obstruction due to Crohn disease

• Stricture or active enteritis is an important cause of bowel obstruction in Crohn disease, especially the fibrostenotic subtype. Crohn disease is discussed on the

following page.

Obstruction due to gallstone

• Gallstone ileus is due to a gallstone that has eroded through into the small bowel, causing the classic Rigler’s triad of

pneumobilia (from cholecystoduodenal fistula), small bowel obstruction, and ectopic gallstone within the small bowel.

Enteritis

• Enteritis is inflammation of the small bowel.

The most common CT manifestation of enteritis is bowel wall thickening.

Mesenteric stranding or free fluid may also be present.

Crohn disease

• Crohn disease is a chronic granulomatous inflammatory condition that may affect any part of the gastrointestinal tract from the mouth to the anus.

Involvement is discontinuous, with characteristic skip lesions of intervening normal GI tract.

The most common site of involvement is the small bowel, especially the terminal ileum.

• The earliest histologic changes occur in the submucosa, seen on imaging as aphthous ulcers due to lymphoid hyperplasia and lymphedema.

• Endoscopy and barium fluoroscopy (small bowel follow-through, enteroclysis, and barium enema) have historically been the modalities to evaluate Crohn disease.

More recently, however, CT and MR enterography are emerging as the exams of choice.

The advantages of CT and MRI are the ability to visualize beyond the bowel lumen to evaluate the bowel wall, presence of extraintestinal complications, and the vasculature.

The disadvantages of CT and MRI compared to fluoroscopy and endoscopy are reduced spatial resolution and limited sensitivity for detecting subtle early signs of disease.

• The most common imaging finding on all modalities is wall thickening of the terminal ileum.

• Fluoroscopic findings include thickened, nodular folds in the affected regions of small bowel, luminal narrowing, mucosal ulceration, and separation of bowel loops.

The typical *cobblestone* appearance seen on endoscopy and fluoroscopy is a result of crisscrossing deep ulcerations.

• The fibrostenotic subtype of Crohn disease may clinically present with bowel obstruction.

Asymmetric bowel fibrosis from ulcerations of the mesenteric side of the bowel produces pseudosacculations on the antimesenteric side.

The fibrosis can lead to a segmental stricture, called the *string* sign.

• Complications of Crohn disease include bowel strictures, fistulae, and abscesses.

Scleroderma

• Scleroderma is a systemic disease characterized by the deposition of collagen into multiple internal organs and the skin.

• The primary insult to the gastrointestinal tract in scleroderma is impaired motility due to replacement of the muscular layers with collagen, which leads to slowed transit and subsequent bacterial overgrowth, progressive dilation, and pseudo-obstruction.

• Radiographic findings are sacculations on the antimesenteric border (side opposite where the mesentery attaches) and a *hidebound* bowel due to thin, straight bowel folds stacked together.

• Treatment is with antibiotics for bacterial overgrowth and prokinetic drugs such as erythromycin or octreotide for bowel motility.

Celiac disease (sprue, gluten-sensitive enteropathy)

• Celiac disease, also known as sprue and gluten-sensitive enteropathy, is an autoimmune, proximal enteritis caused by a T-cell-mediated immune response triggered by antigens in ingested gluten.

• The primary sites of involvement are the duodenum and jejunum.

• The most characteristic imaging finding of celiac disease is reversal of jejunal and ileal fold patterns.

Normally, the jejunum has more folds than the ileum.

However, in celiac disease, the loss of jejunal folds causes a compensatory increase in the number of ileal folds.

• Villous atrophy causes the loss of jejunal folds and hypersecretion of intraluminal fluid that creates *flocculations* of barium due to lack of contrast adhesion to the bowel wall.

The *moulage* (french for casting) sign is seen on a barium study and refers to a castlike appearance of the featureless jejunum.

• The CT findings of celiac disease include dilated, fluid-filled bowels, often with intra-luminal flocculations of enteric contrast.

Contrast can be seen both insinuated between the small bowel folds and centrally within the bowel, with a peripheral layer

of low-attenuation secretions.

Other CT findings of celiac disease include mesenteric adenopathy and engorgement of mesenteric vessels.

• Unlike other causes of enteritis, diffuse bowel wall thickening and ascites are less common.

• An important complication of celiac disease is small bowel **T-cell lymphoma**, which may manifest as an exophytic mass, circumferential bowel wall thickening, or enlarged mesenteric lymph nodes.

• Other complications of celiac disease include:

Intussusception, thought to be due to uncoordinated peristalsis, without a lead-point mass.

Pneumatosis intestinalis, thought to be due to dissection of intraluminal gas through the inflamed bowel wall.

Pneumatosis in the setting of celiac disease is not thought to reflect bowel ischemia.

Splenic atrophy.

Increased risk of **venous thromboembolism**.

Lab abnormalities include anemia (secondary to malabsorption), leukopenia, and immunoglobulin deficiency.

Skin abnormalities include the characteristic dermatitis herpetiformis rash.

Cavitating mesenteric lymph node syndrome (CMLNS) is a very rare complication of celiac disease, with only 36 reported cases in the literature.

The central portion of affected lymph nodes shows low attenuation due to liquid necrosis. CMLNS is thought to be highly specific for celiac disease when seen in combination with villous atrophy and splenic atrophy.

The differential diagnosis of low attenuation mesenteric lymph nodes includes tuberculosis, Whipple disease,

treated lymphoma, and CMLNS.

Infectious enteritis

• Several bacterial, viral, and fungal organisms may cause enteritis.

• *Yersinia* and tuberculosis have a propensity to affect the terminal ileum, mimicking Crohn disease.

• *Salmonella* is the most common cause of food-borne gastroenteritis and causes segmental distal small bowel thickening on CT and segmental nodular thickened folds

on fluoroscopy.

Radiation enteritis

• Long-term effects of radiation to the pelvis include adhesive and fibrotic changes to the mesentery and small bowel.

• Clues to the diagnosis of radiation enteritis include a history of radiation therapy and regional involvement of bowel loops not confined to a vascular territory.

• Imaging findings include mural thickening and mucosal hyperenhancement with narrowing of the lumen.

Radiation enteritis may be a cause of small bowel obstruction.

Whipple disease

• Whipple disease is due to infection by *Tropheryma whippelii*, which manifests in the GI tract as malabsorption and abdominal pain.

Whipple disease may cause arthralgias and increased skin pigmentation.

• Whipple disease characteristically causes low attenuation adenopathy that may appear similar to the cavitating mesenteric lymph node syndrome seen in celiac disease.

• **Radiographically**, Whipple disease causes thickening and nodularity of duodenal and proximal small bowel folds.

In contrast to celiac disease, there is typically no hypersecretion.

Graft versus host disease (GVHD)

• Graft versus host disease is a complication of bone marrow transplantation.

The skin, liver, and gastrointestinal tract are most commonly affected.

• Imaging findings of GVHD include nonspecific wall thickening and effacement of the normal small bowel fold pattern.

While the classic barium finding is the *ribbon* bowel,

this is not often seen.

Large bowel

Colitis

Overview of colitis

• Colitis is inflammation of the colon that may be caused by several unrelated etiologies, often with overlapping imaging findings.

• The primary imaging feature of colitis is bowel wall thickening.

Generally, a full clinical evaluation, stool studies, and sometimes colonic biopsy are required for a definitive diagnosis.

• Incidental colonic wall thickening is found in as many as 10% of CT scans.

Ischemic colitis

• Colonic ischemia can be caused by acute arterial thrombus, chronic arterial stenosis, low-flow states (e.g., congestive heart failure), and venous thrombosis.

• The splenic flexure is the watershed region between the superior and inferior mesenteric arteries and is especially susceptible to ischemia in low-flow states.

• The rectum is supplied by a dual blood supply and is almost never affected by ischemia.

The superior rectal artery (terminal branch of the IMA) and the inferior and middle rectal arteries (arising from the internal iliac artery anterior division) form perirectal collaterals.

• A suggestive CT finding of ischemic colitis is segmental, continuous thickening of the affected colon in a vascular distribution, with sparing of the rectum.

If arterial thromboembolic disease is suspected, one should evaluate for the presence of aortic atherosclerotic disease or a left atrial thrombus in the setting of atrial fibrillation.

If chronic arterial stenosis is suspected, one should evaluate for atherosclerosis of the mesenteric vessels.

Infectious colitis

• Infectious colitis can be bacterial, tubercular, viral, or amoebic. There is a large overlap in the clinical presentation and imaging findings of the various pathogens.

• In general, infectious colitis features pericolonic stranding and ascites in addition to the colonic wall thickening seen in all forms of colitis.

• *Yersinia*, *Salmonella*, and colonic tuberculosis affect the right colon. Tuberculosis is known to involve the ileocecal valve, resulting in a desmoplastic reaction that mimics Crohn disease.

• *E. coli*, CMV, and *C. difficile* colitis (discussed below) most commonly cause pancolitis.

Pseudomembranous colitis

• Pseudomembranous colitis is an especially prevalent form of infectious colitis caused by overgrowth of *Clostridium difficile*, most commonly due to alteration in colonic bacterial flora after antibiotic use.

]Pseudomembranous colitis may also occur without a history of antibiotics, especially in hospitalized or nursing home patients.

• A key imaging finding is marked thickening of the colonic wall, typically with involvement of the entire colon (pancolitis). The *accordion* sign describes severe colonic wall thickening combined with undulation of enhancing inner mucosa.

It signifies severe colonic edema but is not specific to *C. difficile*.

Thumbprinting is a fluoroscopic finding of thickened haustra and is also due to edema.

Ulcerative colitis (UC)

• Ulcerative colitis (UC) is an idiopathic inflammatory bowel disease that begins distally in the rectum and spreads proximally in a continual manner (unlike Crohn disease, which features skip areas).

Of note, it is possible for the rectum to appear normal with more proximal colonic involvement present if the patient has been treated with corticosteroid enemas.

• Patients with UC have an increased risk of primary sclerosing cholangitis, colon cancer, and cholangiocarcinoma.

• Extra-abdominal manifestations of UC include sacroiliitis, iritis, erythema nodosum (tender red subcutaneous nodules), and pyoderma gangrenosum (cutaneous ulcers).

• UC does not extend more proximally than the cecum; however, a *backwash ileitis* caused by reflux of inflammatory debris into the ileum may mimic Crohn disease.

• Imaging of ulcerative colitis features circumferential wall thickening with a granular mucosal pattern that is best seen on barium enema.

Pseudopolyps may be present during acute inflammation, representing islands of normal mucosa surrounded by inflamed mucosa.

A *collar-button* ulcer is nonspecific but represents mucosal ulceration undermined by submucosal extension.

• Chronic changes of ulcerative colitis include a featureless and foreshortened *lead pipe* colon.

Similar to Crohn disease, fat-attenuation of the colonic wall suggests chronic disease, as seen in the case above.

• Toxic megacolon is a severe complication of ulcerative colitis (and less commonly, Crohn disease) caused by inflammation extending through the muscular layer.

Imaging of toxic megacolon shows dilation of the colon to greater than 6 cm in association with an adynamic ileus. Colonic perforation may occur and colonoscopy is contraindicated in suspected toxic megacolon.

Typhlitis (neutropenic enterocolitis)

• Typhlitis is a right-sided colitis seen in immunocompromised patients.

• Treatment is with broad-spectrum antibiotics and antifungals.

Polyposis syndromes affecting the bowel

Familial adenomatous polyposis (FAP)

• Familial adenomatous polyposis (FAP) is an autosomal-dominant syndrome featuring innumerable premalignant adenomatous polyps in the colon and to a lesser extent the

small bowel.

Prophylactic colectomy is the standard of care to prevent colon cancer.

• Gastric polyps are also present, although the gastric polyps are hyperplastic and are not premalignant.

• **Gardner** syndrome is a variant of FAP. In addition to colon polyps, patients also have:

Desmoid tumors.

Osteomas.

Papillary thyroid cancer.

Epidermoid cysts.

Mnemonic: DOPE Gardner

• **Turcot** syndrome is another variant of FAP. In addition to colon polyps, patients also have CNS tumors (gliomas and medulloblastomas).

Hereditary nonpolyposis colon cancer syndrome (HNPCC) = Lynch syndrome

• Hereditary nonpolyposis colon cancer (HNPCC) syndrome (also called Lynch syndrome) is an autosomal dominant polyposis syndrome caused by DNA mismatch repair, leading to colon cancer from microsatellite instability on a molecular level.

• Similar to FAP, the colon polyps of HNPCC are adenomatous.

• HNPCC is associated with other cancers, including endometrial, stomach, small bowel, liver, and biliary malignancies.

Peutz–Jeghers

• Peutz–Jeghers is an autosomal dominant syndrome that features multiple hamartomatous pedunculated polyps, usually in the small bowel.

These polyps may act as lead points and cause intussusception.

• Characteristic skin manifestations include perioral mucocutaneous blue/brown pigmented spots on the lips and gums.

• Peutz–Jeghers is associated with gynecologic neoplasms as well as gastric, duodenal, and colonic malignancies.

Cowden syndrome

• Cowden syndrome is an autosomal dominant syndrome of multiple hamartomatous polyps most commonly found in the skin and external mucous membranes, but also in the gastrointestinal tract.

• Cowden syndrome is associated with an increased risk of thyroid cancer (usually follicular), as well as skin, oral, breast, and uterine malignancies.

Cronkhite–Canada

• Cronkhite–Canada is a non-inherited disorder (the only polyposis syndrome in this list that is not autosomal dominant) consisting of hamartomatous polyps throughout the

gastrointestinal tract.

• Cutaneous manifestations include abnormal skin pigmentation, alopecia, and onychodystrophy (malformation of the nails).

Acute bowel

Appendicitis

• Appendicitis is the most common surgical cause of acute abdomen.

Acute inflammation of the appendix is thought to be due to obstruction of the appendiceal lumen, leading to venous congestion, mural ischemia, and bacterial translocation.

• Appendicitis represents a spectrum of severity ranging from *tip appendicitis* (inflammation isolated to the distal appendix) to *gangrenous appendicitis* with abscess if the disease is not diagnosed until late.

• Greater than 97% of patients undergo a preoperative CT prior to appendectomy, with resultant decrease in negative appendectomy rate from 23% in 1990 to 1.7% in 2007.

• Imaging of appendicitis relies on direct and indirect imaging findings.

• **Direct findings of appendicitis** are due to abnormalities of the appendix itself:

Distended, fluid-filled appendix: 6 mm is used as cutoff for normal diameter of the appendix, although there is wide normal variability and 6 mm is from the ultrasound literature using compression. A normal appendix distended with air can measure >6 mm; therefore, some authors advocate using caution with a numeric cutoff in an otherwise normal-appearing appendix filled with air or enteric contrast.

Appendiceal wall-thickening.

Appendicolith, which may be a cause of luminal obstruction; however, appendicoliths are commonly seen without associated appendicitis.

• **Indirect findings of appendicitis** are due to the spread of inflammation to adjacent sites:

Periappendiceal fat stranding.

Cecal wall thickening.

Hydroureter.

Small bowel ileus.

• Appendicitis can also be evaluated by ultrasound, with the key sonographic finding a tubular, blind-ending, non-compressible right lower quadrant structure measuring >6 mm in diameter.

Is generally necessary to use graded compression to evaluate

for compressibility.

Secondary findings of appendicitis can be evaluated by ultrasound, including free fluid and periappendicular abscess.

Diverticulitis

• Diverticulitis is microperforation and acute inflammation of a colonic diverticulum.

CT is the primary modality for diagnosis, triage, and evaluation of severity and complications.

• The left colon is affected far more commonly than the right.

• It is often impossible to distinguish acute diverticulitis from microperforated colon cancer.

Many authors recommend follow-up colonoscopy after the acute episode has resolved, although this recommendation is somewhat controversial and varies by institution.

• **Uncomplicated diverticulitis** does not have any imaging evidence of bowel perforation (even though histopathologically all diverticulitis is associated with bacterial translocation across the bowel wall).

CT findings of uncomplicated diverticulitis include bowel wall thickening and pericolonic fat stranding, usually centered around a culprit diverticulum.

• **Complicated diverticulitis** implies the presence of an additional complication, including:

Pericolonic or hepatic abscess.

Extraluminal air.

Bowel obstruction.

Bowel fistula (colovesical fistula most common, apparent on imaging as gas in the bladder not explained by Foley catheter placement).

Mesenteric venous thrombosis.

• Uncomplicated diverticulitis is typically treated conservatively.

• Abscesses can usually be drained percutaneously.

• Indications for surgery include the presence of a fistula or recurrent diverticulitis, with two prior episodes of diverticulitis treated conservatively.

Epiploic appendagitis

• Epiploic appendagitis is a benign, clinical mimic of diverticulitis caused by torsion of a normal fatty tag (appendage) hanging from the colon.

• Epiploic appendagitis has a pathognomonic imaging appearance of an oval fat attenuation lesion abutting a normal colonic wall, with mild associated fat stranding.

A central hyperdense dot in cross-section represents the thrombosed central vein of the epiploic appendage.

• Treatment is with anti-inflammatories, *not* antibiotics or surgery.

Mesentery and peritoneum

Anatomy

Peritoneum

• The peritoneum is a thin membrane consisting of a single layer of mesothelial cells that are supported by subserosal fat cells, lymphatic cells, and white blood cells.

• The visceral peritoneum lines the surface of all intraperitoneal organs, while the parietal peritoneum lines the outer walls of the peritoneal cavity.

• The most dependent portion of the peritoneal cavity (both supine and upright) is the pouch of Douglas in women and the retrovesical space in men.

Mesentery

• There are three true mesenteries, which each supply a portion of the bowel and connect to the posterior abdominal wall. Each mesentery consists of a network of blood vessels and lymphatics, sandwiched between the peritoneal layers.

The three true mesenteries are:

Small bowel mesentery: Supplies both the jejunum and ileum. Oriented obliquely from the ligament of Treitz in the left upper quadrant to the ileocecal junction in the right lower quadrant.

Transverse mesocolon: Mesentery to the transverse colon, connecting the posterior transverse colon to the posterior abdominal wall

Sigmoid mesentery: Mesentery to the sigmoid colon.

• The greater and lesser omentum are specialized mesenteries that attach to the stomach.

The greater and lesser omentum do not connect to the posterior abdominal wall.

Greater omentum: Large, drape-like mesentery in the anterior abdomen, which connects the stomach to the anterior aspect of the transverse colon.

Lesser omentum: Connects stomach to liver.

Flow of peritoneal fluid

• Peritoneal fluid is constantly produced, circulated, and finally resorbed around the diaphragm, where it eventually drains into the thoracic duct.

“Misty” mesentery

Overview of the “misty” mesentery

• As previously discussed, the abdominal mesenteries are fatty folds through which the arterial supply and venous and lymphatic drainage of the bowel run.

• The mesenteries themselves are not seen on CT because they are made primarily of fat and blend in with intra-abdominal fat. However, the vessels which course through the mesentery are normally seen.

• Infiltration of the mesentery by fluid, inflammatory cells, tumor, or fibrosis may increase the attenuation of the mesentery and cause the mesenteric vasculature to appear indistinct.

These findings are often the first clue to certain pathologies.

Mesenteric edema

• Edema of the mesentery may be secondary to either systemic or intra-abdominal etiologies.

• Systemic causes of edema include congestive heart failure, low protein states, and third-spacing, all of which can lead to diffuse mesenteric edema.

• Focal mesenteric edema may be secondary to an intra-abdominal vascular cause, such as mesenteric vessel thrombosis, Budd–Chiari syndrome, or IVC obstruction.

Abdominal vascular insults may cause bowel ischemia, which manifest on imaging as bowel wall thickening, pneumatosis, or mesenteric venous gas.

Mesenteric inflammation

• The most common cause of mesenteric inflammation in the upper abdomen is acute pancreatitis.

However, any focal inflammatory process such as appendicitis,

inflammatory bowel disease, and diverticulitis may cause local mesenteric inflammation leading to the “misty” mesentery appearance.

• Mesenteric panniculitis is an idiopathic inflammatory condition, which may cause a diffuse “misty” mesentery.

Intra-abdominal hemorrhage

• Intra-abdominal hemorrhage tends to be localized, surrounding the culprit bleeding vessel unless large. Hemorrhage may be secondary to trauma, post-procedural, or due to anticoagulation.

Neoplastic infiltration

• Neoplastic infiltration of the mesentery may cause the “misty” mesentery.

The most common tumor involving the mesentery is non-Hodgkin lymphoma, which typically also causes bulky adenopathy.

• Mesenteric involvement may be especially apparent after treatment, where the “misty” mesentery is limited to the portion of the mesentery that contained the treated lymph nodes.

• Other tumors that may involve the mesentery include pancreatic, colon, breast, gastrointestinal stromal tumor, and mesothelioma.

Mesenteric masses

Overview of mesenteric masses

• Primary mesenteric tumors are rare, although the mesentery is a relatively common site of metastasis.

Carcinoid

• Gastrointestinal carcinoid is a relatively rare tumor compared to other gastrointestinal malignancies, but is the most common small bowel tumor.

It typically occurs in the distal ileum.

• Carcinoid usually arises as an intraluminal mass and may secondarily spread to the mesentery either by direct extension or lymphatic spread.

Up to 80% of carcinoids spread to the mesentery.

• A classic imaging appearance of carcinoid affecting the mesentery is an enhancing soft-tissue mass with radiating linear bands extending into the mesenteric fat.

Calcification is common.

The radiating linear bands do not represent infiltrative tumor but are the result of an intense desmoplastic reaction caused by the release of serotonin by the tumor.

• The differential diagnosis of a sclerosing mesenteric mass includes:

Carcinoid.

Desmoid tumor.

Sclerosing mesenteritis.

Desmoid tumor

• Desmoid tumor is a benign, locally aggressive mass composed of proliferating fibrous tissue.

• Desmoid may be sporadic, but mesenteric desmoid tumors are more common in patients with Gardner syndrome (a variant of familial adenomatous polyposis).

• On **CT**, most desmoids are isoattenuating to muscle, but large tumors may show central necrosis.

A characteristic imaging feature is strands of tissue radiating into the adjacent mesenteric fat, similar to mesenteric carcinoid and sclerosing mesenteritis.

Sclerosing mesenteritis

• Sclerosing mesenteritis is a rare inflammatory condition that leads to fatty necrosis and fibrosis of the mesenteric root.

• Imaging of sclerosing mesenteritis shows mesenteric masses with striations of soft tissue extending into the adjacent fat. Calcification may be present.

• Mesenteric panniculitis is a variant where inflammation predominates and presents as acute abdominal pain.

On **CT**, there is a “misty” mesentery, sometimes with linear

bands of soft tissue representing early fibrosis.

Mesenteric metastases and lymphoma

• Gastric, ovarian, breast, lung, pancreatic, biliary, colon cancer, and melanoma can metastasize to mesenteric lymph nodes.

• Mesenteric lymphoma can produce the *sandwich* sign, where the mesenteric fat and vessels (the sandwich filling) are engulfed on two sides by bulky lymphomatous masses (the bread).

Diffuse peritoneal disease

Peritoneal carcinomatosis

• Peritoneal carcinomatosis represents disseminated metastases to the peritoneal surface.

• The term *omental caking* describes the replacement of omental fat by tumor and fibrosis.

• Mucinous adenocarcinoma is the most common tumor type to cause peritoneal carcinomatosis. peritoneal carcinomatosis due to mucinous adenocarcinoma should not be confused with pseudomyxoma peritonei.

Pseudomyxoma peritonei

• Pseudomyxoma peritonei is a low-grade malignancy characterized by copious mucus in the peritoneal cavity.

• In general, pseudomyxoma peritonei is thought to be produced by a mucin-producing adenoma or adenocarcinoma of the appendix; however, there is some controversy as to whether the ovary or colon can be a primary site as well.

Pseudomyxoma peritonei is often associated with an ovarian mass (up to 30% of female patients), but it is thought that these are most often metastatic deposits.

• Pseudomyxoma peritonei was previously thought to be produced by a benign appendiceal mucocele, which is now believed to occur much less commonly than originally thought.

20% of all appendiceal adenomas or adenocarcinomas will cause pseudomyxoma peritonei.

Only 2% of all appendiceal mucoceles (which occur slightly less commonly than appendiceal adenomatous lesions) will cause pseudomyxoma peritonei.

• Tumor deposits tend to be spread throughout the entire peritoneal cavity due to intraperitoneal fluid currents.

• Clinically, pseudomyxoma peritonei presents with recurrent mucinous ascites.

The surgeons refer to the mucinous ascites as a “jelly belly.”

• **CT** shows lobular ascites that is typically of slightly higher attenuation (5–20 Hounsfield units) compared to fluid ascites.

Occasionally, mucus can be seen in the region of the appendix, but the flow of peritoneal contents tends to spread the

mucinous ascites diffusely throughout the peritoneum.

• Advanced disease shows pathognomonic scalloping of the hepatic margin.

• Treatment continues to evolve, but the best outcomes are primarily with surgical treatment and hyperthermic intraperitoneal chemotherapy lavage.

Genitourinary imaging

Retroperitoneum

Retroperitoneal anatomy

• The retroperitoneum can be separated into three compartments by the anterior and posterior renal fascia and the lateral conal fascia.

• The adrenals and kidneys are located within the perirenal space of the retroperitoneum.

• The ascending and descending colon, the second and third portions of the duodenum, and the pancreas are located in the anterior pararenal space of the retroperitoneum.

• The third compartment of the retroperitoneum, the posterior pararenal space, is a potential space that is clinically important as a pathway for potential disease spread due to secondary involvement of inflammation or neoplasm.

Retroperitoneal disease

Liposarcoma

• Liposarcomas are a diverse group of neoplasms that make up the most common primary retroperitoneal tumors.

10–15% of all liposarcomas arise from the retroperitoneum.

• The most common type of liposarcoma is the well-differentiated group, which is composed of adipocytic, sclerosing, and inflammatory subtypes.

Adipocytic liposarcoma resembles a lipoma, predominantly composed of fat with strands of tissue representing collagen bands.

• In order of increasing malignancy, liposarcomas may also be myxoid, round-cell, pleomorphic, or dedifferentiated.

The more aggressive subtypes may have minimal or no areas of macroscopic fat and may be indistinguishable from other malignant soft-tissue masses.

Retroperitoneal fibrosis

• Retroperitoneal fibrosis is a rare inflammatory disorder causing increased fibrotic deposition in the retroperitoneum, often leading to ureteral obstruction.

• Unlike malignant retroperitoneal adenopathy, retroperitoneal fibrosis tends *not* to elevate the aorta off the spine.

Adrenal glands

Anatomy

• The adrenal glands are inverted Y-shaped endocrine glands, which primarily mediate the stress response by releasing cortisol and catecholamines.

The adrenals are also a site of secondary sex hormone synthesis and blood pressure regulation (with aldosterone).

• The two distinct components to the adrenal glands are the cortex and the medulla, which are derived from completely different embryological origins (the cortex is derived from mesothelium; the medulla is derived from neural crest) and are susceptible to different diseases.

Adrenal cortex

• The adrenal cortex synthesizes the steroid hormones aldosterone, glucocorticoids, and androgens, which are all biochemical derivatives of cholesterol.

• Each of the three layers of the adrenal cortex synthesizes one type of hormone:

Zona glomerulosa (most superficial): Produces aldosterone.

Zona fasciculata: Produces glucocorticoids in response to pituitary adrenocorticotropic hormone (ACTH).

Zona reticularis (deepest; closest to the adrenal medulla): Produces androgens.

• Pathology of the adrenal cortex that can be diagnosed on imaging includes adrenal hyperplasia, adrenal adenoma, and adrenal cortical carcinoma.

Adrenal medulla

• The adrenal medulla is the central portion of the adrenal gland and produces the catecholamines norepinephrine and epinephrine, which are derived from tyrosine.

• Pathology of the adrenal medulla includes pheochromocytoma and the neuroblastic tumors (ganglioneuroma, ganglioneuroblastoma, and neuroblastoma). Neuroblastoma

is the most common extracranial solid tumor of childhood and is discussed in the pediatric imaging section.

Biochemical approach to adrenal lesions

• A patient may be suspected of having a hyperfunctioning adrenal lesion based on clinical symptoms or lab abnormalities. However, not all adrenal lesions produce adrenal hyperfunction.

Adrenal hyperfunction

• **Cushing syndrome** is excess cortisol production from non-pituitary disease, such as idiopathic adrenal hyperplasia, adrenal adenoma, or ectopic/paraneoplastic ACTH (e.g., from small cell lung cancer).

• **Cushing disease** is excess cortisol production driven by excessive *pituitary* ACTH.

• **Conn syndrome** is excess aldosterone production, most commonly from an adrenal adenoma, which causes hypertension and hypokalemia.

The adenomas implicated in Conn syndrome are typically small and may be difficult to detect on CT.

Localizing the side of excess hormone production with venous sampling may be a helpful diagnostic adjunct.

• **Adrenal cortical carcinoma** is a very rare adrenal malignancy that arises from the cortex and typically causes a disordered increase in all cortical adrenal hormones and precursors.

• **Pheochromocytoma** is a usually benign tumor of the adrenal medulla that causes an increase in catecholamines.

Adrenal hypofunction

• Significant destruction of the adrenals is required to produce adrenal insufficiency.

• Although usually not an imaging diagnosis, Addison disease represents chronic adrenocortical insufficiency and may be caused by autoimmune destruction of the adrenal glands or as a sequela of infection.

• Waterhouse–Friderichsen syndrome is post-hemorrhagic adrenal failure secondary to *Neisseria meningitidis* bacteremia.

• Idiopathic adrenal hemorrhage is usually unilateral and rarely causes adrenal hypofunction.

Imaging of adrenal adenoma and the indeterminate adrenal mass

Adrenal adenoma

• Adrenal adenoma is a benign tumor of the adrenal cortex. Adenomas are usually incidental, but they may occasionally produce excess aldosterone to cause secondary hypertension (Conn syndrome).

Non-contrast imaging of the adrenal glands is the best test to evaluate for the presence of an adrenal adenoma in the presence of suspicious clinical symptoms or lab values.

• A common clinical scenario is the need to differentiate between an adrenal adenoma and an adrenal metastasis in the staging of a patient with known malignancy.

The diagnosis of an adenoma is made by the detection of intracellular lipid.

• An adrenal nodule attenuating ≤10 Hounsfield units (HU) can be reliably diagnosed as an adenoma with no further imaging or follow-up needed.

Most (80%) adenomas are lipid-rich and will attenuate below this cutoff.

Up to 20% may be lipid-poor adenomas, which attenuate >10 HU and are not able to be diagnosed on a noncontrast CT.

An indeterminate (>10 HU), small, homogeneous adrenal lesion in a patient without a known malignancy is overwhelmingly

likely to represent a lipid-poor adenoma, and advanced imaging is usually not required in such cases.

• If the nodule in question attenuates >10 HU and clinical confirmation of an adenoma is necessary for clinical management (for instance, in a patient with lung cancer and

no evidence of metastatic disease but with an indeterminate adrenal nodule), then an adrenal washout CT or in- and out-of-phase MRI may be helpful to characterize the lesion.

• A *collision tumor* represents metastasis into an adrenal gland with a pre-existing adenoma.

If an “adenoma” appears heterogeneous or has shown an interval increase in size, then a collision tumor should be considered in a patient with a known primary even if a region attenuates <10 HU.

MRI adrenal imaging: Chemical shift imaging = in- and out-of-phase imaging

• Adenomas contain intracytoplasmic lipid due to steroid production.

MRI is able to detect even a small amount of intracytoplasmic lipid that may be undetectable on CT by taking advantage of the fact that protons resonate at different frequencies in fat and in water.

Chemical shift imaging consists of images obtained both in-phase and out-of-phase. When fat and water are contained within the same voxel, out-of-phase images show fat drop-out of

signal because fat protons are more shielded and resonate at a slower frequency.

Chemical shift imaging is based on T1 images.

• Adenomas suppress on out-of-phase images, while metastases generally do not.

• A short list of malignancies do contain intracytoplasmic lipid and thus would *also* lose signal on out-of-phase images:

Well-differentiated adrenocortical carcinoma (very rare).

Clear cell renal cell carcinomas metastatic to the adrenal gland.

Hepatocellular carcinoma metastatic to the adrenal gland.

Liposarcoma (typically a predominantly fatty mass that is rarely confused with adrenal adenoma).

CT Imaging: Adrenal washout CT

• Adrenal adenomas demonstrate more rapid contrast washout than metastases do.

The more rapid contrast washout of benign adenomas appears to be true even compared to adrenal metastases of hypervascular primaries.

• The timing of the washout phase remains controversial, with recent evidence suggesting 15-minute washout has greater sensitivity than 10 minutes.

• >**60% absolute washout is diagnostic of adenoma**.

Adrenal adenoma. Axial images from adrenal washout-protocol CT show an adrenal nodule measuring 16 HU precontrast, 112 HU at 60 seconds, and 46 HU on 15 minute washout.

Using the washout formula E - D/E - U:

(112 - 46) / (112 - 16) = 69% washout >60% washout is diagnostic of an adenoma.

Note is made of two large simple cysts of the left kidney.

• If unenhanced CT is not available or not performed due to concern for radiation exposure, >40% relative washout **is diagnostic of adenoma:**

• In a patient with a known primary malignancy, lesions that do not demonstrate benign washout kinetics are suspicious for, but not diagnostic of, metastasis.

Role of biopsy of an adrenal mass

• Adrenal mass biopsy is indicated for an indeterminate adrenal mass after full imaging workup remains nondiagnostic.

• Biopsy is safe and generally very accurate.

Myelolipoma

• An adrenal myelolipoma is a benign neoplasm consisting of myeloid cells (i.e., erythrocyte precursors – *not* “myo” as in muscle) and fat cells.

• An adrenal mass with any discrete focus of macroscopic fat is virtually diagnostic of a myelolipoma.

Exceedingly rare cases of adrenocortical carcinoma and metastatic carcinoma have been reported to contain macroscopic fat.

A retroperitoneal liposarcoma may mimic a myelolipoma, although liposarcoma typically presents as a large mass that may displace, rather than arise from, the adrenal.

• An adrenal myelolipoma should not be confused with a *renal angiomyolipoma (AML).*

These two entities are unrelated, although they do have similar names, are located in adjacent organs, and are both diagnosed by the presence of macroscopic fat.

Adrenal cyst

• Adrenal cysts are uncommon but have imaging characteristics typical of cysts elsewhere (thin, smooth, nonenhancing wall, and water-attenuation internal contents).

• Endothelial adrenal cysts are the most common (45%) type and may be lymphatic or angiomatous in origin.

• Pseudocysts secondary to adrenal hemorrhage represent 39% of adrenal cysts and lack an epithelial lining.

Peripheral calcification may be present.

• Epithelial cysts are rare, comprising only 9% of adrenal cysts.

• Occasionally an adrenal cyst may have a complex appearance that may be difficult to differentiate from a cystic/necrotic neoplasm.

In such a case, percutaneous aspiration or surgical resection may be considered.

• Small, asymptomatic, simple cysts can be ignored. A cyst may rarely grow so large as to cause symptoms, such as dull pain or compression of the stomach/duodenum, in which case surgery may be indicated.

• Very rarely, hydatid disease may affect the adrenal glands, typically producing a complex cystic lesion with an internal membrane.

Malignant (or potentially malignant) adrenal masses

Pheochromocytoma: Potentially malignant

• Pheochromocytoma is a neoplasm of chromaffin cells, usually arising from the adrenal medulla.

Pheochromocytoma may cause hypertension and episodic headaches/diaphoresis.

• The “rule of 10’s” is a general rule characterizing the features of pheochromocytomas:

10% are extra-adrenal.

10% are bilateral.

10% are malignant.

10% are familial or syndromic.

• Pheochromocytoma is associated with several syndromes:

Multiple endocrine neoplasia (MEN) 2A and 2B: Typically bilateral intra-adrenal

pheochromocytomas.

von Hippel–Lindau.

Neurofibromatosis type 1.

Carney's triad (gastric leiomyosarcoma, pulmonary chondroma, and extra-adrenal pheochromocytoma).

• An extra-adrenal pheochromocytoma is a paraganglioma. The most common intraabdominal location of a paraganglioma is the organ of Zuckerkandl, located at the aortic bifurcation.

A rare intra-abdominal location of a paraganglioma is the bladder, producing the distinctive clinical presentation of post-micturition syncope (syncope after urination).

• Paragangliomas occur in the head and neck in characteristic locations.

Paragangliomas of the head and neck are generally called glomus tumors and may be associated with the tympanic membrane (glomus tympanicum), the jugular foramen (glomus jugulare), the carotid body (called a carotid body tumor), or the vagus nerve (glomus vagale).

• Nuclear medicine studies can be used in the workup of pheochromocytoma.

Of note, I-123 MIBG is used for metastatic workup of adrenal pheochromocytoma and Indium-111 pentetreotide (an analog of octreotide) is used as tracer for localization of a paraganglioma.

• In theory, pheochromocytoma should be diagnosed by urine/plasma metanephrines before imaging is performed, with imaging used for localization and staging.

In clinical practice, CT is often employed based on suspicious symptoms (such as episodic hypertension or other symptoms of catecholamine excess).

• The classic MRI appearance of pheochromocytoma is a hyperintense mass on T2-weighted

images. When large, pheochromocytoma may appear heterogeneous on MRI and CT.

Adrenal cortical carcinoma

• Adrenal cortical carcinoma is a very rare malignancy, with a prevalence of approximately 1/1,000,000.

Approximately 66% are functional, producing a disordered array of hormones that may manifest as Cushing syndrome, hyperaldosteronism, and virilization.

• Adrenal cortical carcinoma usually presents on imaging as a large, heterogeneous mass.

Central necrosis and hemorrhage are typical.

Metastasis

• Autopsy studies show adrenal metastases are present in >25% of patients with a known primary.

Lung cancer and melanoma are the most common adrenal metastases.

Lymphoma

• Primary adrenal lymphoma is rare.

Diffuse adrenal disorders

Adrenal hyperplasia

• Adrenal hyperplasia is caused by prolonged stress response or ectopic ACTH secretion.

Adrenal hemorrhage

• Adrenal hemorrhage can be spontaneous or due to anticoagulation.

When secondary to anticoagulation, the hemorrhage typically occurs within the first few weeks of beginning anticoagulation. Hemorrhage involves the right adrenal gland more commonly than the left.

• Hemorrhage may appear mass-like and is often of heterogeneous attenuation on CT.

The most important clue is a new adrenal mass within a short time interval if priors are available.

• Hemorrhage does not enhance and decreases in size on follow-up studies.

Adrenal calcification

• Adrenal calcification rarely causes adrenal hypofunction. Adrenal calcification can be due to Wegener granulomatosis, tuberculosis, histoplasmosis, or old hemorrhage.

Kidneys

Diagnostic approach to a renal mass

Renal mass protocol multiphase CT

• A renal mass protocol CT consists of at least three phases of data acquisition, with each phase providing important information to aid in the diagnosis of a renal mass.

• **Unenhanced** phase: Necessary as a baseline to quantify enhancement.

• **Nephrographic** phase (100 second delay): The nephrographic phase is the critical phase for evaluating for enhancement, comparing to the unenhanced images.

• **Pyelographic** phase (15 minute delay; also called the excretory phase): The pyelographic phase is helpful for problem solving and to diagnose potential mimics of cystic renal masses.

The pyelographic phase can distinguish between hydronephrosis (will show dense opacification in the pyelographic phase) and renal sinus cysts (will not opacify).

Reflux nephropathy may cause a dilated calyx that can simulate a cystic renal mass on the nephrographic phase.

The pyelographic phase would show opacification of the dilated calyx.

The pyelographic phase is also useful to demonstrate a calyceal diverticulum and to show the relationship of a renal mass to the collecting system for surgical planning.

• Optionally, a vascular phase can be performed for presurgical planning.

Evaluating enhancement (CT and MRI)

• The presence of enhancement is the most important characteristic to distinguish between a benign and malignant non-fat-containing renal mass (a lesion containing intralesional fat is almost always a benign angiomyolipoma, even if it enhances).

• **On CT**, enhancement is quantified as the *absolute* increase in Hounsfield units on postcontrast images, compared to pre-contrast:

<10 HU: No enhancement. 10–19 HU: Equivocal enhancement. ≥20 HU: Enhancement.

• **On MRI**, enhancement is quantified as the *percent* increase in signal intensity as measured on post-contrast images:

<15%: No enhancement. 15–19%: Equivocal enhancement. ≥20%: Enhancement.

• Lesions are considered “too small to characterize” if the lesion diameter is smaller than twice the slice thickness. For instance, using 3 mm slices, a lesion less than 6 mm cannot be accurately characterized based on attenuation or enhancement.

Renal mass biopsy

• After full imaging workup is complete, there are several well-accepted indications for percutaneous renal mass biopsy:

• To distinguish renal cell carcinoma from metastasis in a patient with a known primary.

• To distinguish between renal infection and cystic neoplasm.

• To definitively diagnose a hyperdense, homogeneously enhancing mass (after MRI has been performed), which may represent a benign angiomyolipoma with minimal fat versus a renal cell carcinoma.

• To definitively diagnose a suspicious renal mass in patient with multiple comorbidities for whom nephrectomy would be high risk.

• To ensure correct tissue diagnosis prior to renal mass ablation.

Solid renal masses

Renal cell carcinoma (RCC)

• Renal cell carcinoma (RCC) is a relatively uncommon tumor that arises from the renal tubular cells.

It represents 2–3% of all cancers. Risk factors for development of RCC include smoking, acquired cystic kidney disease, von Hippel–Lindau (VHL), and tuberous sclerosis.

• **Clear cell** is the most common RCC subtype (~75%), with approximately 55% 5-year survival.

Clear cell RCC tends to enhance more avidly than the less common subtypes.

Clear cell can be sporadic or associated with von Hippel–Lindau.

• **Papillary** RCC is a hypovascular subtype, with a 5-year survival of 80–90%.

Papillary RCC tends to enhance only mildly due to its hypovascularity.

A renal “adenoma” is frequently seen on autopsy specimens and is a papillary carcinoma ≤5 mm.

• **Chromophobe** is the subtype with the best prognosis, featuring a 90% 5-year survival.

• **Collecting duct carcinoma** is rare and has a poor prognosis.

• **Medullary carcinoma** is also rare, but is known to affect mostly young adult males with sickle cell trait.

Medullary carcinoma is an extremely aggressive neoplasm, with a mean survival of 15 months, not helped by chemotherapy.

• Staging of renal cell carcinoma is based on the Robson system, which characterizes fascial extension and vascular/lymph node involvement. Stages I–III are usually resectable, although the surgical approach may need to be altered for venous invasion (stages IIIA and IIIC).

Stage I: Tumor confined to within the renal capsule.

Stage II: Tumor extends out of the renal capsule but remains confined within Gerota’s fascia.

Stage III: Vascular and/or lymph node involvement.

IIIA: Renal vein involvement or IVC involvement.

IIIB: Lymph node involvement.

IIIC: Venous *and* lymph node involvement.

Stage IVA: Tumor growth through Gerota's fascia; **Stage IVB**: Distant metastasis.

Angiomyolipoma (AML)

• Angiomyolipoma (AML) is the most common benign renal neoplasm, composed of fat, smooth muscle, and disorganized blood vessels.

The majority are sporadic, but 40% are associated with tuberous sclerosis (where AMLs are bilateral, with multiple renal cysts).

• AML has a risk of hemorrhage when large (≥4 cm), thought to be due to aneurysmal change of the vascular elements. Small, asymptomatic AMLs are not typically followed

or resected.

• A early pathognomonic imaging finding is the presence of macroscopic fat in a non-calcified renal lesion.

The non fat-containing portion enhances avidly and

homogeneously. Calcification is almost never present.

• **On MRI**, the fat component will follow retroperitoneal fat on all sequences and will saturate out on fat-saturated sequences. Intracytoplasmic lipid is not a feature of AML, so there should be no significant signal drop-out on dual-phase MRI.

• Approximately 4% of AMLs will not contain visible macroscopic fat and will appear as a hyperdense enhancing mass. MRI is the next step, with the T2-weighted images the

most useful to distinguish from renal cell carcinoma in some cases.

A T2 hyperintense mass suggests RCC (clear cell subtype) and the patient can proceed to surgery.

A T2 hypointense mass is nonspecific and can represent either RCC (papillary type) or AML with minimal fat.

Although an AML typically would enhance more avidly than a papillary RCC, biopsy is warranted for definitive diagnosis.

• AML appears hyperechoic on ultrasound, although up to 1/3 of renal cell carcinomas may also be hyperechoic and ultrasound is thus unreliable to distinguish AML from RCC.

Oncocytoma

• Oncocytoma is the most commonly resected benign renal mass and has overlapping imaging findings with renal cell carcinoma.

• Imaging features can *suggest* oncocytoma, but imaging features are not specific and cannot be reliably differentiated from RCC.

The imaging features suggestive of oncocytoma are homogeneous enhancement and a central scar.

• Complicating the pathologic diagnosis, oncocytic cells can sometimes be found in the rare chromophobe RCC subtype.

The pathologist can usually distinguish oncocytoma from the more common clear cell and papillary renal cell carcinoma subtypes.

Renal lymphoma

• Primary renal lymphoma is rare, as the kidneys do not contain native lymphoid tissue.

However, the kidneys may become involved from hematogenously disseminated disease or spread from the retroperitoneum.

• Renal involvement of lymphoma has several patterns of disease:

Multiple lymphomatous masses (most common pattern; seen in 50% of cases of renal lymphoma).

Solitary renal mass.

Diffuse lymphomatous infiltration, causing nephromegaly.

Direct extension of retroperitoneal disease.

Non-neoplastic solid renal masses

• When evaluating a potential renal mass, it’s important to always consider that an apparent solid renal mass may represent a non-neoplastic lesion.

• **Infection**, especially focal pyelonephritis, can masquerade as a solid renal mass.

Renal abscess may be difficult to differentiate on imaging from a cystic renal cell carcinoma.

• **Renal arteriovenous malformation** (AVM) will avidly enhance and can mimic a hypervascular renal mass.

One clue to the presence of an AVM would be asymmetric

enhancement of the renal vein on the affected side, due to early shunting of venous blood.

Renal pseudotumors

• Renal pseudotumors are normal variations of renal morphology that may mimic a renal mass.

• **Hypertrophied column of Bertin**: The columns (septa) of Bertin are normal structures that anchor the renal cortex to the hilum and create the separations between the renal pyramids. When hypertrophied, the columns of Bertin may mimic a renal mass.

• **Persistent fetal lobation/lobulation**: In normal fetal development, the fetal kidneys are divided into discrete lobes. Occasionally these lobulations persist in adulthood, producing an indentation of the renal cortex.

This indentation can cause an adjacent focal bulge that simulates a renal mass.

This pseudomass can usually be distinguished from a true mass by the presence of septa of Bertin on either side.

Syndromes with renal masses (all have increased risk of RCC)

von Hippel–Lindau (VHL)

• von Hippel–Lindau (VHL) is an autosomal dominant multiorgan syndrome caused by a mutation in the VHL tumor suppressor gene on chromosome 3, which leads to cysts and neoplasms in multiple organs.

• The primary manifestation of VHL in the genitourinary system is bilateral or multifocal renal cell carcinomas, most commonly the clear-cell subtype.

• Other genitourinary manifestations of VHL include multifocal pheochromocytoma and renal cysts.

• Central nervous system manifestations of VHL include hemangioblastoma of the brainstem, cerebellum, or spinal cord.

• Pancreatic and hepatic manifestations include malignant neuroendocrine pancreatic tumor, pancreatic serous cystadenoma (a benign neoplasm), and pancreatic/hepatic cysts.

Birt–Hogg–Dube

• Birt–Hogg–Dube is an autosomal dominant syndrome of dermatologic lesions, cystic lung disease, and multiple renal oncocytomas and renal cell carcinomas.

Tuberous sclerosis (TS)

• Tuberous sclerosis is an autosomal dominant neurocutaneous disease caused by a tumor suppressor gene mutation.

It manifests clinically with seizures, developmental delay, and (mostly) benign tumors in multiple organ systems.

• The most common renal manifestation of tuberous sclerosis is multiple bilateral renal angiomyolipomas (AMLs). Approximately 50% of patients with TS will have at least

one AML.

• Renal cysts can be seen in ~25%.

• The relative risk of renal cell carcinoma is increased in patients with TS, and occurs in approximately 2–3% of patients. Diagnosis of renal cell carcinoma is complicated by the abnormal kidneys that may have multiple cysts and/or AMLs.

• In the heart, the most common neoplasm is a rhabdomyoma. A cardiac rhabdomyoma may be present during fetal life and can be detected by fetal ultrasound.

• In the lung, a process of smooth muscle proliferation identical to lymphangioleiomyomatosis can occur, causing cystic replacement of lung parenchyma.

It has been suggested that the abnormal smooth muscle in the lung in patients with TS represents genetically identical metastatic smooth muscle from a renal angiomyolipoma.

Approach to a cystic renal mass

• A cystic renal mass may be neoplastic or infectious; the two most common entities to cause a cystic renal mass are renal cell carcinoma and renal abscess.

Neoplastic differential of a cystic renal mass

• **Cystic renal cell carcinoma.** Although renal cell carcinoma most commonly presents as a solid renal mass, it can also manifest as a cystic renal mass.

• **Multilocular cystic nephroma** is a benign cystic neoplasm with enhancing septa that occurs in a bimodal age distribution in baby boys and middle-aged women.

A characteristic but nonspecific feature is the propensity to herniate into the renal pelvis, causing hydronephrosis.

In adults, multilocular cystic nephroma can be indistinguishable from cystic renal cell carcinoma.

In children, multilocular cystic nephroma can be indistinguishable from cystic Wilms tumor.

• **Mixed epithelial and stromal tumor (MEST)** is a benign neoplasm composed of epithelial and mesenchymal elements, typically found in middle-aged women.

MEST may appear as either a solid or cystic mass.

Non-neoplastic differential of a cystic renal mass

• **Renal abscess** is a contained purulent collection within the kidney.

• **Hemorrhagic renal cyst**, which will not have any enhancing component.

Role of MRI in evaluation of a complex cystic renal mass

• MRI has a limited role in the evaluation of a cystic renal mass. The key advantage of MRI is more accurate enhancement characterization, as MRI does not suffer from the CT phenomenon of pseudoenhancement due to beam hardening from adjacent, densely enhancing renal parenchyma.

The increased accuracy of MRI to characterize enhancement is most useful to distinguish a Bosniak IIF from a Bosniak III lesion.

Thickening of a septation or cyst wall that shows measurable enhancement defines a Bosniak III lesion.

The Bosniak classification is discussed on the following page.

• MRI is more sensitive for detecting septations compared to CT.

• Calcifications are more difficult to detect with MRI.

Renal cysts and cystic renal masses

Simple renal cyst

• Simple renal cysts are extremely common, found in approximately 50% of patients over age 50.

A simple renal cyst is an incidental lesion that requires no follow-up, even when large.

• **On CT** a simple cyst must attenuate close to 0 Hounsfield units, not contain any enhancing components, and have a thin imperceptible wall.

• **On MRI**, a simple cyst must be hypointense on T1-weighted images, hyperintense on T2-weighted images, and not contain any enhancing component.

Renal sinus cyst

• Cysts in the renal sinus may be classified as parapelvic and peripelvic cysts.

A parapelvic cyst is a renal cortical cyst that herniates into the renal sinus.

These cysts are usually large but solitary.

Peripelvic cysts, in contrast, are lymphatic in origin and usually small and multiple.

Hyperdense cyst

• A homogeneous renal cyst with an attenuation of >70 Hounsfield units on noncontrast imaging represents a benign hyperdense cyst, likely secondary to prior hemorrhage.

• A hyperdense cyst cannot be diagnosed if only post-contrast imaging is available as there is no way to distinguish a hyperdense cyst from an enhancing renal mass.

Bosniak classification of cystic renal masses

• The Bosniak classification risk-stratifies cystic renal masses, with increasing risk for cystic renal cell carcinoma with increasing Bosniak category.

Classification is based on morphology, not size (except for hyperdense cysts in categories II and IIF).

Category I and II: No risk of malignancy. No follow-up necessary.

Category IIF: Small risk of malignancy. Imaging follow-up is needed.

Category III and IV: Surgical lesions, concerning for cystic renal cell carcinoma.

Multicystic renal disease and risk for renal neoplasm

Autosomal dominant polycystic kidney disease (ADPKD)

• Autosomal dominant polycystic kidney disease (ADPKD) is responsible for 10% of patients on long-term dialysis. Patients typically present in their third to fourth decades, initially presenting with upper abdominal pain and a clinical course of

progressive renal failure.

The kidneys may become so enlarged as to be palpable.

Hypertension and hematuria (thought to be due nephrolithiasis or rupture of a renal cyst into the collecting system) are common.

Approximately one half of patients will have a saccular aneurysm in the circle of Willis.

• **On imaging**, the kidneys are markedly enlarged and feature multiple cysts of varying attenuation or signal intensities due to hemorrhage.

• The traditional teaching is that ADPKD does not increase the risk of renal cell carcinoma.

However, some authors propose that there is a slightly increased risk.

Renal cell carcinoma associated with ADPKD tends to occur at a younger age and is more often bilateral, multifocal, and sarcomatoid.

Diagnosis of renal malignancy is complicated by the presence of multiple (often hemorrhagic) cysts and frequently concomitant renal insufficiency, which may preclude the use of intravenous contrast.

Acquired cystic kidney disease (due to end-stage kidney disease)

• Dialysis-associated cystic renal disease does have an increased risk of renal cell carcinoma (~2–3% prevalence, compared to 1/10,000 prevalence in the general population).

Renal infection and inflammation

Pyelonephritis

• Pyelonephritis is infection of renal parenchyma and is the most common bacterial infection of the kidney.

The bacteria usually are ascending from the bladder.

• The imaging findings of pyelonephritis are nonspecific and imaging may be normal in up to 75%.

Various patterns may be seen, including a unilaterally enlarged kidney, wedge-shaped or striated regions of decreased enhancement, and perinephric stranding.

The urothelium may also be thickened and hyperenhancing.

• The differential diagnosis of a unilateral enlarged kidney includes pyelonephritis, acute ureteral obstruction, renal vein thrombosis, and compensatory hypertrophy.

• A striated nephrogram describes linear lucencies extending from the renal cortex to the medulla on a contrast-enhanced study.

The differential diagnosis for a striated nephrogram is similar to that of a wedge-shaped perfusion defect and includes:

Pyelonephritis.

Renal infarct.

Renal vein thrombosis or vasculitis.

Renal contusion (typically focal).

Acute urinary obstruction.

Renal tumor (especially lymphoma if infiltrative).

Radiation nephritis.

• Focal pyelonephritis (previously called *focal lobar nephronia*) may mimic a renal mass.

• Mild hydronephrosis can be seen on the affected side, thought to be due to a bacterial endotoxin causing reduced peristalsis, and should not be confused with obstructive

uropathy.

Pyonephrosis

• Pyonephrosis is the infection of an obstructed collecting system and is colloquially referred to as “pus under pressure.” Treatment is emergent percutaneous nephrostomy.

Renal abscess

• Renal abscess is a localized purulent collection within the kidney that most commonly results from coalescence of small microabscesses in the setting of acute bacterial pyelonephritis. An abscess may simulate a cystic renal mass.

• The treatment is conservative if <3 cm and percutaneous drainage if larger.

Emphysematous pyelonephritis

• Emphysematous pyelonephritis is a severe renal infection characterized by gas replacing renal parenchyma, caused both by gas-forming organisms and renal infarction.

• Emphysematous pyelonephritis is seen almost exclusively in diabetic patients.

• Emphysematous pyelonephritis is a surgical emergency, requiring emergent nephrectomy.

Xanthogranulomatous pyelonephritis

• Xanthogranulomatous pyelonephritis (XGP) is a chronic renal infection due to obstructing calculi, leading to replacement of renal parenchyma with fibrofatty inflammatory tissue.

• *Proteus mirabilis* and *Escherichia coli* are the two most common organisms.

• The clinical presentation of XGP includes flank pain and nonspecific constitutional symptoms, such as fever and weight loss.

Anemia and hematuria are also common.

• XGP can be diffuse (85%) or localized. The localized form, also known as “tumefactive XGP,” may mimic a renal mass.

• CT is the primary modality for imaging, which demonstrates fatty replacement of the renal parenchyma, marked perinephric inflammatory standing, and staghorn calculi.

• The *bear paw* sign represents the configuration of the hypoattenuating fibrofatty masses arranged in a radial pattern, reminiscent of a bear’s paw.

• Primary differential considerations include acute obstructing calculus with pyonephrosis or renal/transitional neoplasm with calcification.

• Treatment is nephrectomy.

Renal tuberculosis

• *Mycobacterium tuberculosis* infection of the renal parenchyma results from hematogenous dissemination. Active pulmonary TB is present in approximately 10%.

• Although initial renal TB infection typically involves both kidneys, chronic changes tend to be unilateral.

• Imaging findings include parenchymal calcification, scarring, papillary necrosis, and infundibular strictures.

End-stage renal TB produces autonephrectomy and the

characteristic *putty kidney* appearance, which represents an atrophic, calcified kidney.

Nephrolithiasis and ureterolithiasis

• Nephro/ureterolithiasis is a common problem that presents with renal colic.

Hematuria is usually present, but may be absent if the stone is completely obstructing.

Calcium containing stones (comprised of calcium oxalate and phosphate, pure calcium oxalate, or the less common pure calcium phosphate stones) together represent 73% of urinary lithiasis.

• Uric acid, xanthine, matrix, pure struvite, and indinavir (seen in HIV patients on antiretroviral therapy) stones are lucent on radiographs.

Virtually all renal stones are radiopaque on CT except for indinavir stones and the very rare uncalcified matrix stones made of mucin.

• Secondary signs of ureteral obstruction from a ureteral stone include ipsilateral hydronephrosis and perinephric stranding surrounding the affected kidney.

The *soft tissue rim* sign helps to distinguish a phlebolith from a ureteral stone.

The presence of a small amount of soft tissue surrounding the calcification, thought to represent the edematous ureteral wall, suggests a ureteral stone rather than a vascular calcification.

Papillary necrosis

• Papillary necrosis is necrosis and sloughing of renal papillary tissue, which clinically can cause gross hematuria and may lead to chronic renal insufficiency.

• There are numerous causes of papillary necrosis, most commonly NSAIDs, sickle cell anemia, diabetes, and renal vein thrombosis.

The commonly used POSTCARD mnemonic may be helpful to remember all causes:

Pyelonephritis.

Obstruction.

Sickle cell disease.

Tuberculosis.

Cirrhosis.

Analgesics (NSAIDS).

Renal vein thrombosis.

Diabetes mellitus.

• On the delayed phase of CT urography, papillary necrosis causes multiple small poolings of extra-calyceal contrast adjacent to the renal calyces.

• Three classic uroradiologic signs of papillary necrosis include the *ball on tee* sign, *lobster claw* sign (not to be confused with the *bear paw* sign of xanthogranulomatous pyelonephritis), and *signet ring* sign, which describe patterns of papillary excavation.

• The *ball on tee* sign describes contrast filling a central papilla.

• The *lobster claw* sign describes contrast filling only the periphery of the papilla.

• The *signet ring* sign describes contrast surrounding the sloughed papilla.

Renal imaging patterns

Delayed (prolonged) nephrogram

• A delayed (prolonged) nephrogram describes slow renal parenchymal uptake of intravenous contrast, prolonged enhancement, and delayed urine excretion.

• A unilateral prolonged nephrogram can be due to acute ureteral obstruction, renal vein thrombosis, and renal artery stenosis.

• Bilateral prolonged nephrograms can be seen in bilateral obstruction, contrast nephropathy, systemic hypotension, and myeloma kidney.

Medullary nephrocalcinosis

• Medullary nephrocalcinosis represents calcification of the renal medullary pyramids, usually with preserved renal function. Medullary nephrocalcinosis can be caused by:

Hypercalcemic state (e.g., hyperparathyroidism, sarcoidosis, etc).

Medullary sponge kidney (cystic dilation of distal collecting ducts; may be unilateral or segmental).

Distal (type 1) **renal tubular acidosis** (RTA).

Furosemide therapy in a child.

Cortical nephrocalcinosis

• Cortical nephrocalcinosis is dystrophic peripheral calcification of the renal cortex, with sparing of the medullary pyramids.

• Causes of cortical nephrocalcinosis include:

Acute cortical necrosis.

Chronic glomerulonephritis.

Chronic transplant rejection.

Hyperoxaluria.

Alport syndrome (hereditary nephropathy and deafness).

• Cortical necrosis is a rare form of renal injury from acute ischemic necrosis of the renal cortex.

Cortical necrosis may lead to cortical nephrocalcinosis.

Chronic renal failure develops in up to 50% of patients.

• Ischemia may be due to small vessel vasospasm or systemic hypotension.

Predisposing factors include hemolytic–uremic syndrome and thrombotic microangiopathy.

Extracalyceal contrast medium

• Contrast shouldn’t normally be seen beyond the calyces on excretory urogram.

Papillary necrosis, tubular ectasia, and calyceal diverticulum may cause this appearance.

• **Tubular ectasia** causes paintbrush-like streaks of contrast that extend from the papillae into the tubules on excretory urogram. **Medullary sponge kidney** is tubular

ectasia with associated calcifications of the renal medullary pyramids.

• **Calyceal diverticulum** is an outpouching of the collecting system into the corticomedullary region.

A dependent sediment or multiple small stones may be present.

• **Papillary necrosis**, previously discussed, may also cause extracalyceal contrast.

Renal trauma

Organ Injury Scale (OIS) - American Association for the Surgery of Trauma (AAST)

• The OIS scale from the AAST is the most commonly used system for classifying renal trauma.

It is a surgical classification but correlates well with the CT findings.

• **Grade I** injury is by far the most common type of renal injury (95%) and describes a renal contusion or subcapsular hematoma. Treatment is conservative.

• **Grade II** injury is a superficial laceration (<1 cm) or confined perinephric hematoma, without urinary extravasation. Treatment is conservative.

• **Grade III** injury is a deeper laceration (>1 cm), without urinary extravasation.

Treatment is typically conservative.

A potential pitfall of a grade III injury is that a clot at the collecting system may prevent urinary extravasation initially, but urinary extravasation may occur later as the clot is lysed by urinary urokinase.

• **Grade IV** is a deep laceration that extends into the collecting system (causing urinary extravasation), or injury to the renal artery or vein with contained hemorrhage.

Urinary extravasation is typically treated with surgical repair to prevent later development of urinoma or abscess formation. Vascular grade IV injury can be treated endovascularly.

• **Grade V** is a shattered kidney, or avulsion of the renal hilum. Treatment is variable but typically surgical.

CT description of renal trauma

• The OIS classification described above is somewhat limited as there are several important renal injuries, including traumatic renal artery thrombosis, renal artery pseudoaneurysm, and ureteral avulsion (most commonly occurring at the ureteropelvic junction) that are not included in the OIS classification but which can affect prognosis.

• Traumatic renal artery thrombosis is due to tearing of the intima, which initiates thrombosis.

There is a permanent loss of renal function after approximately two hours of ischemia.

• Pseudoaneurysm is an arterial injury with a high risk of fatal rupture.

On imaging, a pseudoaneurysm will be of similar density to the aorta, with arterial enhancement in the arterial phase and washout on delays.

Treatment is endovascular embolization.

Page kidney

• A Page kidney (named after the doctor who performed experiments wrapping animal kidneys with cellophane) is a rare cause of secondary hypertension due to prior trauma.

• A subcapsular hematoma compresses the renal parenchyma and decreases its blood flow.

These altered hemodynamics induce increased renin secretion, which can lead to hypertension.

It usually takes several months for hypertension to develop.

• Imaging shows a subcapsular hematoma causing deformation and flattening of the kidney

• Percutaneous drainage of the hematoma may be effective treatment.

Ureter

Overview of ureteral imaging

CT urography (CTU) indications and protocol

• The goal of CT urography (CTU) is to evaluate the kidneys, ureters, and bladder.

The key to successful imaging is to maximally distend and opacify the ureters and bladder.

• One of the most common indications for CTU is for the evaluation of microscopic or macroscopic hematuria.

Hematuria may be caused by a urinary tract calculus, renal

mass (e.g., renal cell carcinoma), or urothelial tumor (e.g., transitional cell carcinoma).

• Protocols vary by institution. Typically, patients are given 900 mL of water PO and either 250 mL of IV saline or 10 mg of IV furosemide to optimally distend the ureters and bladder.

• In adults ≥40 years of age, CTU is performed as a three-phase exam: Unenhanced CT of the abdomen and pelvis.

Nephrographic phase through the kidneys (100 seconds after IV contrast administration).

Excretory phase of the abdomen and pelvis (15 minutes after IV contrast).

• A split-bolus, dual-phase exam decreases radiation exposure in patients under age 40:

Unenhanced CT of the abdomen and pelvis.

Combined nephrographic/excretory phase (8 minutes delay after first IV contrast bolus and 100 seconds after the second bolus).

Malignant ureteral disease

Transitional cell carcinoma (TCC)

• Although upper-tract malignancy is rare, transitional cell carcinoma is the most common ureteral neoplasm.

• The typical imaging appearance is a single filling defect on CT urography; however, multiple filling defects may be seen in 40%.

Less commonly, there may be focal thickening of the ureteral wall.

Given the propensity of transitional cell carcinoma for

multifocality, the bladder should be evaluated for a synchronous mass.

Benign ureteral masses

Fibroepithelial polyp

• Fibroepithelial polyp is the most common benign tumor of the ureter.

It typically affects the proximal ureter.

Fibroepithelial polyp features a long stalk and appears as an elongated smooth tubular lesion.

CT urography best shows the lesion on the coronal images in the pyelographic phase.

Urothelial papilloma

• Urothelial papilloma is a rare benign neoplasm that may involve the bladder or ureter.

The mass may become quite large and mimic a malignancy.

Inverted papilloma

• Inverted papilloma is a benign mass with a central core of urothelium.

Inflammatory and infectious ureteral disease

Ureteritis cystica

• Ureteritis cystica is a benign response to chronic urinary tract inflammation, such as chronic infection or stone disease.

Several small subepithelial cysts are found unilaterally in the proximal third of the ureter and renal pelvis.

Ureteritis cystica does not have any malignant potential.

• Imaging characteristically shows multiple tiny filling defects in the renal pelvis or ureter.

• The same disease entity affecting the bladder is called cystitis cystica, which shows multiple rounded contour defects at the base of the bladder.

Leukoplakia (squamous metaplasia)

• Leukoplakia, also known as squamous metaplasia, is a rare urothelial inflammatory condition named for the characteristic white patch that is produced.

Leukoplakia is not thought to be premalignant when the renal collecting system is involved, although there is an association between squamous cell carcinoma and bladder leukoplakia.

• Imaging shows a flat mass or focal thickening of the renal pelvic or ureteral wall that may produce a characteristic *corduroy* appearance.

Malacoplakia

• Malacoplakia is an inflammatory condition associated with chronic urinary tract infection (usually *Escherichia coli*) that is typically seen in middle-aged women.

It is not premalignant.

• Imaging shows multiple flat filling defects that characteristically involve the distal ureter.

Ureteral tuberculosis

• Multifocal ureteral stenoses are suggestive of ureteral tuberculosis, even more so if there is also evidence of renal tuberculosis (parenchymal calcification and scarring) and/or bladder tuberculosis (small capacity bladder with a thickened wall).

Differential diagnosis of a ureteral filling defect

• *The primary concern of a ureteral filling defect on CT urography is ureteral malignancy.*

• **Ureteral malignancy**, of which transitional cell

carcinoma is by far the most common.

• Ureteral calculus, which is almost always visible

on pre-contrast images.

• Blood clot.

• Malacoplakia (multiple flat defects).

• Leukoplakia.

• Infectious debris (e.g., a

mycetoma).

• Sloughed renal papilla.

• Benign ureteral mass (e.g.,

fibroepithelial polyp).

Structural ureteral lesions

Ureteropelvic junction obstruction (UPJ obstruction)

• Obstruction of the ureteropelvic junction (UPJ) can be either primary or secondary to infection, stones, or prior surgery.

• Primary UPJ obstruction may be due to a congenital aperistaltic segment of ureter, high insertion of the ureter on the renal pelvis, or crossing vessels causing extrinsic compression.

• The key imaging finding is a dilated renal pelvis with a normal caliber ureter.

Ureterocele

• A ureterocele is a focal dilation of the most distal portion of the ureter that protrudes into the bladder.

A ureterocele may be orthotopic or ectopic.

• An **orthotopic ureterocele** is seen with a normally inserting ureter, and is seen most commonly in adults. Orthotopic ureteroceles are also known as *simple*, *adult-type*, and *intravesicular* ureteroceles.

Orthotopic ureteroceles are usually asymptomatic.

• An **ectopic ureterocele** is seen in the setting of a duplicated collecting system, with ectopic insertion of the upper pole ureter into the bladder, and is usually diagnosed in children.

• A **pseudoureterocele** represents intussusception of the distal ureter into the bladder, which may be due to tumor, radiation cystitis, or ureterovesicular junction stone.

Bladder

Bladder stones

• Risk factors for bladder stones include urinary stasis (most commonly bladder outlet obstruction) and chronic inflammation (e.g., from infection or foreign body).

• An off-midline bladder stone should raise concern for displacement of the stone by a bladder mass or enlarged prostate, or a stone within a ureterocele or a bladder diverticulum.

Bladder transitional cell carcinoma (TCC)

• Transitional cell carcinoma is by far the most common bladder cancer.

The bladder is the most common site of malignancy in the urinary tract.

• TCC is a disease of older adults with

a male predominance.

Risk factors include smoking and aromatic amines.

It presents with painless hematuria.

• Bladder cancer spreads through the wall of the bladder.

Organ-confined disease can be divided into nonmuscle-

invasive (70%; typically resected endoscopically) and

invasive (25%; typically treated with radical cystectomy/nodal dissection).

• Metastatic bladder cancer (5%) is treated with systemic therapy.

• The presence of pelvic lymph nodes portends a poorer prognosis.

• If bladder cancer is clinically suspected, a negative CT urogram does not obviate the need for cystoscopy.

Bladder adenocarcinoma

• Adenocarcinoma of the bladder is rare but is associated with a urachal remnant.

• The fetal urachus extends from the bladder dome to the umbilicus.

It should be obliterated after birth, but may persist as a urachal anomaly.

Bladder trauma

• CT cystography is the standard test to evaluate for suspected bladder rupture.

• Full distension of the bladder is necessary to evaluate for bladder rupture.

Delayed imaging of an intravenous contrast study with opacification of excreted urine is *not* sensitive enough, and is not the standard of care.

• To perform a CT cystogram, a total volume of at least 350 mL (or as much as the patient can tolerate) of dilute water-soluble contrast (50 mL of IV contrast mixed in 500 mL of warm saline) is instilled into the bladder by gravity, with the bag raised 40 cm above the bladder.

• Male patients with bladder injury may have associated urethral injury.

If there is blood at the urethral meatus or if there is gross hematuria, a retrograde urethrogram should be performed prior to Foley catheter placement.

• Bladder injury can be classified as extraperitoneal (most common), intraperitoneal, or combined.

Extraperitoneal bladder rupture

Extraperitoneal bladder rupture: Unenhanced CT (left image) shows fluid stranding within the retroperitoneum (yellow arrows).

• Extraperitoneal bladder rupture is defined as rupture of the bladder outside of the peritoneal space.

Extraperitoneal bladder rupture is at least twice as common as

intraperitoneal rupture.

• Extraperitoneal bladder rupture is more commonly associated with pelvic fractures compared to intraperitoneal rupture. Extraperitoneal bladder rupture is typically caused by direct laceration of the bladder by a bone fragment.

• The *molar tooth* sign describes the characteristic inverted U appearance of extravasated contrast in the extraperitoneal space of Retzius, which mimics a molar tooth.

• Extraperitoneal bladder rupture is typically managed conservatively, by placement of

a urinary catheter.

Intraperitoneal bladder rupture

• Intraperitoneal bladder rupture occurs with disruption of the bladder dome and peritoneum, causing resultant extravasation of urine into the peritoneal space.

• The mechanism of intraperitoneal bladder rupture is thought to be through pressure forces on a full bladder causing bursting at the dome into the peritoneum.

• The pathognomonic imaging finding on CT cystogram is intraperitoneal contrast interdigitating between loops of bowel.

• Intraperitoneal bladder rupture is typically treated surgically.

Urethra

Male urethral anatomy

Prostatic urethra (posterior urethra)

• The prostatic urethra courses within the prostate and is lined with transitional epithelium.

• The verumontanum is a prominent ridge of smooth muscle in the posterior portion of the prostatic urethra, and is the site of the paired ejaculatory duct orifices.

The verumontanum is also the site of obstruction in posterior urethral valves in children.

The prostatic utricle is a Müllerian duct derivative and is the blind-ending male homologue of the uterus and vagina, which is also located at the verumontanum.

Membranous urethra (posterior urethra)

• The membranous urethra is the shortest, least mobile urethral segment.

• The membranous urethra is contained within the urogenital diaphragm, which contains the external urethral sphincter and the paired Cowper’s glands.

Bulbous urethra (anterior urethra)

• The bulbar urethra is the site of drainage of Cowper’s glands.

Penile urethra (anterior urethra)

• The penile urethra is the longest urethral segment. It is lined with squamous epithelium.

• The distal portion of the penile urethra is dilated at the glans penis.

This dilation is called the fossa navicularis.

• The glands of Littré are small mucous glands of the penile urethra.

Normally, small ducts would be occluded by balloon during a retrograde urethrogram, and therefore would not opacify with injected contrast.

The glands of Littré tend to opacify more prominently when inflamed in the setting of urethritis.

Imaging of the male urethra

Retrograde urethrogram (RUG)

• Retrograde urethrogram (RUG) provides excellent evaluation of the anterior urethra and may be performed to evaluate for suspected urethral injury, stricture, or fistula.

• To perform a RUG, the fossa navicularis is cannulated with a sterile balloon-tipped catheter that is inflated with 1–2 mL saline. Subsequently, approximately 10 mL of contrast is hand-injected under fluoroscopy.

Voiding cystourethrogram (VCUG)

• Voiding cystourethrogram (VCUG) best evaluates the posterior urethra and is typically performed for evaluation of bladder and voiding function.

• To perform a VCUG, a Foley catheter is sterilely placed in the bladder and subsequently contrast is instilled into the bladder. The patient initiates urination during fluoroscopy.

Urethral stricture

• Urethral strictures secondary to sexually transmitted disease (most commonly chronic urethritis from *Neisseria gonorrhoea*) occur most commonly in the bulbous urethra.

A complication of chronic urethral infection is a periurethral abscess, which may result in a urethroperineal fistula.

• Post-traumatic saddle injury strictures also tend to occur in the bulbous urethra.

• In contrast, an iatrogenic stricture from a Foley catheter tends to occur in the penile urethra at the penile–scrotal junction.

Urethral trauma

• In the setting of trauma, if there is blood at the meatus, painful urination, or inability to void, a RUG should be performed emergently.

If the RUG shows evidence of urethral injury, a suprapubic catheter is typically placed.

• There are five types of urethral injury. Type III, pictured above, is the most common, with disruption of the urogenital diaphragm and rupture of the bulbomembranous urethra.

Contrast extravasates both into the pelvis and out into the perineum in a type III injury.

Female urethra

Anatomy of the female urethra

• The female urethra is much shorter than the male urethra. Unlike the male urethra, the female urethra is not divided into discrete segments.

• The paraurethral glands of Skene are homologous to the male prostate and secrete mucus into the urethra.

The proximal third of the urethra is lined by transitional epithelium, while the distal portion of the urethra is lined with a stratified squamous epithelium.

Urethral diverticulum

• Urethral diverticulum presents clinically with postvoid dribbling, urethral pain, and dyspareunia.

Often, however, the symptoms may be vague and nonspecific.

• Diverticula are thought to arise from glandular dilation caused by inflammation and chronic infection of the paraurethral glands of Skene.

• Urethral diverticula are prone to develop calculi due to urinary stasis.

• Very rarely, adenocarcinoma may develop within a urethral diverticulum.

MRI of the prostate

Prostate anatomy

• From an imaging standpoint, there are two components to the prostate that can be resolved on MRI:

The **peripheral zone** and

the **central gland**.

The central gland refers to both the central zone and the transition zone, as they cannot be distinguished on MRI.

• In younger men, the central gland is composed mostly of the central zone; however, the transition zone enlarges as benign prostatic hyperplasia develops.

These changes result in the central gland becoming predominantly composed of transition zone in older males.

Prostate cancer

• MRI is able to clearly delineate the prostatic zonal anatomy (central gland versus peripheral zone) with T2-weighted sequences. Imaging is enhanced with an endorectal coil.

• MRI is inappropriate for screening due to cost and low sensitivity and specificity.

• The typical MRI appearance of prostate cancer is a T2 hypointense region within the T2 hyperintense peripheral zone.

• MRI may not detect all prostate cancer: Some cancer is not hypointense on T2-weighted images, central zone cancers are difficult to detect on T2-weighted images, and cancer conspicuity is decreased if the peripheral zone is not T2 hyperintense.

• MRI is also not specific: In addition to prostate cancer, the differential diagnosis of a region of peripheral zone T2 hypointensity includes prostatitis, hemorrhage, and

involutional changes from androgen-deprivation therapy. Advanced MRI techniques, such as MRI spectroscopy, dynamic contrast-enhanced imaging, and diffusion imaging may increase specificity.

• MRI spectroscopy of prostate cancer may show elevated choline and depressed citrate peaks compared to normal prostate.

• Dynamic contrast-enhanced MRI typically shows prostate cancer to have early enhancement relative to the peripheral zone.

• Prostate cancer typically shows restricted diffusion.

• The most important goal of MRI is to distinguish between surgical and nonsurgical disease.

Cancer that is contained within the gland (tumor stage T2) is generally amenable to radical prostatectomy, while cancer that has spread outside of the gland (T3 and above) is typically treated non surgically (e.g., anti-androgen and radiation therapy).

• **T-staging**:

T1: Tumor apparent by biopsy only.

T2: Tumor confined within the prostate.

T2a: <50% of one lobe; **T2b**: >50% of one lobe; **T2c**: Tumor involves both lobes.

T3: Tumor extends through the prostate capsule. May involve seminal vesicles.

T4: Tumor invades adjacent structures other than seminal vesicles.

• **N-staging**: Any regional lymph node metastasis is N1; however, extra-pelvic nodes are M1a.

• **M-staging**:

M0: No metastases.

M1a: Nonregional lymph nodes; **M1b**: Bone metastasis; **M1c**: Other metastasis.

• Staging example: T2a prostate cancer, which can be treated with radical prostatectomy.

• Staging example: T3b N1 prostate cancer, which is typically treated non surgically.

MRI of the uterus and adnexa

Uterine anatomy

Normal T2 zonal anatomy

• T2-weighted MRI can distinguish the three layers of the uterus.

• **Endometrial stripe:** Hyperintense on T2.

• **Junctional zone** (first zone of myometrium): T2 hypointense.

The hypointense T2 signal is due to the extremely compact smooth muscle.

The junctional zone should measure ≤12 mm: Thickening of the junctional zone is seen in adenomyosis.

• **Outer myometrium:** Relatively T2 hypointense, although less so than junctional zone.

Benign uterine disease

Adenomyosis

• Adenomyosis represents ectopic endometrial glands within the myometrium.

In contrast to endometriosis, the ectopic endometrial tissue seen in adenomyosis is nonfunctioning.

• Adenomyosis can present with similar symptoms to leiomyomas, with pain and bleeding.

• Adenomyosis is best seen on T2-weighted images as a thickened junctional zone (>12 mm), often with multiple small foci of T2 hyperintensity.

Borderline thickening of the junctional zone (8–12 mm) may be due to adenomyosis, but is not diagnostic of the condition.

• Focal adenomyosis may mimic a leiomyoma, appearing as a localized low-signal mass on T1- and T2-weighted images. Typically, adenomyosis features indistinct margins, in contrast to the characteristically sharp margins of a leiomyoma.

However, the imaging features between these two entities do sometimes overlap.

Leiomyoma (fibroid)

• A leiomyoma, commonly known as a fibroid, is an extremely common benign tumor of smooth muscle, which affects up to 40% of reproductive-age women.

• Fibroids are often multiple and may be intramural (within the myometrial wall), submucosal (directly underneath the endometrial mucosa), or subserosal (directly underneath the outer uterine serosa).

• Small leiomyomas are hypointense on T2-weighted images due to compact smooth muscle.

However, cystic or myxoid degeneration may increase T2 signal

heterogeneously. Carneous or hemorrhagic degeneration may appear hyperintense on T1-weighted images.

• Malignant leiomyosarcoma is very rare and may arise de-novo or from malignant degeneration of a fibroid. Imaging cannot reliably differentiate between leiomyoma and leiomyosarcoma unless clearly malignant behavior is identified (such as invasion of adjacent structures or metastases).

In the absence of obvious malignant imaging findings, an unusual-looking fibroid is overwhelmingly likely to represent a degenerating benign fibroid rather than a leiomyosarcoma.

• MRI is often performed for treatment planning prior to uterine artery embolization (UAE). Hemorrhagic or necrotic leiomyomas are not treated effectively by UAE.

Surgical myomectomy or hysterectomy would be the preferred treatment in these cases. Additionally, there is less chance of UAE success if an ovarian–uterine artery anastomosis is present.

• A uterine contraction may mimic a leiomyoma.

Malignant uterine disease

Endometrial carcinoma

• Endometrial carcinoma is the most common female gynecologic malignancy and is thought to be caused by prolonged estrogen exposure. Specific risk factors include nulliparity, hormone replacement, and Tamoxifen therapy.

• Endometrial carcinoma typically presents with post-menopausal bleeding.

• MRI can be used for staging once carcinoma is confirmed by histologic sampling.

• The presence and extent of myometrial invasion is key for staging.

In a premenopausal patient, an intact junctional zone confirms that there is no myometrial invasion.

The junctional zone cannot be distinguished in post-menopausal patients, however.

The depth of myometrial invasion highly correlates with the presence of lymph node metastasis.

• Post-contrast images demonstrate the tumor with the highest conspicuity, as endometrial cancer enhances less avidly than the surrounding myometrium.

• The FIGO (International Federation of Gynecology and Obstetrics) staging of endometrial carcinoma was revised in 2010.

Stage I: Tumor confined to the uterus. Stage IA: <50% of myometrial invasion; stage IB: >50% myometrial invasion.

Stage II: Spread to the cervical stroma, but tumor still contained within the uterus.

Involvement of the endocervical glands only is stage I.

Stage III: Spread to adnexa or uterine serosa (IIIA), vagina (IIIB), pelvic lymph nodes (IIIC1), or para-aortic lymph nodes (IIIC2).

Prognosis is worse with para-aortic nodes, even in the absence of pelvic adenopathy.

Stage IVA: Spread to bladder or bowel mucosa.

Stage IVB: Distant metastases or inguinal lymph node spread.

MRI of the cervix

Normal cervical T2 zonal anatomy

• **Endocervical canal**: T2 hyperintense due to mucin, analogous to uterine endometrium.

• **Cervical mucosa:** Intermediate T2 signal intensity.

• **Inner cervical stroma**: Very hypointense on T2, analogous to the uterine junctional zone.

Unlike the uterine junctional zone, however, the decreased T2 signal is due to compact fibrous tissue, not smooth muscle.

The superior aspect of the inner cervical stroma is continuous with junctional zone of the uterus.

Cervical carcinoma

• Cervical carcinoma is the third most common gynecologic malignancy, with a steep decline in prevalence over the past 50 years due to screening with Pap smears.

• A cervical mass >1.5 cm should be evaluated by MRI for staging.

The cervical stroma is the key landmark in the staging of cervical cancer: If tumor extends through the cervical stroma

into the **parametrium**, the cancer is stage **IIB** and treatment is typically non-surgical.

Other key findings to note are involvement of bladder or rectum, which denotes stage IV disease (if shown to extend to the mucosal surface with cystoscopy or endoscopy).

• The FIGO (International Federation of Gynecology and Obstetrics) staging of cervical cancer was revised in 2010. The new staging takes into account lymph node involvement.

Stage I: Confined to cervix or uterus. IA: Microscopic lesion. IB: Clinically visible lesion.

Stage IIA: Spread to upper 2/3 vagina, without parametrial invasion. Typically treated surgically.

Stage IIB: Parametrial invasion. Typically treated non-surgically (e.g., brachytherapy).

Stage IIIA: Spread to lower vagina.

Stage IIIB: Pelvic sidewall extension, hydronephrosis, or pelvic nodal involvement.

Stage IVA: Spread to bladder or rectum; **Stage IVB**: Distant metastasis.

Congenital uterine anomalies

• Müllerian duct anomalies may be a cause of infertility or recurrent pregnancy loss (most commonly in septate uterus). Septate and bicornuate uterus are the most common

uterine anomalies, which may be differentiated by MRI. The American Fertility Society classification of Müllerian duct anomalies is discussed in the ultrasound section.

Septate uterus

• Septate uterus is caused by incomplete resorption of the septum of fused Müllerian ducts.

• A septate uterus has a single external fundus but a fibrous or muscular septation dividing two endometrial canals. Infertility is more common in women with septate uterus compared to bicornuate uterus.

Metroplasty (resection of the septum) can be performed hysteroscopically if the septum is fibrous, or via an open approach if the septum is muscular.

Bicornuate uterus

• Bicornuate uterus is due to incomplete fusion of the Müllerian ducts.

• A bicornuate uterus describes a partially split uterus with two separate uterine fundi.

In contrast to a septate uterus, the fundus of a bicornuate uterus pinches inwards >15 mm.

• If treated, metroplasty must be performed transabdominally, which is a more invasive procedure compared to hysteroscopic metroplasty.

MRI of the adnexa

• MRI can provide additional specificity for adnexal lesions that are indeterminate on ultrasound.

Fat and hemorrhage are both hyperintense on T1-weighted images, but fat-suppressed T1-weighted imaging can distinguish between lesions containing fat (such as a mature cystic teratoma) and containing hemorrhage (such as an endometrioma).

Endometriosis

• Endometriosis represents ectopic foci of endometrial tissue that are hormonally responsive and therefore may be composed of blood products of varying ages.

• The typical MRI appearance of endometriosis is multiple hyperintense masses on T1-weighted images, which demonstrate shading (a gradient of signal intensity) on T2-weighted images. Endometriosis does not suppress on fat-saturated sequences. Less commonly, endometriosis may appear hyperintense on both T1- and T2-weighted images.

• Tiny hemorrhagic endometrial implants may only be apparent as tiny hyperintense foci on T1-weighted images.

• A ruptured endometrioma may be a cause of acute pelvic pain and may produce free fluid that is hyperintense on both T1- and T2-weighted images.

• Laparoscopy is the gold standard for evaluation of suspected endometriosis.

Mature cystic teratoma

• Also known as a dermoid cyst, mature cystic teratoma is the most common benign ovarian neoplasm in young women.

It is composed of differentiated tissue from at least two embryonic cell layers.

• A mature cystic teratoma is typically a unilocular cystic structure filled with sebaceous material, hair follicles, and other tissues.

Less commonly, a mature teratoma may appear as a heterogeneous mass or may be a solid fat-containing mass.

• A Rokitansky nodule is a solid nodule projecting into the cyst cavity, from which hair or teeth may arise.

• On imaging, the sebaceous intracystic component is typically hyperintense on T1-and T2-weighted images, matching fat intensity.

Since both an endometrioma and a teratoma are predominantly hyperintense on T1-weighted images, the fat-suppressed

sequences are key to differentiation. Teratoma will show signal loss on the fat suppressed images.

• Ovaries containing a dermoid cyst are predisposed to torsion.

Ovarian cancer

• Ovarian cancer is the second most common female pelvic malignancy but is one of the most lethal malignancies as 65% of patients present with advanced disease.

• MRI is used to characterize indeterminate adnexal masses, rather than for staging.

• The presence of a solid enhancing component, intra-lesional necrosis, ascites, or peritoneal nodularity suggests a malignant lesion, although no finding is 100% specific.

• MRI is highly sensitive to detect peritoneal implants, which occur most commonly in the pouch of Douglas, paracolic gutters, bowel surface, greater omentum, and liver surface.

• Ovarian cancer may be **epithelial**, **germ cell**, **sex-cord stromal**, or **metastatic** in origin.

• Approximately 90% of malignant tumors are of epithelial origin.

Serous tumors are the most common epithelial subtype, followed by mucinous, endometrioid, and clear cell.

Serous cystadenocarcinomas are frequently bilateral and typically appear as mixed solid and cystic masses.

The solid portions demonstrate avid enhancement.

There is often concomitant ascites.

Mucinous cystadenocarcinomas are large, most commonly unilateral, and occur in older patients compared to serous cystadenocarcinomas.

Mucinous cystadenocarcinomas typically present as a

multiloculated cystic mass containing mucin-rich T1 hyperintense fluid.

Clear cell carcinoma and less commonly **endometrioid carcinoma** are associated with endometriosis.

• Malignant **germ cell** tumors occur in younger patients and include dysgerminoma, endodermal sinus tumor, and immature teratoma.

• **Sex-cord stromal** tumors include granulosa cell (hormonally active) and Sertoli–Leydig (rare).

• **Metastases** are uncommon but may result from gastric cancer (Krukenberg tumor), colon cancer, pancreatic cancer, breast cancer, and melanoma.

Metastases are often bilateral.

Neuroimaging

Introduction

Ventricular anatomy

Cerebrospinal fluid (CSF)

Ventricular anatomy

• The ventricular system consists of two lateral ventricles and midline third and fourth ventricles.

• The foramen of Monro connects the lateral ventricles with the third ventricle.

• The cerebral aqueduct (of Sylvius) connects the third ventricle with the fourth ventricle.

• The fourth ventricle continues inferiorly as the central canal of the spinal cord.

The fourth ventricle also drains into the subarachnoid space and basal cisterns via three foramina:

Paired foramina of Luschka (Luschka is lateral).

Single foramen of Magendie (Magendie is medial).

CSF dynamics

• Cerebrospinal fluid is produced by the choroid plexus, which is located in specific locations throughout the ventricular system:

Body and temporal horn of each lateral ventricle.

Roof of third ventricle.

Roof of fourth ventricle.

There is NO choroid plexus in the cerebral aqueduct or occipital or frontal horns of the lateral ventricles.

• The ventricular volume is approximately 25 mL. The volume of the subarachnoid space is approximately 125 mL, for a total CSF volume of approximately 150 mL.

• CSF production is 500 mL/day, which completely replenishes the total CSF volume 3–4 times per day.

• CSF is absorbed primarily by the arachnoid granulations (leptomeningeal evaginations extending into the dural venous sinuses) and to a lesser extent by the lymphatic system and cerebral veins.

Cerebral edema

• Edema within the brain can be caused by cell death, altered capillary permeability, or hemodynamic forces.

Cytotoxic edema

• Cytotoxic edema is cell swelling caused by damaged molecular sodium–potassium ATPase ion pumps.

It can affect both gray and white matter.

• Cytotoxic edema is caused by cell death, most commonly due to infarct.

Water ions trapped inside swollen cells feature reduced diffusivity.

Vasogenic edema

• Vasogenic edema is interstitial edema caused by increased capillary permeability.

It is seen primarily in the white matter, as there is more interstitial space.

• Vasogenic edema is caused most commonly by neoplasm, infection, or infarct.

Interstitial edema

• Interstitial edema is caused by imbalances in CSF flow, most commonly due to obstructive hydrocephalus.

• Interstitial edema presents on imaging as periventricular fluid, often called “transependymal flow of CSF,” even though it is unlikely that the CSF actually flows across the ependymal cells lining the ventricles.

Herniation

• The total volume in the skull is fixed. Increases in intracranial pressure may lead to herniation across a dural fold.

• Herniation may be due to a mass lesion (such as a neoplasm or hematoma) or may be due to edema secondary to a large stroke. Because the volume of the posterior fossa is especially limited, cerebellar infarcts are prone to herniation.

Subfalcine herniation

• Subfalcine herniation is seen when the cingulate gyrus slides underneath the falx.

• Subfalcine herniation may rarely cause compression of the anterior cerebral artery (ACA) against the falx, resulting in infarction.

• Contralateral hydrocephalus may result from foramen of Monro obstruction, resulting in ventricular entrapment.

Transtentorial (uncal) herniation

• *Downward* transtentorial herniatition results in inferomedial displacement of the medial temporal lobe (uncus) through the tentorial notch, causing compression on the brainstem and adjacent structures.

The ipsilateral cranial nerve III (oculomotor nerve) may be compressed, leading to pupillary dilation and CN III palsy (eye is “down and out”).

Compression of the ipsilateral posterior cerebral artery (PCA) may cause medial temporal/occipital infarct.

Upper brainstem **Duret hemorrhages** are caused by shearing of perforating vessels due to downward force on the brainstem.

Compression of the contralateral cerebral peduncle against Kernohan’s notch causes a hemiparesis ipsilateral to the herniated side.

• *Upward* transtentorial herniatition is superior transtentorial herniatition of the cerebellar vermis due to posterior fossa mass effect.

The main complication of upward transtentorial herniation is obstructive hydrocephalus from aqueductal compression.

Cerebellar tonsillar herniation

• Downward displacement of the cerebellar tonsils through foramen magnum causes compression of the medulla.

• Compression of medullary respiratory centers is often fatal.

Hydrocephalus

• **Communicating hydrocephalus** is ventricular enlargement without an obstructing lesion.

Subarachnoid hemorrhage can cause communicating hydrocephalus by impeding arachnoid granulation reabsorption of CSF.

Normal pressure hydrocephalus (NPH) is a form of communicating hydrocephalus characterized by normal mean CSF pressure and the clinical triad of dementia, ataxia, and incontinence.

NPH is an important diagnosis as it is a treatable and potentially reversible cause of dementia.

Imaging typically shows enlargement of the lateral and third ventricles.

• **Noncommunicating hydrocephalus** is hydrocephalus due to an obstructing lesion, such as a third ventricular colloid cyst or a posterior fossa mass obstructing the fourth ventricle.

Intra-axial and extra-axial compartments

• An intra-axial lesion is within the brain parenchyma itself, underneath the pial membrane.

• An extra-axial lesion is external to the pial membrane. The meninges and subarachnoid space are extra-axial.

Basal cisterns

• The basal cisterns, also known as the perimesencephalic cisterns, are CSF-filled spaces surrounding

the midbrain and pons.

• Compression or effacement of the basal cisterns may be a sign of impending or actual herniation.

• In actuality, not all of the cisterns can be visualized on the same axial slice.

MRI in neuroradiology

• As discussed in the physics section, inherent tissue T1 and T2 characteristics depend on the longitudinal recovery/relaxation (T1) and transverse relaxation (T2) times of the protons in that tissue.

Any tissue signal abnormality is produced by alterations

(prolongation or shortening) of the transverse or longitudinal relaxation.

T1 shortening is hyperintense (bright) on T1-weighted images and T1 prolongation is hypointense (dark).

Conversely, T2 shortening is hypointense on T2-weighted images and T2 prolongation is hyperintense.

• It is technically incorrect to refer to image signal abnormality as “T2 hypo/hyperintense” or “T1 hypo/hyperintense” as it is the *MR image* that may exhibit signal abnormalities, rather than the proton relaxation times.

Correct terminology would include, “a lesion is hyperintense on T2-weighted images” or “a lesion demonstrates T2 prolongation.”

Conventional spin-echo T1

• Most brain lesions are hypointense on T1-weighted images due to pathologic prolongation of the longitudinal recovery.

The presence of hyperintensity on T1-weighted images (caused by T1 shortening) can be an important clue leading to a specific diagnosis.

• Causes of **T1 shortening (hyperintensity)** include:

Most commonly: **Gadolinium**, **fat**, and **proteinaceous** substance.

Some **paramagnetic stages of blood** (both intra- and extracellular methemoglobin).

Melanin.

Mineralization (copper, iron, manganese).

Slowly-flowing blood.

Calcium (rarely; when dispersed, not in bone). It is much more common for calcium to be hypointense.

Conventional spin-echo T2

• Most brain lesions are hyperintense on T2-weighted images. Water has a very long T2 relaxation constant (water is very “bright” on T2-weighted images).

Edema is a hallmark of many pathologic processes and causes T2 prolongation.

• Since most pathologic lesions are hyperintense on T2-weighted images, the clue to a specific diagnosis may be obtained when a lesion is hypointense.

• Causes of **hypointensity** on T2-weighted images include:

Most **paramagnetic stages of blood** (except hyperacute blood and extracellular methemoglobin).

Calcification.

Fibrous lesion.

Highly cellular tumors with a high nucleus:cytoplasm ratio producing low lesional water content (for instance, lymphoma and medulloblastoma).

Vascular flow-void.

Mucin. Desiccated mucin, as seen in desiccated sinus secretions, is hypointense on T2-weighted images.

Conversely, mucinous lesions in the pelvis tend to be hydrated and thus hyperintense.

Fluid attenuation inversion recovery (FLAIR)

• The FLAIR sequence is the workhorse of neuroradiology. FLAIR is a T2-weighted image with suppression of water signal based on water's T1 characteristics.

• A normal FLAIR image may appear similar to a T1-weighted image since the CSF is dark on both.

However, the signal intensities of the gray and white matter are different.

T1: Normal white matter is brighter than gray matter because the fatty myelinated white matter has a shorter T1 time.

FLAIR: White matter is darker than gray matter.

Conventional spin-echo proton density (PD)

• Proton density (PD) images are not used in many neuroradiology MRI protocols, but they do have the highest signal to noise ratio of any MRI sequence.

• PD sequences are useful in the evaluation of multiple sclerosis (MS), especially for visualization of demyelinating plaques in the posterior fossa.

Diffusion weighted images and apparent diffusion coefficient (DWI and ADC)

• Diffusion MRI is based on the principal that the Brownian motion of water protons can be imaged.

Signal is lost with increasing Brownian motion.

Free water (CSF) experiences the most signal attenuation, while many pathologic processes (primarily ischemia) cause reduced diffusivity and less signal loss.

• Diffusion MRI consists of two separate sequences — DWI (diffusion weighted imaging) and ADC (apparent diffusion coefficient), which are interpreted together to evaluate the diffusion characteristics of tissue.

• Diffusion imaging has revolutionized evaluation of cerebral infarct and is approximately 95% sensitive and specific for infarct within minutes of symptom onset.

In the setting of stroke imaging, diffusion restricted tissue represents infarction.

• **DWI** is an inherently T2-weighted sequence (obtained with an echo-planar technique).

On DWI, reduced diffusivity will be **hyperintense** (less Brownian motion g less loss of signal) and lesions are very conspicuous.

• The **ADC** map shows pure diffusion information without any T2 weighting.

In contrast to DWI, reduced diffusivity is **hypointense** on the ADC map.

Because studies have shown that readers are less sensitive to detecting reduced diffusivity using the ADC map alone, DWI is the primary sequence used to detect diffusion abnormalities.

• An important pitfall to be aware of is the phenomenon of *T2 shine through*.

Because DWI images are T2-weighted, lesions that are inherently hyperintense on T2-weighted images may also be hyperintense on DWI even without restricted diffusion.

This phenomenon is called *T2 shine through*. Correlation with the ADC map for a corresponding dark spot is essential before concluding that diffusion is restricted.

• In the brain, diffusion images are obtained in three orthogonal gradient planes to account for the inherent anisotropy of large white matter tracts.

Anisotropy is the tendency of water molecules to diffuse directionally along white matter tracts.

• The b-value is an important concept that affects the sensitivity for detecting diffusion abnormalities.

The higher the b-value, the more contrast the image will provide for detecting reduced diffusivity.

The downside to increasing the b-value is a decrease in the signal to noise ratio, unless scan time is proportionally increased for additional acquisitions.

The previously described ADC map is calculated from a set of at least two different b-value images.

• Although diffusion MRI is most commonly used to evaluate for infarct, the differential diagnosis for reduced diffusion includes:

Acute stroke.

Bacterial abscess.

Cellular tumors, such as lymphoma and medulloblastoma.

Epidermoid cyst.

Herpes encephalitis.

Creutzfeldt–Jakob disease.

Gradient recall echo (GRE)

• Gradient recall echo (GRE) captures the T2* signal. Because the 180-degree rephasing pulse is omitted, GRE images are susceptible to signal loss from magnetic field inhomogeneities.
• Hemosiderin and calcium produce inhomogeneities in the magnetic field, which creates *blooming* artifacts on GRE and makes even small lesions conspicuous.
• The differential diagnosis of multiple dark spots on GRE includes:
Hypertensive microbleeds (dark spots are primarily in the basal ganglia, thalami, cerebellum, and pons).
Cerebral amyloid angiopathy (dark spots are in the subcortical white matter, most commonly the parietal and occipital lobes).
Familial cerebral cavernous malformations (an inherited form of multiple cavernous malformations).
Axonal shear injury.
Multiple hemorrhagic metastases.

Magnetic resonance spectroscopy

• MR spectroscopy describes the chemical composition of a brain region.

In some circumstances, spectroscopy may help distinguish recurrent tumor from radiation necrosis, or may be helpful to differentiate between glioblastoma and metastasis.
Glioblastoma is an infiltrative tumor that features a gradual transition from abnormal to normal spectroscopy.
In contrast, a metastasis would be expected to have a more abrupt transition.
• The ratios of specific compounds may be altered in various disease states. *N*-acetylaspartate (NAA) is a normal marker of neuronal viability that decreases in most abnormalities. In tumors, NAA decreases and choline increases, although this pattern is nonspecific.
Creatine provides information about cellular energy stores.
The peaks of the three principle compounds analyzed occur in alphabetical order: Choline (cho), creatine (cr), and NAA. Canavan disease is a dysmyelinating disorder known for being one of the few disorders with elevated NAA.
• A lactate "doublet" may be seen in high-grade tumors indicating anaerobic metabolism.
• "Hunter's angle" is a quick way to see if a spectrum is close to normal.
A line connecting the tallest peaks should point up like a plane taking off.

Perfusion

• Perfusion MR is an advanced technique where the brain is imaged repeatedly as a bolus of gadolinium contrast is injected.
The principle of perfusion MR is based on the theory that gadolinium causes a magnetic field disturbance, which (counterintuitively) transiently *decreases* the image intensity.

• Perfusion images are echo-planar T2* images, which can be acquired very quickly.
• Perfusion MR may be used for evaluation of stroke and tumors.

Patterns of enhancement in the brain

Blood brain barrier (BBB) and enhancement

• Micro or macro disruption of the blood brain barrier (BBB) produces *parenchymal* enhancement after contrast administration, which may be secondary to infection, inflammation, neoplasm, trauma, and vascular etiologies.
• The BBB is formed by astrocytic foot processes of brain capillary endothelial cells and prevents direct communication between the systemic capillaries and the protected extracellular fluid of the brain.
• Several CNS regions do not have a blood brain barrier, and therefore normally enhance:
Choroid plexus.
Pituitary and pineal glands.
Tuber cinereum (controls circadian rhythm, located in the inferior hypothalamus).
Area postrema (controls vomiting, located at inferior aspect of 4th ventricle).
The dura also lacks a blood brain barrier, but does not normally enhance.
This phenomenon is subsequently explained in the section on pachymeningeal (dural) enhancement.
• *Vascular* enhancement is due to a localized increase in blood flow, which may be secondary to vasodilation, hyperemia, neovascularity, or arteriovenous shunting.
On CT, the arterial phase of contrast injection (for instance a CT angiogram) mostly shows intravascular enhancement.

Parenchymal enhancement, including the dural folds of the falx and tentorium, is best seen several minutes after the initial contrast bolus.

On MRI, routine contrast-enhanced sequences are obtained in the parenchymal phase, several minutes after injection.

Most intracranial vascular MRI imaging is performed with a noncontrast time of flight technique.

• Intracranial enhancement may be intra- or extra-axial. Extra-axial structures that may enhance in pathologic conditions include the dura (pachymeninges) and arachnoid (leptomeninges).

Periventricular enhancement (intra-axial)

• Enhancement of the subependymal surface can be either neoplastic, infectious, or demyelinating in etiology.

• **Primary CNS lymphoma** is a malignant B-cell neoplasm that can have diverse presentations including periventricular enhancement, solitary brain mass, or multiple brain masses.

Primary CNS lymphoma is hyperattenuating on CT and demonstrates low ADC and low signal intensity on T2-weighted MRI due to hypercellularity.

Primary CNS lymphoma rarely involves the meninges. In contrast, the meninges (both pachymeninges and leptomeninges) are commonly involved when systemic lymphoma spreads to the brain.

CNS lymphoma tends to be centrally necrotic in immunocompromised patients, but usually enhances homogeneously in immunocompetent patients.

• **Infectious ependymitis** is most commonly caused by cytomegalovirus.

Infectious ependymitis usually features thin linear enhancement along the margins of the ventricles.

• **Primary glial tumor** may cause periventricular enhancement.
• **Multiple sclerosis** may affect the subependymal surface.
Although the majority of demyelinating lesions do not enhance, an active plaque may demonstrate enhancement.

Gyriform enhancement (intra-axial)

• Superficial enhancement of the cortical (gyral) surface of the brain can be due to either cerebral infection, inflammation, or ischemia.
• Herpes encephalitis is a serious necrotizing infection of the brain parenchyma due to reactivation of latent HSV-1 infection within the trigeminal ganglion.
The medial temporal lobes and cingulate gyrus are usually affected first and demonstrate gyral enhancement due to inflammation, petechial hemorrhage, and resultant BBB breakdown.
The involved areas typically also demonstrate reduced diffusivity.
• Meningitis may cause gyral enhancement in addition to the more typical leptomeningeal enhancement (subsequently discussed).
• Subacute infarct can demonstrate gyriform enhancement lasting approximately 6 days to 6 weeks after the initial ischemic event.
In contrast to the gyriform enhancement of subacute infarct, an acute infarct may demonstrate vascular enhancement due to reactive collateral vasodilation and resultant hyperemia.
• Posterior reversible encephalopathy syndrome (PRES) is a syndrome of vasogenic white matter edema triggered by altered autoregulation that may demonstrate gyral enhancement.
PRES may rarely exhibit restricted diffusion.

Nodular subcortical enhancement (intra-axial).

• Nodular intra-axial enhancement is most commonly due to metastatic disease.

• Hematogenously disseminated metastatic disease is commonly found at the subcortical gray– white junctions.

Tumor emboli become “stuck” at the junction between the simple vasculature of the white matter and the highly branching vasculature of the gray matter.

• Edema is almost always present with metastatic disease of the gray–white junction, although slightly more distal cortical metastases may not show any edema and may be detectable only on the post-contrast images.

• In contrast to the subcortical pattern seen with arterial metastases, venous dissemination of metastasis (e.g., pelvic malignancy spread via the Batson prevertebral venous plexus) leads to posterior fossa disease by transit through the retroclival venous plexus. differential diagnosis of gyriform enhancement

• Peripheral (ring) enhancement is a common presentation with a broad range of differential diagnoses.

The two most common causes are high-grade neoplasm and cerebral abscess.

• The mnemonic MAGIC DR (metastasis, abscess, glioma, infarct, contusion, demyelination, and radiation) may be helpful to remember the wide range of etiologies for ring enhancement, although it is usually possible to narrow the differential based on the pattern of ring enhancement combined with additional MRI sequences and clinical history.

• Metastasis: Hematogenous metastases are typically found at the subcortical gray–white junction.

Metastases are often multiple, but smaller lesions may not be ring-enhancing.

• Abscess: A pyogenic abscess is formed as a result of organization and sequestration of an infection, featuring a central region of viscous necrosis.
The key imaging findings of abscess are reduced diffusivity (bright on DWI and dark on ADC) caused by high viscosity of central necrosis and a characteristic smooth, hypointense rim on T2-weighted images.
• Glioma: High grade tumors such as glioblastoma typically have a thick and irregular wall.
Multivoxel MRI spectroscopy will be abnormal outside the margin of an enhancing high grade glial neoplasm secondary to nonenhancing infiltrative tumor.
This is in contrast to a demyelinating lesion, abscess, and metastasis, where the spectral pattern returns to normal at the margin of the lesion.
Perfusion MRI demonstrates elevated perfusion in a high grade glioma.
• Infarct: Although subacute cortical infarcts often demonstrate gyral enhancement, ring enhancement can be seen in subacute basal ganglia infarcts.
In contrast to neoplasm and infection, a subacute infarct does not have significant mass effect.
• Contusion: Both traumatic and nontraumatic intraparenchymal hemorrhage can show ring enhancement in the subacute to chronic stage.
• Demyelinating disease: The key finding in ring-enhancing demyelinating disease is lack of significant mass effect.
The “ring” of enhancement is often incomplete and “C” shaped.
Multiple sclerosis is the most common demyelinating disease.
Enhancement suggests active disease.

Although the typical finding is an incomplete rim of enhancement, tumefactive demyelinating disease can look identical to a high-grade tumor.

• Radiation necrosis may look identical to a high-grade tumor. On perfusion, cerebral blood volume is generally low in radiation necrosis and typically increased in a high grade glioma.

• The pachymeninges (pachy means thick – a “thick-skinned” elephant is a pachyderm) refers to the dura mater, the thick and leather-like outermost covering of the brain.

• In addition to surrounding the surface of the brain, the dura forms several reflections, including the falx, tentorium, and cavernous sinus.

• The dura does not have a blood brain barrier. Although contrast molecules normally diffuse into the dura on enhanced CT or MRI, dural enhancement is never visualized on CT and is only visualized on MRI in pathologic situations.

Dural enhancement is not seen on CT because both the skull and adjacent enhancing dura appear white.

Enhancement of normal dura is not visible on MRI because MRI visualization of enhancement requires both water protons and gadolinium.

Although gadolinium is present in the dura, there are normally very few water protons.

However, dural pathology often causes dural edema, which provides enough water protons to make the gadolinium visible.

Therefore, **dural enhancement on MRI is an indication of dural edema rather than BBB breakdown.**

Differential diagnosis of pachymeningeal enhancement

• Intracranial hypotension: Prolonged decrease in cerebrospinal fluid pressure can lead to vasogenic edema in the dura.

Intracranial hypotension clinically presents as a postural headache exacerbated by standing upright.
Intracranial hypotension may be idiopathic or secondary to CSF leak from surgery or lumbar puncture.
Imaging shows thick, linear dural enhancement, enlargement of the pituitary gland, and “sagging” of the cerebellar tonsils. There may also be subdural hemorrhage due to traction effect on the cerebral veins.
• Postoperative: Dural enhancement may be seen postoperatively.
• Post lumbar puncture: Diffuse dural enhancement is occasionally seen (<5% of the time) after routine lumbar puncture.
• Meningeal neoplasm, such as meningioma, can produce a focal area of dural enhancement called a dural tail, due to reactive changes in the dura.
Metastatic disease to the dura, most commonly breast cancer in a female and prostate cancer in a male, can cause irregular dural enhancement.
• Granulomatous disease, including sarcoidosis, tuberculosis, and fungal disease, can produce dural enhancement, typically of the basal meninges (meninges of the skull base).
Differential diagnosis of leptomeningeal enhancement
• Meningitis (either bacterial, viral, or fungal) is the primary consideration when leptomeningeal enhancement is seen.
Leptomeningeal enhancement in meningitis is caused by BBB breakdown due to inflammation or infection.
Fine, linear enhancement suggests bacterial or viral meningitis.
Thicker, nodular enhancement suggests fungal meningitis.

• Leptomeningeal carcinomatosis, also called carcinomatous meningitis, is spread of neoplasm into the subarachnoid space, which may be due to primary brain tumor or metastatic disease.
CNS neoplasms known to cause leptomeningeal carcinomatosis include medulloblastoma, oligodendroglioma, choroid plexus tumor, lymphoma, ependymoma, glioblastoma, and germinoma. Mnemonic: MOCLEGG or GEMCLOG (courtesy W. Stephen Poole, MD).
Metastatic tumors known to cause carcinomatosis include lymphoma and breast cancer.
• Viral encephalitis may produce cranial nerve enhancement within the subarachnoid space.
• Slow vascular flow may mimic leptomeningeal enhancement at first glance, but a careful examination shows the distinction.
Slow flow appears as an intravascular distribution of FLAIR hyperintensity due to “unmasking” of the inherent high signal of blood, which remains in the plane of imaging as the entire pulse sequence is obtained.
Slow flow of peripheral vessels in moyamoya disease causes the ivy sign.
The differential diagnosis of FLAIR hyperintensity in the subarachnoid space overlaps with the differential for leptomeningeal enhancement.
Subarachnoid FLAIR hyperintensity may be due to:
Meningitis and leptomeningeal carcinomatosis both have increased subarachnoid FLAIR signal and leptomeningeal enhancement.
Subarachnoid hemorrhage manifests as increased subarachnoid FLAIR signal, without leptomeningeal enhancement. Blooming artifact on GRE or SWI from blood products will help differentiate subarachnoid hemorrhage from carcinomatosis.

Subarachnoid FLAIR signal is artifactually increased when the patient is on oxygen or propofol therapy, without abnormal enhancement.

Brain tumors

Approach to evaluation of a focal brain lesion

Are there any tumor-related complications?

• The three emergent complications of a brain tumor are the three H's: Hemorrhage, hydrocephalus, and herniation.

CT is a good screening method to evaluate for these complications.

• Hemorrhage: Primary or metastatic brain tumors are often associated with neovascularity and tumoral vessels are more prone to hemorrhage than normal vasculature.

The most common primary brain tumor to hemorrhage is a glioblastoma.

Hemorrhagic metastases include melanoma, renal cell carcinoma, thyroid carcinoma, and choriocarcinoma. Although breast and lung cancer metastases are less frequently hemorrhagic on a case-by-case basis, these two malignancies are so common that they should also be considered in the differential of a hemorrhagic metastasis.

• Hydrocephalus: A tumor can cause hydrocephalus by blocking the flow of CSF.

Posterior fossa tumors have increased risk of causing hydrocephalus by effacing the fourth ventricle.

• Herniation: The overall mass effect from tumor is a combination of the tumoral mass and associated vasogenic edema, which may contribute to brain herniation.

Is the mass intra- or extra-axial?

• After evaluation for emergent complications, the next step is to determine if the lesion is intra- or extra-axial. This distinction can sometimes be quite tricky.
• Although metastases may be either intra- or extra-axial, the differential diagnosis for each space is otherwise completely different.
• Findings of an extra-axial mass include a CSF cleft between the mass and the brain, buckling of gray matter, and gray matter interposed between the mass and white matter.
• Findings of an intra-axial mass include absence of intervening gray matter between the mass and the white matter.
• The presence of white matter edema is not specific to intra-axial masses.
In particular, meningioma (an extra-axial dural neoplasm) is known to cause white matter edema of underlying brain.
• Meningeal enhancement is seen more commonly in extra-axial masses (most commonly meningioma), but can also be seen in intra-axial masses.
Where specifically is the lesion located?
• The differential can often be narrowed depending on the location.
For instance, temporal lobe, posterior fossa, pineal, or suprasellar location may help point to a specific diagnosis.
Does the lesion enhance?
• Metastases always enhance due to tumoral neo-vessels, which lack a blood brain barrier.
• Low-grade primary infiltrating tumors may not enhance.
Primary brain tumors may form near-normal CNS capillaries with an intact BBB.
• The degree of enhancement does not correlate with the histologic grade.

Even large high grade primary brain tumors may enhance only slightly.
Conversely, benign juvenile pilocytic astrocytoma features an avidly enhancing nodule, typically associated with an adjacent cyst.
Is there more than one lesion?
• If there is more than one lesion, the diagnosis is overwhelmingly likely to be metastases.
Are there any distinctive MRI signal characteristics?
• As previously described, most brain lesions are hyperintense on T2-weighted images and hypointense on T1-weighted images.
MRI signal characteristics are most helpful if a lesion is either hypointense with T2 weighting or hyperintense with T1 weighting.
• Tumors hypointense on T2-weighted images include:
Metastases containing desiccated mucin, such as some gastrointestinal adenocarcinomas.
Note that mucinous metastases to the brain can have variable signal intensities on T2-weighted images, depending on the water content of the mucin.
Hydrated mucin is hyperintense on T2-weighted images.
Hypercellular tumors, including lymphoma, medulloblastoma, germinoma, and some glioblastomas.
• Tumors hyperintense on T1-weighted images include:
Metastatic melanoma (melanin is hyperintense on T1-weighted images).
Fat-containing tumors, such as dermoid or teratoma.
Hemorrhagic metastasis (including renal cell, thyroid, choriocarcinoma, and melanoma).

• Some tumors contain cystic components, which are isointense to CSF on all sequences.
Note that cysts tend to be at the periphery of enhancing low-grade tumors. In contrast, although intra-tumoral necrosis of a high-grade tumor may also follow CSF signal, necrosis tends to be surrounded by enhancing tumor.

Glial cells

Overview of glial cells

• A glioma is a primary CNS tumor that arises from a glial cell.

Glial cells include astrocytes, oligodendrocytes, ependymal cells, and choroid plexus cells.

• Glioma is not a synonym for a “brain tumor.”

Only a tumor that arises from one of the aforementioned glial cells can accurately be called a glioma.

Astrocyte

• The normal functions of an astrocyte are to provide biochemical support to the endothelial cells that maintain the blood brain barrier, to maintain extracellular ion balance, and to aid in repair after a neuronal injury.

• Astrocytes are normally located throughout the entire brain (primarily in the white matter) and spinal cord.

Oligodendrocyte

• The normal function of an oligodendrocyte is to maintain myelin around CNS axons.
A single oligodendrocyte can maintain the myelin of dozens of axons.

The counterpart in the peripheral nervous system is the Schwann cell, which maintains myelin around a single peripheral nerve.

Unlike the oligodendrocyte, each Schwann cell is in charge of only a single axon.

• Oligodendrocytes are normally located throughout the entire brain and spinal cord.

Ependymal cells

• The normal function of an ependymal cell is to circulate CSF with its multiple cilia.

• Ependymal cells line the ventricles and central canal of the spinal cord.

Choroid plexus cells

• The normal function of a choroid plexus cell is to produce CSF.
A choroid plexus cell is a modified ependymal cell.

• Choroid plexus cells are located intraventricularly, in the body and temporal horn of each lateral ventricle, roof of the third ventricle, and roof of the fourth ventricle.
Juvenile pilocytic (hair-like) astrocytoma (JPA) is a benign World Health Organization (WHO) grade I tumor seen typically in the posterior fossa in children.

• Imaging shows a well-circumscribed cystic mass with an enhancing nodule and relatively little edema.

When in the posterior fossa, JPA may compress the fourth ventricle.

• JPA can also occur along the optic pathway, with up to 1/3 of optic pathway JPA associated with neurofibromatosis type 1.

Posterior fossa JPA is not associated with NF1.

Fibrillary astrocytoma

• Fibrillary astrocytomas are infiltrative tumors that include low-grade astrocytoma, anaplastic astrocytoma, and glioblastoma multiforme (GBM).

• Astrocytomas can occur in the brain or the spinal cord.

Low-grade astrocytoma

• Low-grade astrocytoma is a WHO grade II tumor that typically presents as a hyperintense mass on T2-weighted images, without enhancement. Imaging findings may be subtle.

Anaplastic astrocytoma

• Anaplastic astrocytoma is a WHO grade III tumor. It features a range of appearances from thickened cortex (similar to low-grade astrocytoma) to an irregularly enhancing mass that may appear identical to glioblastoma.
The natural history of the disease is eventual progression to glioblastoma.

Glioblastoma multiforme (GBM)

• Glioblastoma multiforme (GBM) is an aggressive WHO grade IV tumor of older adults.

It is the most common primary CNS malignancy.

GBM has a highly variable appearance (“multiforme”) but typically presents as a white matter mass with heterogeneous enhancement and surrounding nonenhancing T2 prolongation.

Most of the surrounding T2 prolongation is thought to represent infiltrative tumor.

• GBM is an infiltrative disease that spreads through white matter tracts, through the CSF, and subependymally.

Subependymal spread describes spread within the walls of the ventricles under the ependymal cells.

• A GBM that crosses the midline via the corpus callosum is called a butterfly glioma.

The differential diagnosis of a transcallosal mass includes glioblastoma, lymphoma, and demyelinating disease.

Other gliomas

Gliomatosis cerebri

• Gliomatosis cerebri is a diffuse infiltrative mid-grade (WHO II or III) astrocytoma that affects multiple lobes.

• Diagnostic criteria include involvement of at least two lobes plus extra-cortical involvement of structures such as the basal ganglia, corpus callosum, brainstem, or cerebellum.

• Gliomatosis has a poor prognosis and may degenerate into GBM.

• The typical imaging appearance is diffuse T2 prolongation throughout the involved brain.

Diffuse T2 prolongation can be seen in several entities, typically in immunocompromised patients, including lymphoma, progressive multifocal leukoencephalopathy (demyelination caused by JC virus), and AIDS encephalopathy.

• Gliomatosis exerts mass effect but typically does not enhance.

Oligodendroglioma

• Oligodendroglioma is a WHO grade II tumor that usually presents as a slow-growing cortical-based mass.

• The typical patient is a young to middle-aged patient presenting with seizures.

• Oligodendrogliomas have a propensity to calcify (approximately 75% calcify).

Variants such as oligoastrocytoma and anaplastic oligodendroglioma are much more aggressive.

Oligoastrocytoma is a mixed tumor with an astrocytic component.

Although oligoastrocytoma can degenerate into GBM, typically prognosis is better than a pure GBM.

Anaplastic oligodendroglioma is indistinguishable from GBM on imaging and has a poor prognosis.

• An ependymoma is a tumor of ependymal cells that tends to occur in the posterior fossa in children and in the spinal cord in older adults.

• The pediatric posterior fossa ependymoma has been called the toothpaste tumor for its propensity to fill the fourth ventricle and squeeze through the foramina of Magendie or Luschka into the adjacent basal cisterns. Medulloblastoma, the most common

pediatric brain tumor, also usually arises in the posterior fossa but does not typically squeeze through the foramina.

• The adult spinal ependymoma can occur anywhere in the intramedullary spinal cord.

The main differential diagnosis of an intramedullary spinal cord mass is an astrocytoma, which tends to occur in younger patients.

It is not possible to reliably differentiate spinal cord ependymoma from astrocytoma on imaging.

Non-glioma primary brain tumors

Lhermitte–Duclos

• Lhermitte–Duclos, also called dysplastic cerebellar gangliocytoma, is a WHO grade I cerebellar lesion that is part hamartoma and part neoplasm.

• Lhermitte–Duclos is almost always seen in associated with Cowden syndrome (multiple hamartomas and increased risk of several cancers).

• The classical imaging finding is a corduroy or tiger-striped striated lesion in the cerebellar hemisphere.

Enhancement is rare.

Embryonal tumors

• Embryonal tumors represent a spectrum of WHO grade IV, aggressive childhood malignancies that are known as primitive neuroectodermal tumors (PNET).

Intracranial PNET tumors are more commonly located in the posterior fossa but may occur supratentorially.

Atypical teratoid/rhabdoid tumor (ATRT)

• Atypical teratoid/rhabdoid tumor (ATRT) is a WHO IV, aggressive tumor that may appear similar to medulloblastoma, but occurs in slightly younger patients.

The majority occur in the posterior fossa. ATRT is associated with malignant rhabdoid tumor of the kidney.

Medulloblastoma

• Medulloblastoma is a WHO grade IV tumor of small-blue-cell origin.

It is one of the most common pediatric brain tumors.

• Medulloblastoma most commonly occurs in the midline in the cerebellar vermis.

It is slightly hyperattenuating on CT due to its densely packed cells and is accordingly hypointense on T2-weighted images and has low ADC values.

The tumor is avidly enhancing and may appear heterogeneous due to internal hemorrhage and calcification.

The low ADC values can be a useful finding to differentiate medulloblastoma from Ependymoma and pilocytic astrocytoma, the two other most common childhood posterior fossa tumors.

• Leptomeningeal metastatic disease is present in up to 33% of patients. Sugar-coating (Zuckerguss) is icing-like enhancement over the brain surface.

Imaging of the entire brain and spine should be performed prior to surgery.

• When medulloblastoma occurs in a young adult (as opposed to a child), the tumor tends to arise eccentrically in the posterior fossa, from the cerebellar hemisphere.

Tumors with a cyst and an enhancing nodule

• A few low-grade, fluid-secreting tumors present as a cyst with an enhancing mural nodule.
Juvenile pilocytic astrocytoma

Hemangioblastoma

• Hemangioblastoma is a highly vascular WHO grade I tumor associated with von Hippel–Lindau (VHL) syndrome that occurs most commonly in the cerebellum, medulla, or spinal cord. It only rarely occurs supratentorially.

• Although associated with VHL, only 30% of patients with hemangioblastoma have VHL.

Hemangioblastoma in a patient with VHL has a worse prognosis.

• The classic appearance of hemangioblastoma is a cystic mass with an enhancing mural nodule.

Prominent vessels are often seen as tubular areas of flow void.

Less commonly, a hemangioblastoma may be solid or hemorrhagic.
• When in the spinal cord, hemangioblastoma is often associated with a syrinx.

Pleomorphic xanthoastrocytoma (PXA)

• Pleomorphic xanthoastrocytoma (PXA) is a low-grade WHO grade II astrocytoma variant.

• PXA is a rare tumor of childhood and adolescents, typically with history of chronic epilepsy.

• The most common location of PXA is the temporal lobe, where it typically presents as a supratentorial cortical cystic mass with an enhancing mural nodule.

The overlying dura may be thickened and enhancing.

• The main differential consideration, both by imaging and clinical presentation, is ganglioglioma; however, ganglioglioma does not usually cause dural thickening.

Ganglioglioma

• Ganglioglioma is a rare, slow-growing neuroglial tumor that typically presents in an adolescent or young adult with medically refractory temporal lobe epilepsy.

• Ganglioglioma characteristically appears as a temporal lobe cyst and enhancing mural nodule, often with calcification.

Ganglioglioma may cause calvarial remodeling and scalloping.

Intraventricular tumors

Central neurocytoma

• Central neurocytoma is a low-grade tumor likely of neuronal origin that occurs in young adults, from teenagers to young middle aged-patients.

Prognosis is excellent.

• Typical imaging appearance is a lobulated mass attached to the septum pellucidum, with numerous intratumoral cystic areas.

Calcification is common.

Choroid plexus papilloma/carcinoma

• Choroid plexus papilloma is a rare intraventricular tumor.

Choroid plexus papilloma is a low-grade (WHO I) neoplasm arising from choroid plexus epithelial cells.

It is the most common brain tumor in babies <1 year old, but it may also occur in adults.

• T2-weighted images show a lobulated, heterogeneous or hyperintense mass that avidly enhances on T1-weighted MRI.

• In children, the atrium of the lateral ventricle is the most common location.

• In adults, the fourth ventricle is the most common location.

• Less commonly, choroid plexus papilloma may arise from the third ventricle or cerebellomedullary angle.

• Choroid plexus papilloma and carcinoma (WHO grade III) are not reliably distinguishable.

Intraventricular meningioma

• Intraventricular meningioma appears as a solid mass, typically in the trigone of the lateral ventricle.

It tends to occur in older patients, similar to other meningiomas.

• Intraventricular meningiomas are typically hypercellular and homogeneously enhance, distinguishing them from other intraventricular neoplasms.

Subependymal giant cell astrocytoma (SEGA)

• Subependymal giant cell astrocytoma (SEGA) is a low-grade (WHO I) astrocytoma variant that is associated with tuberous sclerosis.

Other findings in tuberous sclerosis include subependymal nodules and hamartomas (cortical and subcortical).

• SEGA classically is an enhancing mass in the lateral ventricle near the foramen of Monro.

Subependymoma

• Subependymoma is a nonenhancing low-grade tumor of unclear origin thought to arise from subependymal astrocytes, ependymal cells lining the ventricles, or common precursor cells.

• Subependymoma is a tumor of middle-aged and older adults. It is often found incidentally.

• The most common locations are the obex of the 4th ventricle (inferior 4th ventricle) or at the foramen of Monro in the lateral ventricle.

The tumor usually doesn’t enhance.

• Despite their similar names, subependymoma is not related to subependymal giant cell astrocytoma (discussed above, associated with tuberous sclerosis) or with ependymoma.

CNS lymphoma

Primary CNS lymphoma (PCNSL): Overview

• Primary CNS lymphoma is lymphoma isolated to the CNS, most commonly diffuse large B-cell lymphoma.

Immature blast cells form lymphoid aggregates around small cerebral blood vessels in a periventricular location.

Note that the brain does not contain native lymphoid tissue.
• PCNSL is known to “melt away” with chemoradiation but tends to recur aggressively.

• The appearance of PCNSL depends on the immune status of the patient.

Regardless of immune status, however, key imaging findings are a periventricular location and high cellularity (hyperattenuating on CT, relatively hypointense on T2-weighted images, and reduced diffusivity).

Primary CNS lymphoma: Immunocompetent patient

• In an immunocompetent patient, PCNSL typically presents as an enhancing periventricular mass, often crossing the corpus callosum to involve both hemispheres.

Involvement of the frontal lobes and basal ganglia is most common.

• The differential diagnosis for a mass involving the corpus callosum includes lymphoma, glioblastoma multiforme, and demyelinating lesion.

• PCNSL in an immunocompetent individual usually enhances homogeneously, without central necrosis.

This is in contrast to PCNSL in an immunocompromised patient, where central necrosis is typical.

Primary CNS lymphoma: Immunocompromised patient

• In an immunocompromised patient, PCNSL typically presents as a periventricular ring-enhancing lesion in the basal ganglia.

The ring enhancement is caused by central necrosis.

The two primary differential considerations for a ring-enhancing basal ganglial mass in an immunocompromised patient are lymphoma and toxoplasmosis.

• Several clinical and imaging options are available to differentiate between lymphoma and toxoplasmosis:

Empirical anti-toxoplasmosis therapy and short-interval follow-up.

Thallium scanning: CNS lymphoma is thallium avid and toxoplasmosis does not take up thallium.

PET: CNS lymphoma tends to be high-grade and metabolically active.
Toxoplasmosis usually does not have avid FDG uptake.

Perfusion scanning: CNS lymphoma has increased relative cerebral blood volume while toxoplasmosis is hypovascular.

Note that lymphoma and toxoplasmosis cannot be reliably differentiated by enhancement. Intra-axial enhancement is a measure of capillary leakage, not perfusion.

Both will enhance.

Secondary CNS lymphoma

• Secondary CNS lymphoma represents involvement of the CNS in a patient with known extra-cerebral lymphoma.

Secondary CNS lymphoma tends to involve the meninges and may cause leptomeningeal carcinomatosis or epidural cord compression.

• Less commonly, secondary CNS involvement of lymphoma may present as a parenchymal mass.

Metastatic disease to the brain

Intra-axial metastasis

• The most common primary tumors to cause parenchymal metastases are lung, breast, and melanoma.

• Most metastases are hematogenous and arise at the gray–white junction, where there is a caliber change in the small arterioles.

• Enhancement is universal, as capillaries produced by an extra-CNS primary tumor do not have a functioning blood brain barrier.

• Larger metastases often feature marked edema, while small metastases may present as tiny enhancing foci that are apparent only on the post-contrast images.

Dural neoplasms

Meningioma

• Meningioma is by far the most common extra-axial tumor. It arises from meningoepithelial cells called arachnoid “cap” cells.

Meningiomas typically occur in elderly adults with a female predominance and are most often asymptomatic.

• The vast majority are benign, but 1–2% are anaplastic or malignant.

Both benign and malignant meningiomas can metastasize, although this is uncommon.

• Multiple meningiomas are seen in neurofibromatosis type 2 or following radiation therapy.

• Meningiomas can occur anywhere in the neuraxis, but are most commonly supratentorial and parasagittal.

• Meningiomas may also be intraventricular (in the trigone/atrium of the lateral ventricle) or intra-osseous.

Intra-osseous meningioma may mimic fibrous dysplasia.

• **On noncontrast CT**, meningiomas are usually hyperattenuating relative to brain and approximately 25% calcify.

• **On MRI**, appearance can be variable with iso- or slightly hypointense signal on T1-weighted images and variable signal intensity on T2-weighted images.

There is typically a broad-based attachment to the dura.

• Meningiomas avidly enhance. An enhancing dural tail is thought to be due to vasoactive substances released by the meningioma rather than tumor spread to the dura.

• Despite the extra-axial location of most meningiomas, there may be extensive white matter edema, thought to be due to vasoactive factors and a pial vascular supply.

There is often a discordance between the size of meningioma and degree of white matter edema, with severe edema possible even with a very small tumor.

Dural metastasis

• The most common tumors to metastasize to the dura are breast (most common), lymphoma, small cell lung cancer, and melanoma.

Cerebellopontine angle (CPA) mass

Overview and anatomy of the CPA

• The cerebellopontine angle (CPA) is region between the pons and cerebellum and the posterior aspect of the petrous temporal bone.

Important structures of the CPA include the 5th (trigeminal), 7th (facial), and 8th (vestibulocochlear) cranial nerves, and the anterior inferior cerebellar artery (AICA).

• Most lesions of the CPA are extra-axial and located in the CPA cistern itself, although some may arise in the internal auditory canal (IAC), temporal bone, or rarely intraaxially from the pons or cerebellum.

CPA masses are more common in adults.

Schwannoma

• Schwannoma of the vestibulocochlear nerve, also known as a vestibular schwannoma, is by far the most common cerebellopontine angle mass, representing greater than 75% of all CPA masses.

• Vestibular schwannoma is hyperintense on T2-weighted images and avidly enhances.

The characteristic ice cream cone appearance describes the "cone" protruding through (and widening) the porus acousticus and the "ice cream" exerting mass effect on the cerebellar–pontine junction.

Schwannoma may become cystic, especially when larger.

• Schwannomas of other cranial nerves in the CPA, including the facial or trigeminal nerves, are less common.

Trigeminal schwannoma may extend into Meckel's cave.

Meningioma

• Although meningioma is overall the most common extra-axial mass in adults, it is only the second most common mass of the CPA, representing approximately 10–15% of all CPA masses.

• Meningiomas often feature a short segment of dural enhancement and may induce adjacent bony hyperostosis.

Approximately 20% calcify, in contrast to schwannomas where calcification is rare.

• In contrast to schwannoma, a CPA meningioma does not enlarge the porus acousticus.

Arachnoid cyst

• An arachnoid cyst is a benign CSF-filled lesion that is usually congenital.

Although most arachnoid cysts are supratentorial, the cerebellopontine angle is the most common infratentorial location.

• An arachnoid cyst will follow CSF signal on all sequences, including complete suppression on FLAIR.

Unlike an epidermoid cyst, an arachnoid cyst does not have restricted diffusion.

Aneurysm

• Vertebrobasilar aneurysm (arising from the posterior inferior cerebellar artery, anterior inferior cerebellar artery, vertebral artery, or basilar artery) may appear as a well-defined, avidly enhancing CPA lesion and may be initially mistaken for a schwannoma or meningioma on contrast-enhanced CT.

• **On MRI**, clues to a vascular etiology would be flow void and pulsation artifacts.

MRA or CTA are diagnostic.

Epidermoid cyst

• An epidermoid cyst is a congenital lesion arising from ectopic ectodermal epithelial tissue.

• Epidermoids progressively enlarge from desquamation of keratinized epithelium lining the cyst.

The mass characteristically insinuates in between structures, encasing cranial nerves and vessels.

Gross pathology features a characteristic “cauliflower-like” surface.

• **On CT**, epidermoid cyst may mimic arachnoid cyst and appear as a water-attenuation cystic structure.

On MRI, an epidermoid cyst has similar signal characteristics to

CSF on T1- and T2-weighted images. Unlike arachnoid cyst, an epidermoid does not usually suppress on FLAIR.

• Diffusion sequences show very bright signal on diffusion-weighted images (a combination of restricted diffusion and T2 shine through).

Postsurgical DWI follow-up is critical to detect any residual focus, which will be DWI bright.

• Rarely, epidermoids may exhibit signal hyperintensity on unenhanced T1-weighted imaging, also known as “white epidermoids.”

Intra-axial neoplasm

• A posterior fossa intra-axial neoplasm may invade laterally into the CPA.

• An exophytic brainstem glioma or metastasis may invade into the CPA.

• Medulloblastoma tends to occur in the midline in children, though lateral involvement of the cerebellar hemispheres can be seen in older children or young adults.

• Ependymoma may extend into the CPA by squeezing through the lateral 4th ventricular foramina (of Luschka).

• Hemangioblastoma, associated with von Hippel–Lindau (VHL) disease, typically presents in the cerebellar hemisphere

as a fluid-secreting tumor with a cyst and enhancing nodule. There are often prominent flow voids feeding the tumor.

The other intracranial manifestation of VHL seen on imaging is an endolymphatic sac tumor, which occurs along the posterior petrous ridge.

Overview and anatomy of the sella, suprasellar region, and cavernous sinus

• The pituitary gland is formed from Rathke's pouch, which is a superior invagination from the primitive oral cavity.

The pituitary gland sits in the sella turcica, a cup-shaped depression of the basisphenoid bone. The pituitary is composed of an anterior and a posterior lobe.

Rathke's pouch closes off to form a vesicle that involutes.

Sometimes, the involution is incomplete and a cleft can be left behind, which may give rise to craniopharyngioma or Rathke's cleft cyst.

• The anterior lobe of the pituitary produces and secretes endocrine hormones, including growth hormone, ACTH, prolactin, TSH, FSH, and LH.

• The posterior lobe of the pituitary is derived from neuroectoderm and is composed of axons from the hypothalamus, through which vasopressin and oxytocin are transported.

• The pituitary gland has a wide range of normal morphology, depending on patient age, sex, and hormonal/pregnancy status.

The gland may be convex superiorly in adolescent or pregnant females.

The normal posterior pituitary is hyperintense on T1-weighted MRI and is called the “posterior pituitary bright spot,” best seen on sagittal images.

• The empty sella is a normal variant when seen in isolation. An empty sella is partially filled with CSF, with the gland flattened against the floor of the sella.

Empty sella is also a component of the constellation of findings in pseudotumor cerebri.

Pseudotumor cerebri, also known as idiopathic intracranial hypertension, is a syndrome associated with elevated CSF pressure, visual changes, and headaches that is typically seen in obese black females.

Imaging findings include empty sella, enlargement of Meckel’s cave, and optic disc protrusion into the posterior globes.

The ventricles are normal in size or slightly reduced in caliber.

The sigmoid or transverse sinus may be stenotic.

Approach to a sellar/parasellar mass

• The first step in evaluation of a sellar region mass is to determine if the mass is intrinsic to the pituitary or if the mass represents an adjacent extra-pituitary lesion.

• The differential for an intrinsic pituitary mass is rather limited and includes pituitary adenoma (by far the most common intrinsic pituitary mass), Rathke's cleft cyst, and hypophysitis (inflammation of the pituitary).

Craniopharyngioma may rarely occur in the sella, but essentially never occurs within the pituitary gland itself.

Intrinsic pituitary mass

Pituitary microadenoma

• A pituitary microadenoma is a pituitary adenoma <10 mm in size.

Patients seek medical attention due to symptoms of hormone excess, not mass effect.

• Most microadenomas are hypoenhancing relative to the pituitary, although ACTHsecreting adenomas may be hyperenhancing.

Pituitary macroadenoma

• A macroadenoma is defined as an adenoma >10 mm in size.

Patients usually present with mass effect (e.g., compression of the optic chiasm) rather than endocrine dysfunction.

• The bony sella is often enlarged. Macroadenomas may encase the carotid, but tend not to narrow it.

In contrast, meningiomas or metastases can narrow the carotid.

Pituitary macroadenoma may bleed after medical treatment, producing a complex MRI appearance.

Intra-tumoral hemorrhage is distinct from pituitary apoplexy.

Pituitary apoplexy is a clinical syndrome of severe headache and endocrine dysfunction caused by hemorrhage into an otherwise normal pituitary.

Lymphocytic hypophysitis

• Lymphocytic hypophysitis is an autoimmune inflammatory disorder usually seen in peripartum women that may affect the pituitary and infundibulum.

It presents with diabetes insipidus, headache, visual impairment, and endocrine dysfunction.

• **MRI shows** thickening and intense enhancement of the pituitary stalk, usually with enlargement of the pituitary gland that may appear similar to a macroadenoma.

• Lymphocytic hypophysitis responds to steroid therapy.

Granulomatous hypophysitis

• Granulomatous inflammation of the pituitary and infundibulum can be secondary to sarcoidosis, Wegener granulomatosis, tuberculosis, and Langerhans cell histiocytosis (LCH).

LCH hypophysitis is a disease of children. In all causes, imaging is identical to lymphocytic hypophysitis.

Rathke's cleft cyst

• Rathke's cleft cyst may be limited to the pituitary gland but is more commonly seen extrinsic to the pituitary.

Suprasellar mass

• The differential diagnosis for an extra-pituitary lesion is broad, but the imaging findings together with clues about the patient's age and clinical presentation can usually narrow the differential diagnosis to a few entities.

• The most common suprasellar lesion in a child is craniopharyngioma, while the most common suprasellar lesion in an adult is a pituitary macroadenoma that has extended superiorly.

• The SATCHMO mnemonic may be helpful to remember the spectrum of extrapituitary masses; however, the order of the entities is NOT based on frequency of occurrence.

Sarcoidosis/Suprasellar extension of an adenoma.

Aneurysm.

Teratoma (dermoid cyst)/Tolosa Hunt.

Craniopharyngioma/Cleft cyst (Rathke's).

Hypothalamic glioma (adults)/Hypothalamic hamartoma (children).

Meningioma/Metastasis.

Optic nerve glioma.

Craniopharyngioma

• Craniopharyngioma is the most common suprasellar lesion of childhood, arising from squamous epithelial remnants of Rathke's pouch that produce keratin.

• Craniopharyngioma occurs in a bimodal age distribution.

The majority of cases are lesions of childhood, but craniopharyngioma may occur uncommonly in late middle age.

• Most involve both the sella and suprasellar regions. Although craniopharyngioma may rarely involve only the sella, it is almost always separate from the pituitary gland.

• Craniopharyngioma has potential for enamel production and almost always calcifies.

The characteristic intracystic machine-oil seen on gross examination is composed of desquamated squamous epithelium, keratin, and cholesterol.

• **MRI shows** a complex cystic mass containing protein or blood products (hyperintense on T1-weighted images).

• There is avid enhancement of the solid elements and cyst walls.

• In contrast to Rathke's cleft cyst, craniopharyngioma almost always enhances, is almost always calcified, and is almost always separate from the pituitary.

Rathke's cleft cyst

• Similar to craniopharyngioma, Rathke's cleft cyst is also a remnant of the embryologic Rathke's pouch, the precursor of the anterior lobe of the pituitary gland.

In contrast to craniopharyngioma, Rathke’s cleft cyst is made of simple columnar or cuboidal epithelium.

• While craniopharyngioma is the most common suprasellar lesion of childhood, Rathke’s cleft cyst is typically seen in middle-aged adults, twice as commonly in females.

• Rathke’s cleft cyst is reportedly very common in autopsy studies (up to 22% incidence), but clinically is usually asymptomatic or discovered incidentally.

• **Imaging appearance** is dependent on the protein content of the cyst.

The intra-cystic fluid may be isointense to CSF if low protein and hyperintense on T1-weighted images if high protein.

High protein content may cause incomplete nulling of the intracystic fluid on FLAIR.

• The claw sign represents enhancing pituitary tissue completely wrapped around the cyst.

• It is usually possible to distinguish craniopharyngioma from Rathke’s cleft cyst.

Unlike craniopharyngioma, Rathke’s cleft cyst does not enhance (although rim enhancement is often seen) and does not calcify. Rathke’s cleft cyst may occasionally be inseparable from the pituitary, but craniopharyngioma is nearly always distinct.

Meningioma

• Meningioma is the second most common suprasellar tumor in adults.

Most common in middle-aged females, it typically presents with visual loss due to optic pathway involvement.

There are several dural reflections in the region of the sella from which a meningioma may arise, including the tuberculum sella, clinoid process, planum sphenoidale, and sphenoid wing.

• **Imaging shows** isointense signal on T1-weighted images, variable signal on T2-weighted images, and uniform, intense contrast enhancement.

There is often an enhancing dural tail.

Meningioma may cause adjacent hyperostosis due to vasoactive factors.

• An important imaging finding of a parasellar meningioma is the tendency to encase and narrow the cavernous or supraclinoid internal carotid artery.

• In contrast to pituitary adenoma, the sella is usually normal and the pituitary can be identified separately.

Astrocytoma (optic pathway glioma)

• An astrocytoma involving the visual pathway (optic nerve, optic chiasm, and optic tract) is the second most common suprasellar mass in children (craniopharyngioma is most common).

A substantial minority of patients with optic pathway glioma have neurofibromatosis type 1.

In contrast to the low-grade tumor of childhood, optic glioma is an aggressive tumor when it occurs in adults.

• Tumors are isointense on T1-weighted images, hyperintense on T2-weighted images, and usually enhance.

Germinoma

• The most common intracranial germ cell tumor is a germinoma, of which 80% arise in the pineal region and 20% arise in the parasellar region.

Germinomas are primarily seen in children and adolescents.

• **Imaging shows** a homogeneous, intensely enhancing midline mass.

The mass is hypointense on T2-weighted images and dark on ADC map due to hypercellularity.

Epidermoid and dermoid cysts

• Epidermoid and dermoid cysts are congenital benign inclusion cysts.

• Epidermoids occur most commonly in middle-aged adults in the cerebellopontine angle, but can be seen less commonly in the parasellar region.

Epidermoids follow CSF signal on T1- and T2-weighted images. In contrast to a simple arachnoid cyst, epidermoid is hyperintense on FLAIR and diffusion sequences show restricted diffusion.

• Dermoids are most common in young adult males in the posterior fossa, but may occasionally occur in the parasellar region.

They may contain intracystic fat which can cause chemical meningitis or ventriculitis with rupture.

Aneurysm

• A saccular supraclinoid internal carotid artery aneurysm may mimic a suprasellar tumor.

• Although parasellar aneurysms are relatively uncommon, it is essential never to biopsy a mass that may represent an aneurysm.

• Pulsation artifact may be present on conventional MRI sequences.

CTA or MRA would be diagnostic.

Hypothalamic hamartoma (hamartoma of the tuber cinereum)

• Hypothalamic hamartoma is not a true neoplasm, but represents ectopic hypothalamic neural tissue.

It is a rare lesion of childhood that classically presents with precocious puberty and gelastic seizures (laughing spells).

• Hypothalamic hamartoma characteristically appears as a sessile mass between the pituitary stalk and the mammillary bodies.

• Hypothalamic hamartoma does not enhance and is isointense to gray matter.

Metastases

• Breast cancer is by far the most common lesion to metastasize to the parasellar region.

Lymphoma

• Parasellar lymphoma is rare but may occur in older adults.

Intrinsic pineal mass

Overview and anatomy of the pineal region

• The pineal gland is located in the midline at the level of the midbrain.

It is situated between the thalami at the posterior aspect of the third ventricle.

The internal cerebral veins and vein of Galen are located superior and posterior to the pineal gland, respectively.

• The principal neuronal cell of the pineal gland is the pinealocyte, which is a modified retinal neuronal cell that is innervated by the sympathetic plexus originating in the retina.

The pineal gland releases melatonin, which modulates the sleep/wake cycle.

The pineal gland does not have a blood brain barrier.

• A mass lesion in the pineal region may cause compression of the midbrain, compression of the cerebral aqueduct of Sylvius, or compression of the tectal plate.

Compression of the tectal plate produces Parinaud syndrome, which is the inability to look up (upward gaze paralysis), pupillary light dissociation, and nystagmus.

• The first step in evaluating a pineal region mass is to determine if the lesion is arising from the pineal gland itself or from an adjacent structure.

• A mass of the pineal gland is extra-axial.

• Tumors of pineal cell origin tend to lift the internal cerebral veins, while tentorial meningiomas tend to depress the internal cerebral veins.

The relationship of any pineal region mass to the internal cerebral veins is key for surgical planning and approach.

Germ cell tumor

• Extragonadal germ cell tumors can be found in the pineal gland as well as other intracranial and extracranial midline locations including the suprasellar region, mediastinum, and sacrococcygeal region.

Extragonadal germ cell tumors are thought to be due to aberrant migration of totipotent germ cells during early embryogenesis.

• Germinoma and teratoma are germ cell tumors, which are the most common and second most common pineal region tumors, respectively.

• Germinoma (extra-gonadal seminoma) is the most common pineal region tumor and has a peak incidence in the second decade of life (age 10-19).

Pineal germinoma is seen much more commonly in males, but suprasellar germinoma does not show a gender predilection.

Germinoma is a highly cellular, avidly enhancing tumor that is slightly hyperdense on CT, isointense on T1- and T2-weighted images, and is dark on ADC map.

Germinoma characteristically “engulfs” the pineal gland and promotes its calcification, resulting in a central area of calcification.

Imaging of the entire neuraxis is recommended as leptomeningeal deposits can occur.

Treatment is radiotherapy, with excellent prognosis.

Pineal germinoma may present with a synchronous suprasellar germ cell tumor.

• Teratoma is the second most common pineal region tumor and confers a worse prognosis than germinoma.

Teratoma has a heterogeneous imaging appearance.

Intralesional fat is suggestive of teratoma. Teratoma is prone to hemorrhage and coarse calcification.

Pineal cyst

• Pineal cysts are seen commonly on MRI and have a prevalence as high as 40% on autopsy series.
They are more common in women. Most pineal cysts are less than 1 cm and asymptomatic, but may cause symptoms due to mass effect.

• Very few pineal cysts grow and follow-up of small pineal cysts is not routinely recommended.

• Pineal cysts are usually not entirely simple. Most cysts do not fully suppress on FLAIR.

Most cysts do display some peripheral enhancement, and rim calcification can be seen about 25% of the time.

• A differential consideration is a pineocytoma, which would demonstrate internal enhancement and may have cystic components.
However, a truly cystic pineocytoma is considered very rare.

A potential pitfall is that if imaging is delayed after contrast administration (by greater than 60 minutes), gadolinium may diffuse into the cyst, causing it to appear solid.

In rare cases, it may not be possible to differentiate a hemorrhagic pineal cyst from pineocytoma.

Pineocytoma

• A pineocytoma is a low-grade (WHO grade I or II), slow-growing pinealocyte tumor.

• Any solid component should enhance. Pineocytoma may feature cystic change, however, which can make differentiation from a pineal cyst difficult.

Pineoblastoma

• Pineoblastoma is a highly malignant WHO grade IV tumor of young children, of the same primitive neuroectodermal tumor (PNET) type as medulloblastoma.

• The term trilateral retinoblastoma is used when bilateral retinoblastomas are also present (both the retina and pineal gland are light-sensing organs).

The sella is additionally involved in quadrilateral retinoblastoma.

• Pineoblastoma often presents with obstructive hydrocephalus.

Pineoblastoma

Characteristically appears as a poorly defined pineal mass that may invade into adjacent structures.

High cellularity causes restricted diffusion.

• In contrast to germinoma, which “engulfs” and induces calcification of the pineal gland, pineoblastoma peripherally calcifies in a pattern that has been likened to “exploded” calcification.

• Pineoblastoma has a propensity for leptomeningeal metastasis and CSF seeding.

Metastases

• Due to the lack of a blood brain barrier, metastases to the pineal gland occur relatively commonly, but rarely in the absence of a known malignancy.

• Leptomeningeal disease is present in two-thirds of patients with pineal metastasis.

Pineal region mass (extra-pineal)

Gliomas

• Gliomas (most commonly astrocytomas) of varying grade may occur in adjacent intraaxial structures such as the tectum, midbrain, or splenium of the corpus callosum.

Vein of Galen aneurysm

• Despite the name, a vein of Galen “aneurysm” is not a true aneurysm.

Instead, it represents dilation of the vein of Galen due to an arteriovenous fistula between the anterior or posterior circulation and the venous plexus leading to the vein of Galen.

Meningioma

• The tentorial apex, adjacent to the pineal gland, is a characteristic location for meningioma.

• As previously discussed, a tentorial meningioma tends to depress the internal cerebral veins, in contrast to a pineal-based mass, which typically elevates the internal cerebral veins.

Quadrigeminal plate lipoma

• A lipoma of the quadrigeminal plate is a rare lesion that be seen in isolation or associated with agenesis or hypoplasia of the corpus callosum.

• The quadrigeminal plate is another name for the tectum.

Cerebral trauma

Extra-axial hemorrhage

• Acute extra-axial hemorrhage (subarachnoid, epidural, or subdural in location) is usually hyperattenuating when imaged by CT; however, blood must clot in order to be hyperattenuating.

Hyperacute unclotted blood (and clotted blood in a patient with severe anemia) may be close to water attenuation on CT.

Subarachnoid hemorrhage (SAH)

• Trauma is the most common cause of subarachnoid hemorrhage (SAH), while aneurysm rupture is the most common cause of non-traumatic SAH.

• Traumatic SAH tends to occur contralateral to the side of direct impact, most often in the superficial cerebral sulci.

Epidural hematoma

• An arterial epidural hematoma is an extra-axial collection of blood external to the dura, classically caused by fracture of the squamous portion of the temporal bone and resultant tearing of the middle meningeal artery.

• An arterial epidural hematoma has a lentiform shape and does not cross the cranial sutures, where the dura is tightly adherent to the cranium.

• The swirl sign describes mixed high and low attenuation blood within the hematoma and suggests active bleeding.

The low attenuation blood is hyperacute unclotted blood while the high attenuation blood is already clotted.

• A large epidural hematoma is a surgical emergency due to mass effect and risk of herniation, although small epidural hematomas can be conservatively managed with serial imaging.

• Venous epidural hematomas are far less common than arterial epidurals and are due to laceration of the dural sinuses, usually occurring in the posterior fossa in children.

Subdural hematoma

• A subdural hematoma is a crescentic extra-axial collection of blood located beneath the dura.

Since it is underneath the dura, the hematoma can extend across the cranial sutures.

Subdural hematomas often extend along the surfaces of the falx cerebri and tentorium cerebelli.

• Subdural hematomas typically result from tearing of cerebral veins.

Patients with atrophic involutional changes are at increased risk of subdural hematoma with even minor trauma, as the cerebral veins stretch to traverse the enlarged CSF spaces.

• A particular danger is a subdural hematoma in a patient with a ventricular shunt because the shunted ventricular system does not function as a natural tamponade.

• An isodense subdural hematoma is isoattenuating to gray matter.

This occurs in the subacute phase approximately 1–3 weeks after the initial injury.

Three important clues alerting to the presence of an isodense subdural are increased mass effect, white matter buckling, and an apparently thickened cortex.

Intraventricular hemorrhage

• Intraventricular hemorrhage can occur due to tearing of subependymal veins or from direct extension of subarachnoid or intraparenchymal hematoma.

• Patients with intraventricular hemorrhage are at increased risk of developing noncommunicating hydrocephalus due to ependymal scarring, which may obstruct the cerebral aqueduct.

Intra-axial injury

Coup/contrecoup mechanism

• The coup/contrecoup mechanism of brain trauma describes the propensity for brain to be injured both at the initial site of impact and 180° opposite the impact site, due to secondary impaction against the cranial vault.

Cortical contusion

• A cortical contusion is caused by traumatic contact of the cortical surface of the brain against the rough inner table of the skull.

Contusions affect the gyral crests and can occur in a coup or a contrecoup location.

• A subacute cortical contusion may demonstrate ring enhancement and should be considered in the differential of a ring enhancing lesion if there is a history of trauma.

Enhancement may continue into the chronic stage.

• A chronic contusion appears as encephalomalacia on CT. MRI is more specific, showing peripheral hemosiderin deposition as hypointense on T2-weighted images and blooming artifact on gradient echo sequences.

Intraparenchymal hematoma

• Traumatic intraparenchymal hematoma can occur in various locations, ranging from cortical contusion to basal ganglial hemorrhage (due to shearing of lenticulostriate vessels).

• Similar to a cortical contusion, a subacute intraparenchymal hematoma may show ring enhancement.

Diffuse axonal injury (DAI)/Traumatic axonal injury (TAI)

• Diffuse axonal injury (DAI) is the result of a shear-strain deformation of the brain.

• The term traumatic axonal injury (TAI) has recently been introduced as this injury pattern is thought to be multifocal rather than diffuse; however, this text will use the term DAI, as that is the more common term.

• DAI is caused by rotational deceleration and subsequent reacceleration force that exceeds the limited elastic capacity of the axons.

The most common locations of DAI include the gray–white matter junction, the corpus callosum, and the dorsolateral midbrain.

The higher the grade, the worse the prognosis.

Grade I DAI involves only the gray–white matter junctions.

Grade II DAI involves the corpus callosum.

Grade III (most severe) DAI involves the dorsolateral midbrain.

• CT is relatively insensitive for detection of DAI, although hemorrhagic DAI may show tiny foci of high attenuation in the affected regions.

• MRI is much more sensitive to detect DAI, although detection relies on multiple
sequences, including FLAIR, GRE, and DWI.

GRE is extremely sensitive for hemorrhagic axonal injury; however, not all DAI is hemorrhagic.
FLAIR is most sensitive for nonhemorrhagic DAI.

Diffusion sequences show restricted diffusion in acute DAI due to cytotoxic edema and cell swelling.

Facial fractures

Zygomaticomaxillary complex fractures

• Commonly but incorrectly known as the tripod fracture, a zygomaticomaxillary complex (ZMC) fracture causes a floating zygoma by disrupting all four of the zygomatic articulations.

• The zygoma normally articulates with the frontal, maxillary, temporal, and sphenoid bones via the zygomaticofrontal, zygomaticomaxillary, zygomaticotemporal, and zygomaticosphenoid articulations.

• A ZMC fracture causes disruption of the zygomatic articulations by fractures through the following structures:
Lateral orbital rim fracture: Zygomaticofrontal disruption.
Inferior orbital rim fracture: Zygomaticomaxillary disruption.
Zygomatic arch fracture: Zygomaticotemporal disruption.
Lateral orbital wall: Zygomaticosphenoid disruption.

• The Le Fort classification describes a predictable pattern of midface fractures, all of which disrupt the pterygomaxillary buttress and cause detachment of the maxilla from the skull base.

All Le Fort fractures are defined by fractures through the pterygoid plates.
• Le Fort I (floating palate) detaches the maxillary alveolus from the skull base.
• Le Fort II dissociates the central midface from the skull, causing the nose and hard palate to be moved as a single unit.
• Le Fort III represents a complete midface dissociation.

Cortical anatomy

Central sulcus

Find the central sulcus:

• The central sulcus separates the motor strip (frontal lobe) from the sensory cortex (parietal lobe).

• To find the central sulcus, follow the cingulate sulcus posteriorly on a slightly offmidline sagittal (left images above).

The cingulate sulcus connects to the marginal ramus.

Directly anterior to the marginal ramus is the paracentral lobule, which contains both the motor strip and the sensory cortex.

• On an axial image (right images above), the central sulcus forms a characteristic upside down omega (Ω).

The corresponding region of motor strip, just anterior to the Ω, controls the hand.

Vascular anatomy

Internal carotid artery (ICA)

Segments of the internal carotid artery

• Cervical (C1): Does not branch within the neck.

• Petrous (C2): Fixed to bone as the ICA enters the skull base, so a cervical carotid dissection is unlikely to extend intracranially.

• Lacerum (C3): No branches.

• Cavernous (C4): The meningohypophyseal trunk arises from the cavernous carotitid to supply the pituitary, tentorium, and dura of the clivus.

The inferolateral trunk also arises from C4 to supply the 3rd, 4th, and 6th cranial nerves, as well as the trigeminal ganglion.

• Clinoid segment (C5): The carotid rings are two dural rings that mark the proximal and distal portions of the clinoid segment of the ICA.

The carotid rings prevent an inferiorly located aneurysm from causing intracranial subarachnoid hemorrhage with rupture.

• Supraclinoid (C6–C7): Gives off several key arteries:

The ophthalmic artery supplies the optic nerve. It takes off just distal to the distal carotid ring in 90% of cases and can be used as a landmark for the distal ring.

Aneurysms located superior to this ring can result in subarachnoid hemorrhage.

Given this risk, these aneurysms are treated more aggressively than aneurysms located proximal to the distal dural ring, which are contained.

The posterior communicating artery (P-comm) is an anastomosis to the posterior circulation.

A fetal posterior cerebral artery (PCA) is a variant supplied entirely by the ipsilateral ICA via an enlarged P-comm.

The anterior choroidal artery supplies several critical structures, despite its small size.

It supplies the optic chiasm, hippocampus, and posterior limb of the internal capsule.

Circle of Willis

Critical small arteries arising from the circle of Willis

• The A1 segment of the anterior cerebral artery (ACA) travels above the optic nerves and give off the recurrent artery of

Heubner, which supplies the caudate head and anterior limb of the internal capsule.

The A1 segment also gives rise to the medial lenticulostriate perforator vessels, which supply the medial basal ganglia.

• Just outside the circle of Willis, the middle cerebral artery (MCA) gives rise to the lateral lenticulostriate perforator vessels to supply the lateral basal ganglia including the lateral putamen, external capsule, and the posterior limb of the internal capsule.

• The posterior communicating artery (P-comm) travels between the optic tract and the 3rd cranial nerve, giving off anterior thalamoperforator vessels.

A P-comm aneurysm
may cause cranial nerve III palsy due to local mass effect.

• The posterior cerebral artery (PCA) gives off thalamoperforators to supply the thalamus.

Artery of Percheron is a variant where there is a dominant thalamic perforator supplying the ventromedial thalami bilaterally and the rostral midbrain, arising from a P1 PCA segment.
]
An artery of Percheron infarct will result in bilateral ventromedial thalamic infarction, with or without midbrain infarction (the infarct may be V shaped if the midbrain is involved).

Deep venous thrombosis may also result in bilateral thalamic infarcts.

• The anterior choroidal artery is the most distal branch of the internal carotid artery.
As discussed on the previous page, it supplies the optic chiasm, hippocampus, and posterior limb of the internal capsule.

Circle of Willis normal anatomic variants

• Normal circle of Willis anatomy is only seen approximately 25% of the time.

Middle cerebral artery (MCA)

Segmental anatomy of the middle cerebral artery

• Although the transition from M1 to M2 is technically defined as the upward point of deflection into the sylvian fissure, in practical terms, the pre-bifurcation MCA is often called M1 and the post-bifurcation MCA is called M2.

Anterior cerebral artery (ACA)

Segmental anatomy of the anterior cerebral artery

• The recurrent artery of Heubner arises most commonly from the A1 segment of the ACA, proximal to the anterior communicating artery.

The recurrent artery of Heubner supplies the head of the caudate and the anterior limb of the internal capsule.

Persistent carotid–basilar connections

Overview of persistent fetal anterior–posterior connections

• A number of carotid to basilar connections are formed during embryogenesis.

These fetal anterior–posterior circulation connections normally regress before birth.

• Occasionally, a fetal carotid–basilar connection may persist after birth.

Each anomalous connection is named for the structures adjacent to its course in the head and neck.

Persistent trigeminal artery

• A persistent trigeminal artery is the most common persistent carotid–basilar connection and has an association with aneurysms.

• The persistent trigeminal artery courses adjacent to the trigeminal nerve.

Angiography shows a characteristic trident or tau sign on the lateral view due to the artery's branching structure.

• Saltzman type I connects to the basilar artery while Saltzman type II connects to the superior cerebellar artery.

Less common carotid to basilar connections

• The otic, hypoglossal, and proatlantal intersegmental arteries are rare persistent carotid–basilar connections.

Stroke

Imaging of stroke

Determine potential candidate for therapy

• The goal of stroke imaging is to determine who would benefit from therapy.

• The goal of stroke therapy is to restore perfusion to the brain.

• In the appropriate patients, intravenous or intra-arterial thrombolysis performed with tissue plasminogen activator (tPA) can have near-miraculous results.

However, there is grave risk of fatal hemorrhage if patients are inappropriately selected for therapy.

The exact exclusion criteria for administration of thrombolytic therapy varies among institutions.
• The American Heart Association (AHA) published guidelines for early management of adults with ischemic stroke (Stroke, 2007) and established recommendations for imaging and treatment of acute stroke.

Imaging of the brain is recommended before thrombolytic therapy is administered and the imaging study should be interpreted by a physician with expertise in reading brain studies.

Initial imaging in suspected acute stroke is usually noncontrast CT, for the primary purpose of excluding hemorrhage.

However, some authors assert that MRI is equally sensitive for detecting hemorrhage using GRE sequences.

Advanced imaging (with CT or MR) includes vascular imaging, diffuse-weighted imaging, and perfusion imaging.

These advanced imaging studies may provide additional information, but are not required before the initiation of thrombolytic therapy.

In fact, advanced imaging should not delay treatment.

Per AHA guidelines, the only CT finding that absolutely precludes intravenous tPA within 3 hours of onset of stroke is the presence of hemorrhage.

Some authors advocate for extending the window for tPA administration to 4.5 hours from stroke onset, although this expanded window is not discussed in the AHA guidelines.

Intra-arterial thrombolysis may be performed for an MCA thrombus within 6 hours of stroke onset in patients who are not candidates for intravenous thrombolysis.

Subsequent to the development of the AHA guidelines, some authors recommend extending the window for intra-arterial treatment of anterior circulation stroke to 8 hours. Similarly, many authors argue for no time limit for intraarterial tPA for posterior circulation infarction because these strokes can be catastrophic if untreated.

• Some institutions add additional exclusion criteria for administration of intravenous
tPA, although these additional criteria are not a part of the AHA guidelines:
Individuals with a large (greater than 1/3 MCA territory) infarct may be excluded from IV tPA.
Occlusion of the distal internal carotid artery and proximal MCA and ACA (a T-shaped occlusion) may preclude treatment with IV tPA.

Absence of a penumbra (discussed below) of salvageable brain that represents at least 20% of the region of abnormal perfusion may preclude treatment with IV tPA.

Perfusion stroke imaging

• The role of perfusion CT or MRI in the management of acute stroke is evolving and remains controversial.

The theoretical goal of perfusion imaging is to characterize the ischemic penumbra, which is the area of vulnerable brain adjacent to the infarct core that may also become infarcted without intervention.

Currently, no clinical guidelines exist regarding the implementation of perfusion imaging.

The penumbra does receive some perfusion, but at a reduced rate compared to normal brain.

Perfusion of the penumbra is <20 mL/100 g tissue per minute in physiologic studies, compared to ~60 mL/100 g tissue per minute for normal gray matter.

Such a low rate of perfusion causes cellular dysfunction and produces a neurologic deficit, which may be restored with therapy.

The infarct core is usually dead tissue, which generally cannot recover even after therapy.

Acute stroke: Noncontrast CT imaging

• Noncontrast CT is the initial test of choice for evaluation of hyperacute infarct when the patient presents within the IV tPA time window (3 hours, or 4.5 hours at some institutions).

• The main purpose of a noncontrast CT is to exclude patients who would be harmed by thrombolytic therapy as discussed above, most importantly to exclude those with hemorrhage.

• Noncontrast CT in the hyperacute stage is relatively insensitive to detect early infarction compared to MRI.

Subtle loss of gray–white differentiation in the insula or basal ganglia may be present on CT, thought to be due to decreased cerebral blood volume.

• The insular ribbon sign describes the loss of gray–white differentiation in the insula.

The gray–white junction becomes most conspicuous at very narrow stroke windows (window 30/level 30).

• Obscuration of the lentiform nucleus (putamen and globus pallidus) is caused by loss of gray–white differentiation at the border of the lentiform nucleus and the posterior limb of the internal capsule.

• The hyperdense artery sign describes direct visualization of the acute intravascular thrombus, most commonly seen in the MCA.

The hyperdense artery sign is specific for ischemia when seen, but relatively insensitive (seen in approximately one third of cases).

Some authors suggest that the presence of the hyperdense artery sign portends a worse prognosis.

Acute stroke: MR imaging

• Detailed MRI imaging of the multiple temporal stages of stroke is discussed on the following pages.

For the initial evaluation, diffusion sequences can detect acute infarction with high sensitivity within minutes of symptom onset.

DWI is more sensitive than FLAIR in the detection of hyperacute stroke.

Evolution of infarction

Hyperacute infarct (0–6 hours)

• Within minutes of critical ischemia, the sodium–potassium ATPase pump that maintains the normal low intracellular sodium concentration fails.

Sodium and water diffuse into cells, leading to cell swelling and cytotoxic edema.

• Calcium also diffuses into cells, which triggers cascades that contribute to cell lysis.

• By far the most sensitive imaging modality for detection of hyperacute infarct is MRI diffusion-weighted imaging (DWI).

DWI hyperintensity and ADC map hypointensity reflect reduced diffusivity, which can be seen within minutes of the ictus.

• Diffusion is reduced in an acute infarct by two factors:
1) Shift from extracellular to intracellular water due to Na/K ATPase pump failure.

2) Increased viscosity of infarcted brain due to cell lysis and increased extracellular protein.

• FLAIR may be normal. Subtle hyperintensity may be seen on FLAIR images in the hyperacute stage.

These changes are seen less than two thirds of the time within the first six hours.

• Perfusion shows decreased cerebral blood volume of the infarct core, with or without a surrounding region of decreased cerebral blood flow, which represents the penumbra.

Acute infarct (6 hours–72 hours)

• The acute infarct phase is characterized by increase in vasogenic edema and mass effect.

• Damaged vascular endothelial cells cause leakage of extracellular fluid and increase the risk of hemorrhage.

• **On imaging**, there is increased sulcal effacement and mass effect.

The mass effect peaks at 3–4 days, which is an overlap time between the acute and early subacute phases.

• MRI shows hyperintensity of the infarct core on T2-weighted images, best seen on FLAIR.

The FLAIR abnormality is usually confined to the gray matter.

DWI continues to show restricted diffusion.

• There may be some arterial enhancement, due to increased collateral flow.

• Perfusion images most commonly show increase in size of the infarct core with resultant decrease in size of the penumbra.

Early subacute infarct (1.5 days–5 days)

• In the early subacute phase, blood flow to the affected brain is re-established by leptomeningeal collaterals and ingrowth of new vessels into the region of infarction.
• The new vessels have an incomplete blood brain barrier, causing a continued increase in vasogenic edema and mass effect, which peaks at 3–4 days.

• MR imaging shows marked hyperintensity on T2-weighted images involving both gray and white matter (in contrast to the acute stage, which usually involves just the gray matter).

• The ADC map becomes less dark or even resolves if there is extensive edema; however, the DWI images typically remain bright due to underlying T2 shine through.

• Perfusion imaging shows continued expansion of the infarct core and further reduction in the ischemic penumbra.

Late subacute infarct (5 days–2 weeks)

• The subacute phase is characterized by resolution of vasogenic edema and reduction in mass effect.

• A key imaging finding is gyriform enhancement, which may occasionally be confused for a neoplasm.

Unlike a tumor, however, a subacute infarction will not typically demonstrate both mass effect and enhancement simultaneously.

Enhancement can be seen from approximately 6 days to 6 weeks after the initial infarct.

The enhancement of a subacute infarct has also been described by the “2-2-2” rule, which states that enhancement begins at 2 days, peaks at 2 weeks, and disappears by 2 months.

• DWI may remain bright due to T2 shine through, although the ADC map will either return to normal or show increased diffusivity.

Chronic infarct

• In the chronic stage of infarction, cellular debris and dead brain tissue are removed by macrophages and replaced by cystic encephalomalacia and gliosis.

• Infarct involvement of the corticospinal tract may cause mass effect, mild hyperintensity on T2-weighted images, and eventual atrophy of the ipsilateral cerebral peduncle and ventral pons due to Wallerian degeneration.

These changes can first be seen in the subacute phase, with atrophy being the predominant feature in the chronic stage.

• DWI has usually returned to normal in the chronic stage.

• Occasionally, cortical laminar necrosis can develop instead of encephalomalacia.

Cortical laminar necrosis is a histologic finding characterized by deposition of lipidladen macrophages after ischemia that manifests on imaging as hyperintensity on both T1- and T2-weighted images.

Vascular malformations

High-flow lesions (lesions with shunting)

Arteriovenous malformation (AVM)

• An arteriovenous malformation (AVM) is a congenital high-flow vascular malformation consisting of directly connecting arteries and veins without an intervening capillary bed.
AVM occurs intra-axially and 85% are supratentorial.

AVM usually presents with seizures or bleeding (usually parenchymal hemorrhage, rarely subarachnoid).

Aneurysms of the feeding arteries or intra-nidal arteries are often seen, which predispose to bleeding.

• The Spetzler–Martin scale helps to evaluate surgical risk for AVM resection.

A large
AVM draining to a deep vein in eloquent cortex is high risk, while a small AVM draining to a superficial vein in non eloquent cortex is low risk.

• On imaging, AVM is characterized by a vascular nidus (“nest”) containing numerous serpentine vessels that appear as black flow-voids on MRI.

There are usually adjacent changes to the adjacent brain including gliosis (T2 prolongation), dystrophic calcification, and blood products (blooming on T2* gradient imaging).

The gliosis/encephalomalacia or mineralization seen in the adjacent brain is due to alteration in vascular flow from the AVM.

• AVM replaces rather than displaces brain. It causes minimal mass effect.

• Uncommonly, a bleeding AVM may be angiographically occult if the malformed vessels are compressed by the acute hematoma.

• Factors that increase bleeding risk that are detectable by imaging include intra-nidal aneurysm, venous ectasia, venous stenosis, deep venous drainage, and posterior fossa location.

• Treatment can be with embolization, stereotactic radiation, or surgical resection.

• Vein of Galen malformation is a type of vascular malformation characterized by arteriovenous fistulae from thalamoperforator branches into the deep venous system.

The enlarged vein is actually an enlarged median prosencephalic vein.

In childhood, a vein of Galen malformation is the most common extracardiac cause of high output cardiac failure.

Vein of Galen malformation may also be seen in adults, but clinically would be either asymptomatic or may be the cause of Parinaud syndrome due to mass effect in the pineal region.

Dural arteriovenous fistula (dAVF)

• Dural arteriovenous fistulas are a complex group of high-flow lesions characterized by arteriovenous shunts between the meningeal arterioles and dural venules.

• The primary prognostic feature is the presence and degree of cortical venous drainage.

The Cognard classification I through IV describes lesions with progressively increased risk of bleeding.

Type V is reserved for spinal dAVFs.

Type I: No cortical venous drainage.
Lowest risk of bleeding.
Type IIA: Reflux into dural sinus but not cortical veins.
Type IIB: Reflux into cortical veins:
10–20% hemorrhage rate.
Type III: Direct cortical venous drainage:
40% hemorrhage rate.

Type IV: Direct cortical venous drainage with venous ectasia: 66% hemorrhage rate.
Type V: Spinal venous drainage.

May cause myelopathy.

• Carotid–cavernous fistula (CCF) is a subtype of dAVF that is often caused by trauma with resultant fistula between the cavernous carotid artery and the cavernous sinus.

Enlargement of the superior orbital vein and shunting within the cavernous sinus can lead to eye symptoms, such as proptosis and cranial nerve palsy.

Low-flow lesions (lesions without shunting)

Cavernous malformation (cavernoma)

Cavernous malformation and associated DVA: Two axial contrast-enhanced T1-weighted images show a hypointense, centrally hyperintense nonenhancing cavernous malformation (yellow arrow) in the left cerebellar hemisphere.

Directly superior to the cavernoma (right image) is an enhancing vascular structure with caput medusa morphology (red arrow), representing a developmental venous anomaly.

• A cavernous malformation (also called a cavernoma) is a vascular hamartoma with a very small but definite bleeding risk.

The clinical course of a cavernous malformation is variable and the lesion may cause seizures even in the absence of significant hemorrhage.

• Cavernous malformation is often associated with an adjacent developmental venous anomaly (DVA).

There is increased risk of bleeding if a DVA is present.

However, the DVA itself does not have any bleeding risk.

• When multiple, cavernous malformations represent an inherited disorder called familial cavernomatosis.

• Cavernous malformations can be induced by radiation treatment to the brain.

• Noncontrast CT shows a well-circumscribed rounded hyperattenuating lesion.

The hyperattenuation is due to microcalcification within the cavernoma.

• MRI shows characteristic “popcorn-like” appearance of lobular mixed signal on T1-and T2-weighted images from blood products of various ages.

There is a peripheral rim of hemosiderin which is dark on GRE and T2-weighted image.

There is typically no enhancement, but intense enhancement may be seen with a long delay after contrast administration.

Cavernous malformations may range in size from tiny (single focus of susceptibility artifact) to giant.

• Cavernous malformations are usually occult by vascular imaging (CTA or angiography).

Developmental venous anomaly (DVA), also called venous angioma

• A developmental venous anomaly (DVA) is an abnormal vein that provides functional venous drainage to normal brain.

• DVA can usually only be seen on contrast-enhanced images, where it appears as a radially oriented vein with a characteristic caput medusa appearance.

• A DVA is a Do Not Touch lesion. If resected, the patient will suffer a debilitating venous infarct.

The DVA must be preserved if an adjacent cavernous malformation is resected.

Capillary telangiectasia

• A capillary telangiectasia is an asymptomatic vascular lesion composed of dilated capillaries with interspersed normal brain.

A capillary telangiectasia is another Do Not Touch lesion.

• Post-contrast MRI shows a faint, brush-stroke-like enhancing lesion in the brainstem or pons, without mass effect or surrounding edema.

GRE may show blooming due to susceptibility.

• Similar to cavernous malformation, capillary telangiectasia is angiographically occult.

Subarachnoid hemorrhage and aneurysms

Subarachnoid hemorrhage (SAH)

• Overall, the most common cause of subarachnoid hemorrhage (SAH) is trauma.

]Aneurysm rupture is by far the most common cause of non-traumatic subarachnoid hemorrhage.

No cause of the subarachnoid hemorrhage is identified in up to 22% of cases.

• Clinically, non-traumatic subarachnoid hemorrhage presents with thunderclap headache and meningismus.

• Noncontrast CT is the initial imaging modality in suspected subarachnoid hemorrhage.

On CT, subarachnoid blood appears as high attenuation within the subarachnoid space.

High attenuation material in the subarachnoid space may be due to SAH (by far the most common cause), meningitis, leptomeningeal carcinomatosis, or prior intrathecal contrast administration.

• Noncontrast CT is >95% sensitive for detecting subarachnoid hemorrhage within the first six hours, with sensitivity slowly decreasing to 50% by day 5.

If clinical suspicion for subarachnoid hemorrhage is high with a negative CT scan, the standard of care is to perform a lumbar puncture to look for xanthochromia.

• If SAH is present on imaging or lumbar puncture shows xanthochromia, catheter angiography is the gold standard to evaluate for the presence of an aneurysm.

Several recent studies have shown, however, that CT angiography is equivalent to catheter angiography in the search for a culprit aneurysm in cases of SAH.

• **On MRI**, acute subarachnoid hemorrhage appears hyperintense on FLAIR and demonstrates susceptibility artifact on gradient sequences.

The differential diagnosis for increased FLAIR signal in the subarachnoid space is similar to the differential for high attenuation subarachnoid material seen on CT, including SAH, meningitis, leptomeningeal carcinomatosis, and residual contrast material.

Note that meningitis and carcinomatosis will typically show leptomeningeal enhancement in addition to the abnormal FLAIR signal.

Recent oxygen or propofol administration will also cause increased subarachnoid FLAIR signal.

Distribution of subarachnoid hemorrhage

• The pattern of subarachnoid hemorrhage may provide a clue to the location of the ruptured aneurysm.

However, multiple aneurysms are seen in up to 20% of patients with SAH, and subarachnoid blood may redistribute if the patient was found down.

• Hemorrhage in the anterior interhemispheric fissure suggests an anterior communicating artery aneurysm (33% of intracranial aneurysms).

• Hemorrhage in the suprasellar cistern suggests a posterior communicating artery aneurysm (also 33% of intracranial aneurysms).

Rarely, P-comm aneurysm rupture can result in isolated subdural hemorrhage.

• Hemorrhage in the sylvian fissure suggests a middle cerebral artery aneurysm (20% of intracranial aneurysms).

• Hemorrhage in the perimesencephalic cistern suggests either a basilar tip aneurysm (5% of intracranial aneurysms), which has a high morbidity, or the relatively benign nonaneurysmal perimesencephalic subarachnoid hemorrhage (subsequently discussed).

Grading of subarachnoid hemorrhage

• The Hunt and Hess score is the clinical grading scale for aneurysmal subarachnoid hemorrhage and is based solely on symptoms, without imaging. Grade I is the lowest grade, with only a mild headache.

Grade V is the most severe, with coma or extensor posturing.
• **The Fisher grade** classifies the CT appearance of SAH. Grade 1 is negative on CT;
grades 2 and 3 are <1 mm thick and >1 mm thick, respectively, and
grade 4 is diffuse SAH or intraventricular or parenchymal extension.

Complications of subarachnoid hemorrhage

• Vasospasm is the most common cause of morbidity and mortality in patients who survive the initial episode of subarachnoid hemorrhage.

The peak incidence of vasospasm occurs approximately 7 days after the initial ictus.

Vasospasm may lead to stroke or hemorrhage.

• Approximately 20–30% of patients with subarachnoid hemorrhage will develop acute hydrocephalus, due to obstruction of arachnoid granulations.

Treatment is ventriculostomy.

• **Superficial siderosis** is a condition caused by iron overload of pial membranes due to chronic or repeated subarachnoid bleeding.

Clinically, patients with superficial siderosis present with sensorineural deafness and ataxia.

On imaging, the iron causes hypointensity on T2-weighted images outlining the affected sulci.

• Imaging workup includes cranial and spinal imaging to evaluate for a source of bleeding.

Perimesencephalic subarachnoid hemorrhage

• Perimesencephalic subarachnoid hemorrhage is a type of nonaneurysmal subarachnoid hemorrhage that is a diagnosis of exclusion with a much better prognosis than hemorrhage due to a ruptured aneurysm.

• The hemorrhage must be limited to the cisterns directly anterior to the midbrain.

The standard of care is to perform catheter angiography twice, one week apart.

Both angiograms must be negative.
Although the cause of the hemorrhage is unknown, it is thought to represent angiographically occult venous bleeding.

• Although the clinical presentation of perimesencephalic subarachnoid hemorrhage is similar to aneurysmal SAH (thunderclap headache), patients generally do well without residual neurological deficit.

Some patients may experience mild to moderate vasospasm.

Reversible cerebral vasoconstriction syndrome (RCVS)

• Reversible cerebral vasoconstriction syndrome (RCVS) is a cause of nontraumatic, nonaneurysmal subarachnoid hemorrhage and ischemia.

RCVS presents with thunderclap headache and is characterized by prolonged (but reversible) vasoconstriction.

Aneurysm morphology and pathology

Saccular aneurysm

• A saccular (also called berry) aneurysm is a focal outpouching of the arterial wall, most commonly arising at a branch point in the circle of Willis.

The aneurysm points in the direction of blood flow leading into the branch point.

Saccular aneurysms are seen almost exclusively in adults, with a slight female predominance.

Saccular aneurysms are caused by a combination of hemodynamic stress and acquired degeneration of the vessel wall.

• Non-inherited risk factors for the development of saccular aneurysm include hypertension and inflammatory vascular disease such as Takayasu or giant cell arteritis.

• Inherited diseases that predispose to aneurysm formation include connective tissue diseases such as Marfan and Ehlers–Danlos, polycystic kidney disease, and neurofibromatosis type I.

• The aneurysm neck is the opening that connects the aneurysm to the parent vessel and the aneurysm body is the aneurysm sac.

The neck:body ratio affects treatment options.

Aneurysms with relatively small necks are generally easier to treat endovascularly with coils.

• Saccular aneurysms can be classified as small (<1 cm), medium (>1 cm and <2.5 cm), and giant (>2.5 cm).

The larger the size, the greater the risk for rupture.

Giant aneurysms often present with mass effect, causing cranial nerve palsy.

Fusiform aneurysm

• A fusiform aneurysm is segmental arterial dilation without a defined neck.

Fusiform aneurysms are usually due to atherosclerosis, but may arise from chronic dissection.

• In contrast to saccular aneurysms, fusiform aneurysms do not occur at branch points.

• The vertebrobasilar system is affected more commonly than the anterior circulation.

• Fusiform aneurysms are much less common than saccular aneurysms.

Since there is no aneurysm neck to exclude from the systemic circulation, fusiform aneurysms are more difficult to treat.

Fusiform aneurysms of the vertebrobasilar system pose particular challenges, as critical perforating vessels may arise directly from the diseased artery.

Mycotic (infectious) aneurysm

• Mycotic aneurysms account for only 2–4% of all intracranial aneurysms and are due to septic emboli.

Bacterial endocarditis is the most common embolic source.

• In contrast to saccular aneurysms, mycotic aneurysms form in the distal arterial circulation, beyond the circle of Willis.

Mycotic aneurysms are fragile and have a high risk of rupture.

Oncotic aneurysm

• An oncotic aneurysm is an aneurysm caused by neoplasm.

• A benign left atrial myxoma may peripherally embolize and cause a distal oncotic aneurysm.

Traumatic pseudoaneurysm

• Aneurysms due to trauma are most commonly pseudoaneurysms, which don’t contain the typical three

histologic layers of the vessel wall. Usually the vessel will exhibit abnormal luminal narrowing proximal to the aneurysm. Similar to mycotic aneurysms, traumatic pseudoaneurysms tend to occur distally.

• Arteries close to bony structures (such as the basilar and vertebral artery) are prone to dissecting aneurysms.

Cerebral venous disease

Venous anatomy

Dural sinuses

• The superior sagittal sinus (and its tributaries) drains the motor and sensory strips.

• The paired transverse sinuses are usually asymmetric, with the left transverse sinus often hypoplastic.

• The sigmoid sinus connects to the jugular bulb.

• The torcular Herophili is the confluence of the superior sagittal sinus, the transverse sinus, and the straight sinus. The word torcular is from the Greek word for wine press, and Herophilus was a Greek anatomist.

Technically, the term torcular Herophili refers to the depression on the inner table of the skull produced by the confluence of

sinuses, but in general use, torcular Herophili refers to the actual confluence of sinuses.

Deep cerebral veins

• The deep cerebral veins consist of the paired internal cerebral veins, the basal vein of Rosenthal, and the vein of Galen.

• The venous angle (red dot in the diagram above) is the intersection of the septal vein and the thalamostriate veins.

The venous angle is the angiographic landmark for the foramen of Monro.

Superficial cerebral veins

• The vein of Trolard connects superficial cortical veins to the superior sagittal sinus.

• The vein of Labbé drains the temporal convexity into the transverse or sigmoid sinus.

Retraction injury to the vein of Labbé during surgery may lead to venous infarction and aphasia.

Venous disease

Venous thrombosis

Thrombosis of a cortical vein or a deep venous sinus is one of the more common causes of stroke in younger patients.

Risk factors for venous thrombosis include pregnancy, oral contraceptives, thrombophilia, malignancy, and infection.

• A clue to the diagnosis of venous thrombosis on noncontrast CT is increased density within the thrombosed sinus or cortical vein (the cord sign).

On contrast-enhanced CT, the empty delta sign signifies a filling defect in the superior sagittal sinus.

• MR venogram will show lack of flow in the thrombosed vein or dural venous sinus.

• Venous thrombosis leads to venous hypertension, which may cause infarction and parenchymal hemorrhage.

There are three characteristic patterns of venous infarction, dependent on the location of the thrombosed vein:
Superior sagittal sinus thrombosis → infarction of the parasagittal high convexity cortex.

Deep venous system thrombosis → infarction of the bilateral thalami.

Transverse sinus thrombosis → infarction of the posterior temporal lobe.

Intraparenchymal hemorrhage

Imaging of hemorrhage

CT imaging of hemorrhage

• Noncontrast CT is usually the first imaging study performed in the emergency setting for a patient with a sudden neurologic deficit, headache, seizure, or altered level of consciousness.

• CT is highly sensitive for detection of hyperacute/acute intracranial hemorrhage, which appears hyperattenuating relative to brain parenchyma and CSF.

Acute hemorrhage may be nearly isoattenuating to water in severe anemia (hemoglobin <8 mg/dL).

MR imaging of hemorrhage

• MR imaging of hemorrhage is complex. The characteristics of blood products change on T1-and T2-weighted sequences as the iron in hemoglobin evolves through physiologic stages:
Intracellular oxyhemoglobin g deoxygenation →
Intracellular deoxyhemoglobin g oxidation →
Intracellular methemoglobin g cell lysis →
Extracellular methemoglobin g chelation →
Hemosiderin and ferritin

• Each stage of this evolution adheres to a reasonably constant time course in the intraaxial space and allows the radiologist to "date" the hemorrhage based on the unique signal characteristics on T1- and T2-weighted images for each stage.

• In general, all stages of hemorrhage are isointense or slightly dark on T1-weighted images, except for the methemoglobin stages, which are bright.

• Methemoglobin is bright on both T1- and T2-weighted images, except for intracellular methemoglobin, which is dark on T2-weighted images.

• In general, non hyperacute hemorrhage is dark on T2-weighted images, with the exception of extracellular methemoglobin, which is hyperintense on T2-weighted images.

A hyperacute hematoma, containing primarily oxyhemoglobin, is slightly hyperintense on T2-weighted images but features a characteristic dark rim representing deoxygenation at the periphery of the clot.

• The inherent slight hyperintensity of oxygenated blood on T2-weighted images becomes apparent in slow flow states, as seen in venous hypertension and moyamoya disease.

Slowly flowing blood is not susceptible to the flow-void artifact and the resultant apparently "enhancing" vasculature really represents unmasking of the normal blood signal.

• The expected evolution of blood products is highly dependent on macrophage elimination of blood breakdown products.

These rules of thumb are not applicable to extra-axial blood and timing is generally not given for extra-axial blood, such as a subdural hematoma.

Treatment of hemorrhage

• In most cases, imaging is performed to evaluate for a treatable cause of hemorrhage, such as AVM or aneurysm.

The mainstay of treatment of intraparenchymal hemorrhage is supportive, including blood pressure control and normalization of any coagulopathy.

• Larger hemorrhages can be evacuated surgically if there is significant mass effect or risk of herniation.

In particular, a hemorrhage >3 cm in the posterior fossa would generally be treated surgically as there is increased risk of brainstem compression or hydrocephalus from fourth ventricular obstruction.

Specific stages of parenchymal hematoma on MR

Hyperacute hematoma (<6 hours): Intracellular oxyhemoglobin

• A few hours after red cell extravasation, a hyperacute hematoma is primarily composed of intact red cells containing oxygenated hemoglobin, which is diamagnetic.

• The center of the hematoma will be isointense on T1-weighted images and iso- to slightly hyperintense on T2-weighted images.

• The key imaging finding of a hyperacute hematoma is a peripheral rim of hypointensity on T2-weighted images due to deoxygenation of the most peripheral red cells.

This peripheral dark rim is most conspicuous on GRE sequences.

Acute hematoma (6–72 hours): Intracellular deoxyhemoglobin

• After the red cells desaturate (lose oxygen), the entire hematoma becomes hypointense on T2-weighted images and iso- to mildly hypointense on T1-weighted images.

Early subacute hematoma (3 days to 1 week): Intracellular methemoglobin

• The subacute phase is characterized by methemoglobin, which is paramagnetic and undergoes proton–electron dipole–dipole interactions (PEDDI) with water.

PEDDI shortens T1 to cause hyperintensity on T1-weighted images.

Intracellular and extracellular methemoglobin are both hyperintense on T1-weighted images.

• In the early subacute phase, blood remains hypointense on T2-weighted images due to the paramagnetic effects of methemoglobin, which remains trapped in the red cells.

Late subacute hematoma (1 week to months): Extracellular methemoglobin (after RBC lysis)

• The methemoglobin PEDDI effect persists after cell lysis, causing continued hyperintensity on T1-weighted images.

• Paramagnetic effects of methemoglobin lessens. Signal intensity on T2-weighted images increases to that of CSF, due to RBC lysis and decrease in protein concentration.

• There may be peripheral enhancement of a subacute to chronic infarct.

Chronic sequela of hemorrhage: Hemosiderin and ferritin

• Salvaged iron atoms are deposited into hemosiderin and ferritin, which become permanently trapped in the brain parenchyma after the blood brain barrier is restored.

• Susceptibility effects of the stored iron produce characteristic hypointensity on T2-weighted and GRE images.

• Chronic hemorrhage may have peripheral enhancement.

Causes of intraparenchymal hemorrhage

Hypertensive hemorrhage

• Chronic hypertension is the most common cause of spontaneous adult intraparenchymal hemorrhage and is due to the secondary microangiopathic effects of chronic hypertension.

• Chronic hypertension causes arteriolar smooth muscle hyperplasia, which eventually leads to smooth muscle death and replacement with collagen.

The resultant vascular ectasia predisposes to hemorrhage.

• Hypertensive hemorrhage occurs in characteristic locations in the basal ganglia, thalamus, and cerebellum.

• In addition to location, imaging (MRI or CT) findings suggestive of a hypertensive bleed include additional stigmata of hypertensive microangiopathy, such as periventricular white matter disease and prior lacunar infarcts.

• An additional MR-specific finding suggesting hypertensive hemorrhage is the presence of microhemorrhages on T2* (GRE or SWI) in the basal ganglia or brainstem.

Cerebral amyloid angiopathy (CAA)

• Cerebral amyloid angiopathy (CAA) is amyloid accumulated within the walls of small and medium arteries, ultimately causing vessel weakness and increased risk of hemorrhage.

• While the spontaneous form of CAA occurs almost exclusively in elderly adults (in which population it is the second most common cause of nontraumatic hemorrhage), a hereditary variant has an earlier age of onset.

• In addition to being a risk factor for hemorrhage, CAA can also occlude the lumens of small vessels and contribute to microangiopathy.

• The main clinical clue that a hemorrhage is secondary to CAA is that the patient is a normotensive elderly adult.

• The primary imaging feature to suggest CAA is the location of hematoma, which is almost always lobar or cortical, usually in the parietal or occipital lobes.

• An important secondary MRI finding is multiple microhemorrhages seen on T2* (GRE or SWI) within the brain parenchyma.

In contrast to the microhemorrhages associated with hypertension, CAA microhemorrhages are in the cortex, not in the basal ganglia.

Aneurysmal hemorrhage

• As discussed, aneurysmal hemorrhage is by far the most common cause of nontraumatic subarachnoid hemorrhage.

If an intraparenchymal hematoma is due to an aneurysm, the hematoma is usually adjacent to the ruptured aneurysm dome.

• The pattern of subarachnoid blood may help localize the aneurysm; however, if the patient was found down, then the blood will settle in the dependent portion of the brain, confounding localization.

• CT angiography is the study of choice for further evaluation of nontraumatic subarachnoid hemorrhage.

Arteriovenous malformation (AVM)

• An arteriovenous malformation is a congenital lesion consisting of abnormal high-flow arteriovenous connections without intervening normal brain.

• In case of AVM rupture, the resultant hematoma is usually parenchymal.

In contrast to amyloid angiopathy, a hematoma from a bleeding AVM tends to affect younger patients.

Dural AV fistula (dAVF)

• Dural AV fistulas are a group of high-flow vascular malformations characterized by a fistulous connection between a meningeal artery and a venous sinus or cortical vein.

Cavernous sinus (cavernous–carotid fistula) and posterior fossa dAVFs are the most common types.

• Imaging may show enlarged feeding arteries and enlarged or occluded dural sinuses, or enlarged cortical veins.

Venous thrombosis

• Thrombosis of cortical veins or deep venous sinuses leads to venous hypertension, which may cause infarction and parenchymal hemorrhage.

Hemorrhagic neoplasm

• Occasionally, the initial presentation of a brain tumor may be acute hemorrhage.

• The most common primary brain tumor to cause hemorrhage is glioblastoma.

• There are a relatively limited number of extracranial primary tumors known to cause hemorrhagic metastases, including:
Choriocarcinoma.
Melanoma.
Thyroid carcinoma.
Renal cell carcinoma.
Although breast and lung cancer rarely cause hemorrhage on a per-case basis, they are such common cancers overall that they can always be considered when a hemorrhagic neoplasm is suspected.

• Patients treated with bevacizumab (trade name Avastin®, Genentech) may be at increased risk for hemorrhagic metastasis.

• Clues to the diagnosis of an underlying tumor causing hemorrhage include morethan-expected edema surrounding a hyperacute hematoma and heterogeneous blood product signal, suggesting varying breakdown stages of hemoglobin.

• The presence of multiple enhancing masses strongly suggests metastatic disease.

• In cases where the diagnosis is unclear, a follow-up MRI should be performed once the initial hemorrhage improves.

If tumor is present, the MRI may show a delay in the expected evolution of blood products, persistent edema, and enhancement of the underlying tumor.

Cavernous malformation (cavernoma)

• A cavernous malformation is a vascular hamartoma that consists of low-flow endothelial-lined blood vessels without intervening normal brain.

• Although non-hemorrhagic cavernomas have a characteristic MRI appearance (with “popcorn-like” lobular mixed/high signal on T1- and T2-weighted images and a dark peripheral hemosiderin rim), once bleeding occurs, the resultant hematoma has nonspecific imaging features.

The presence of a developmental venous anomaly adjacent to a hematoma may suggest the diagnosis of a recently hemorrhaged cavernoma.

• Angiography plays no role in the diagnosis. Cavernous malformations are angiographically occult.

Hemorrhagic transformation of infarct

• In most cases, the clinical or imaging diagnosis of stroke is made before hemorrhagic transformation occurs; however, hemorrhage may occasionally be the presenting feature of an infarct.

• More commonly, symptomatic hemorrhage occurs post-infarct in approximately 6–12% of patients receiving thrombolytic therapy.

• Noncontrast CT can identify risk factors for hemorrhagic transformation after thrombolytic therapy, including a relatively large region of hypoattenuation and a dense artery sign.

Note that per AHA guidelines, neither of these findings is an exclusion criterion for administration of IV tPA.

Vasculitis

• Vasculitis affecting the CNS may be primary or secondary to systemic vasculitides.

• The most common presentation of vasculitis is cerebral ischemia.

Less commonly, vasculitis may present with frank hemorrhage.

• Standard MRI imaging shows of vasculitis shows multiple foci of T2 prolongation in the basal ganglia and subcortical white matter.

• Noninvasive vascular imaging (CTA or MRA) is relatively insensitive to small vessel involvement, but may show irregularity of involved large or medium vessels.

• Angiography is the most sensitive test and shows multifocal areas of stenosis and dilation.

Moyamoya

• Moyamoya is a non-atherosclerotic vasculopathy characterized by progressive stenosis of the intracranial internal carotid arteries and their proximal branches, which leads to proliferation of fragile lenticulostriate collateral vessels.

• Angiography of the enlarged basal perforating arteries gives a puff of smoke appearance.

• The ivy sign on FLAIR MRI represents tubular branching hyperintense structures within the sulci, representing cortical arterial branches that appear hyperintense due to slow collateral flow.

• Patients with moyamoya disease are susceptible to aneurysm formation, especially in the posterior circulation.

• Perfusion studies show decreased flow in the affected vascular regions.

White matter disease

White matter overview

White matter imaging overview

• The typical MRI appearance of white matter injury is T2 prolongation of the affected white matter.

Less commonly, tumefactive demyelination may be mass-like, enhance, and look very similar to a tumor.

• The key imaging finding of demyelinating disease is minimal mass effect relative to the lesion size.

• A frequent pattern of white matter disease consisting of scattered foci of T2 prolongation in the subcortical, deep, and periventricular white matter is seen very commonly, especially in older adults.

In older patients, this pattern is most likely due to chronic microvascular ischemia.

In younger patients, a similar pattern can be seen in chronic migraine headaches, as sequelae of prior infectious or inflammatory disease, and with demyelination.

Normal structures may mimic white matter disease on T2 images

• Virchow–Robin spaces are tiny perivascular spaces that follow deep penetrating vessels into the subarachnoid space.

Virchow–Robin spaces follow CSF signal on all sequences, including FLAIR.

Enlarged Virchow–Robin spaces and a J-shaped sella can be seen in the mucopolysaccharidoses.

• Ependymitis granularis represents frontal horn periventricular hyperintensity on T2-weighted images due to interstitial CSF backup.

Despite the name ("-itis"), ependymitis granularis is not associated with inflammation.

Idiopathic/autoimmune/inflammatory white matter disease

Multiple sclerosis (MS)

• Multiple sclerosis (MS) is idiopathic inflammatory destruction of CNS axons in the brain and spinal cord.

MS is likely autoimmune in etiology and may be associated with other autoimmune diseases such as Graves disease and myasthenia gravis.

• MS is the most common chronic demyelinating disease. It often leads to severe disability.

• MS is more common in middle-aged Caucasian females from northern latitudes.

• There are two main clinical presentations of multiple sclerosis:
1) Relapsing–remitting (most common): Partial or complete resolution of each acute attack.
2) Progressive: No resolution or incomplete resolution between acute attacks.

Primary progressive: Slow onset without discrete exacerbations.
Secondary progressive: Similar to relapsing–remitting but with less complete resolution between attacks, leading to progressive disability.

• Optic neuritis may represent the first sign of MS. The purpose of a brain MRI after optic neuritis is to look for other lesions, which may be clinically silent.

• Histopathologically, destruction of myelin is caused by lymphocytes attacking oligodendrocytes (which make CNS myelin).

• Although MRI imaging is highly sensitive, there are no pathognomonic imaging findings.

The McDonald criteria, last revised in 2010, describe strict imaging findings to diagnose MS.

The McDonald criteria are most useful for clinically ambiguous cases.

• In order to make the diagnosis of MS, there must be lesions separated in space (different areas of the CNS) and in time (new lesions across scans).

Suggestive imaging findings include periventricular ovoid foci of T2 prolongation that “point” towards the ventricles, called Dawson fingers.

The corpus callosum is often affected, best seen on sagittal FLAIR.

• In general, an enhancing lesion is suggestive of active demyelination, as enhancement is thought to be due to inflammatory blood brain barrier breakdown.

• Lesions that are dark on T1-weighted images are called “black holes” and are associated with more severe demyelination and axonal loss.

• Chronic MS leads to cortical atrophy, thinning of the corpus callosum, and changes in MRI spectroscopy, with decreased NAA, increased choline, increased lipids, and increased lactate.

• Tumefactive MS describes the occasionally seen ring enhancement and mass-like appearance of an active MS plaque.

In contrast to a brain tumor, the demyelinating lesion will not have any significant mass effect, and the ring of enhancement is usually incomplete.

• MS involves the spinal cord in a substantial minority of patients and the spine is routinely evaluated in patients with MS.

Spinal MS involvement is usually short-segment and unilateral.

Isolated spinal cord involvement is seen in up to 20% of cases of MS.