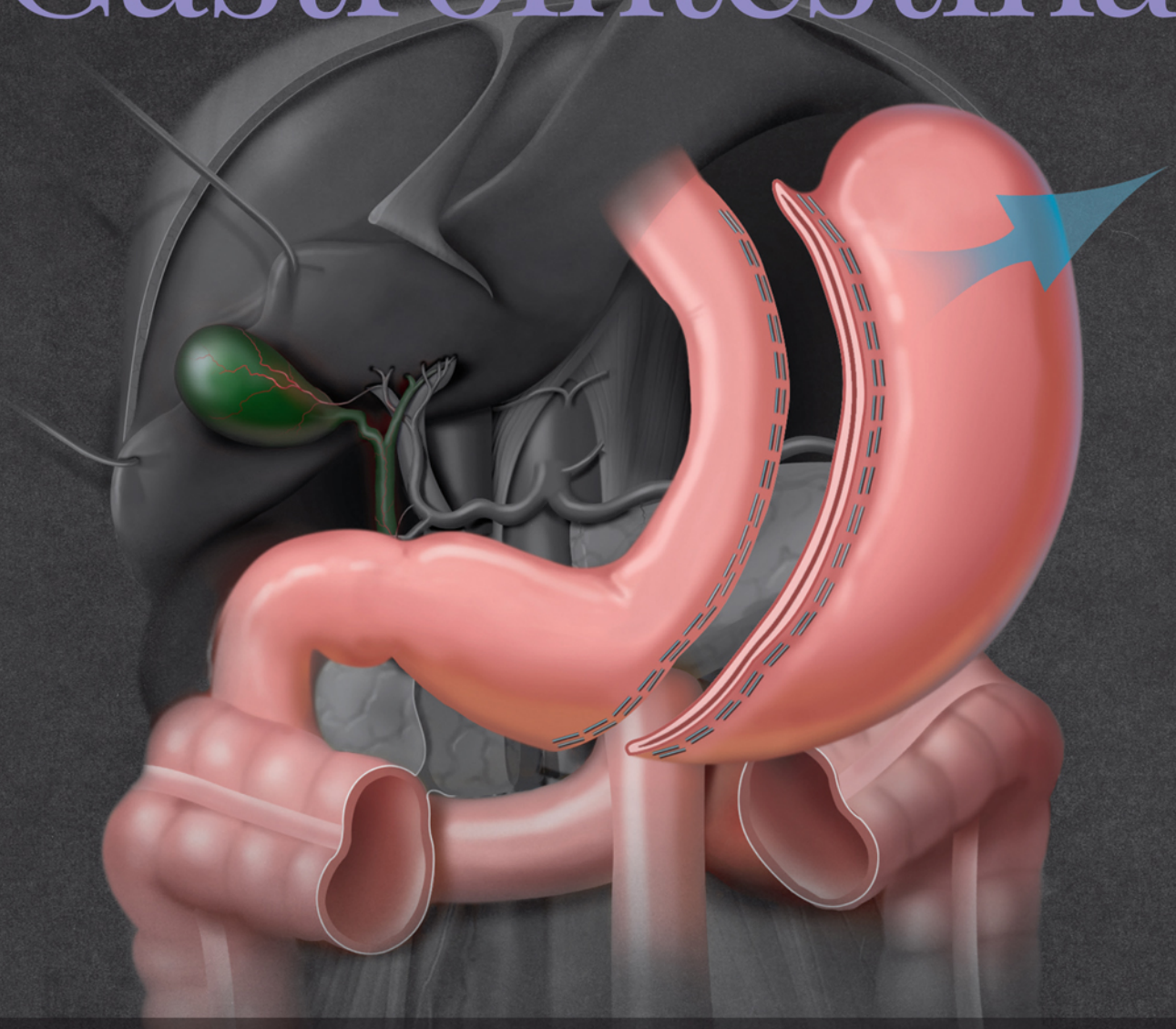
Diagnostic Imaging
Gastrointestinal



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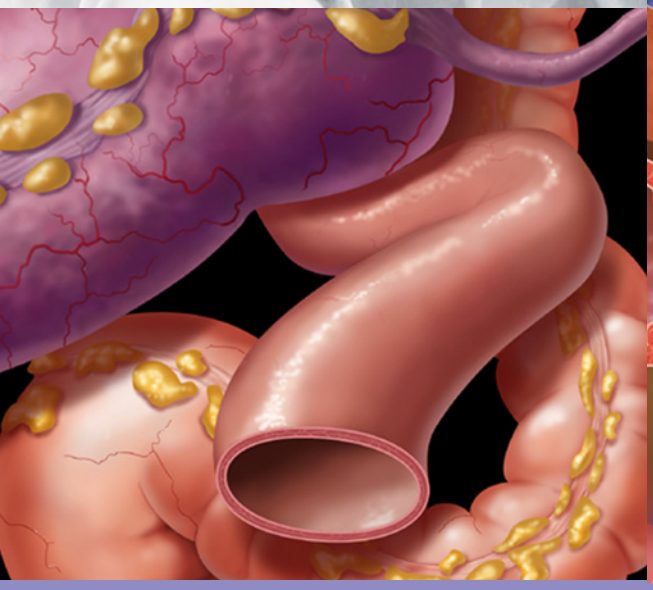

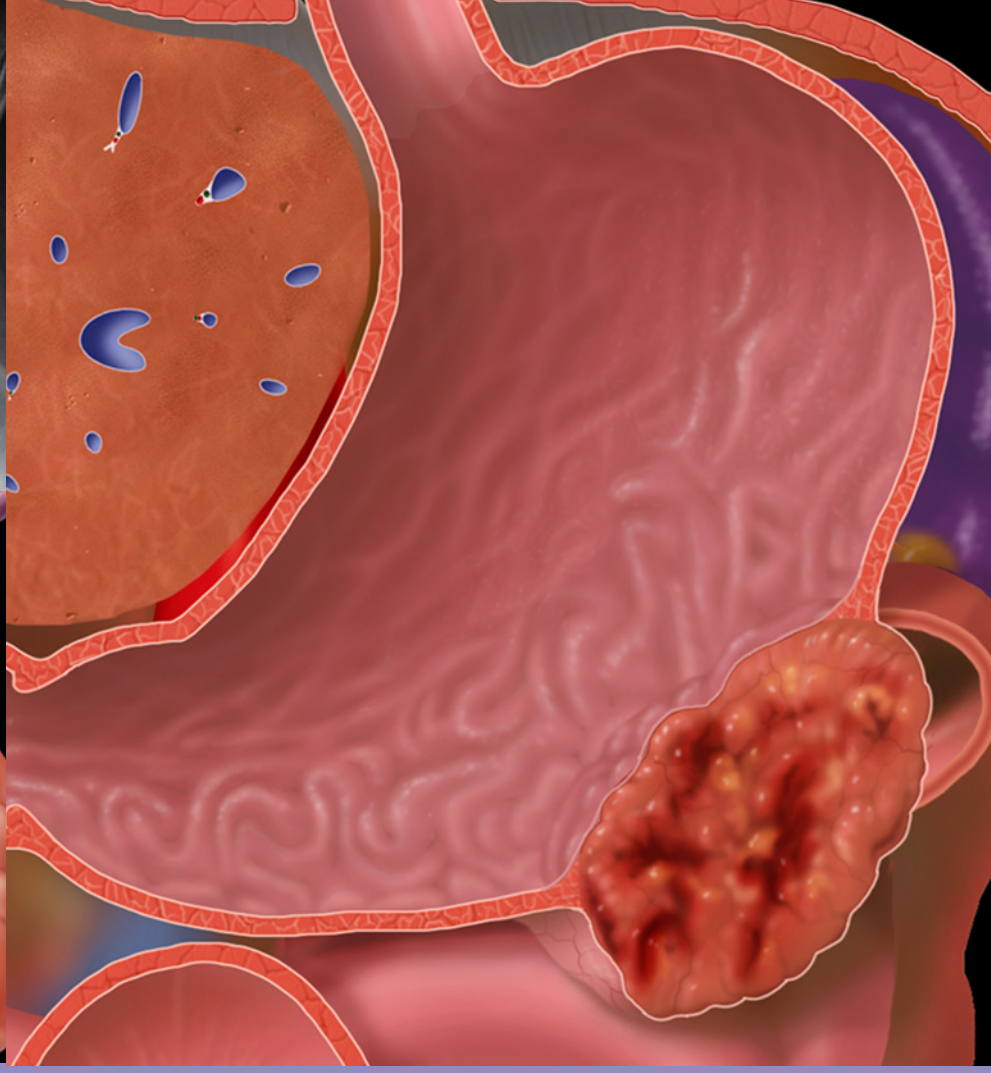

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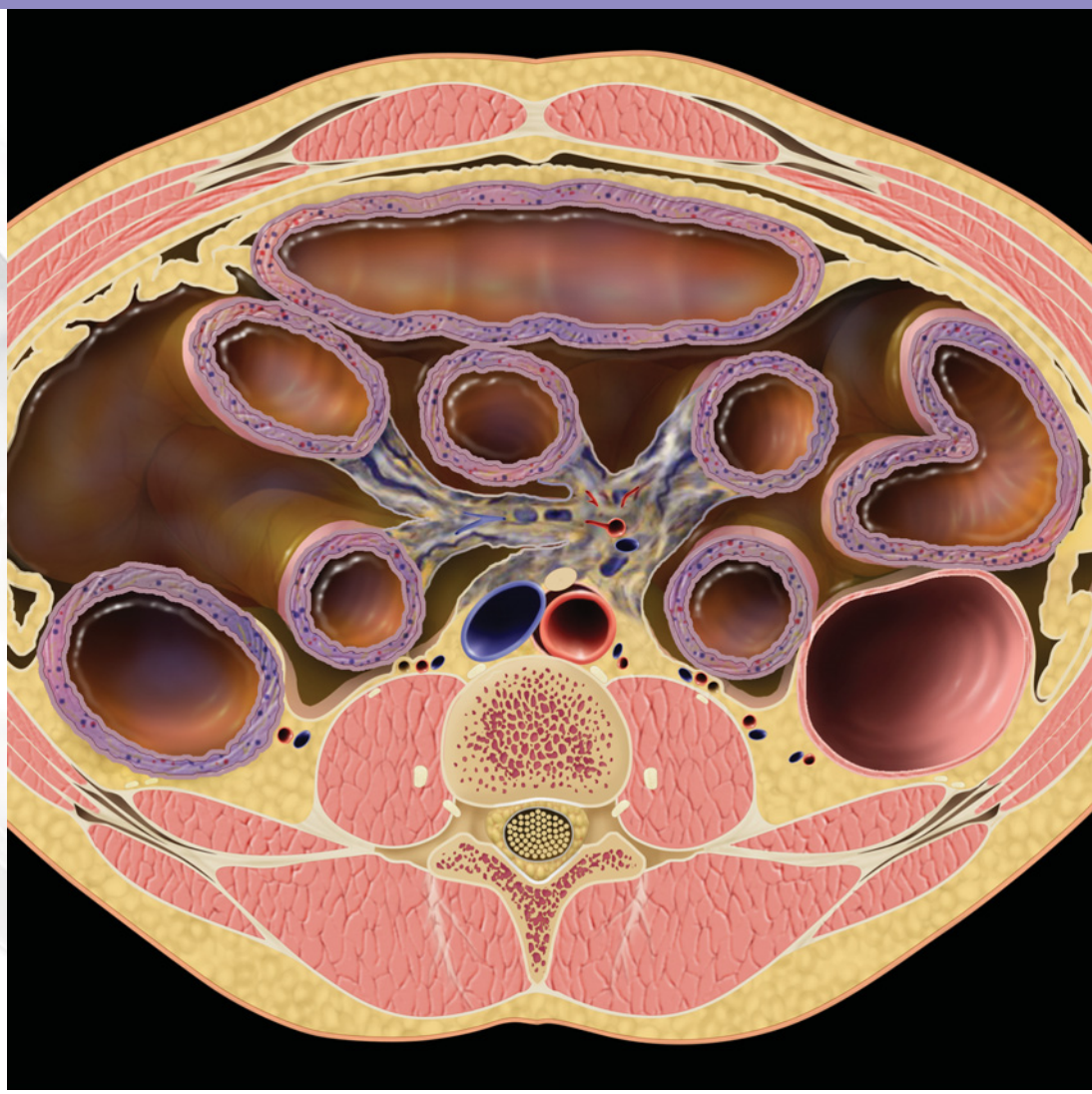

Diagnostic Imaging

Gastrointestinal

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Dedications

This book is dedicated to the phenomenal referring physicians at Stanford and Johns Hopkins Medical Centers who attract and care for some of the most challenging and interesting patients, often referred specifically to them because of their well-earned reputations for excellence. They keep us "on our toes," and the most rewarding part of our jobs is to participate with them in advancing the evaluation and care of patients with abdominal disease and disorders.

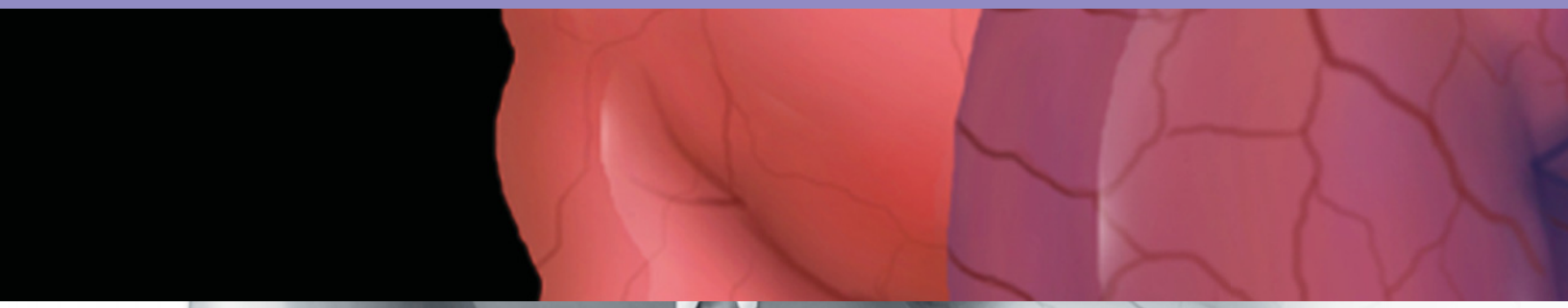
MPF

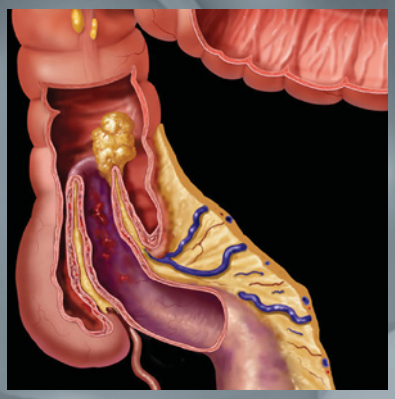
To my loving wife, Janani Venkateswaran, for her boundless understanding and patience.

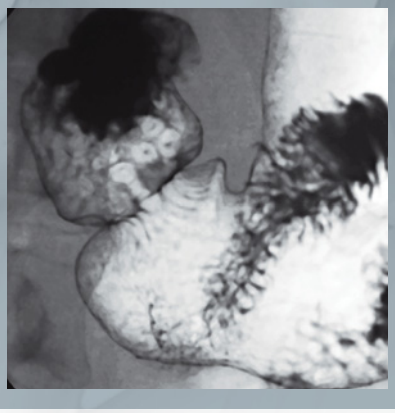
To my parents, Raghu and Visali, for their support throughout my entire career and education.

To all my colleagues at Johns Hopkins.

SPR







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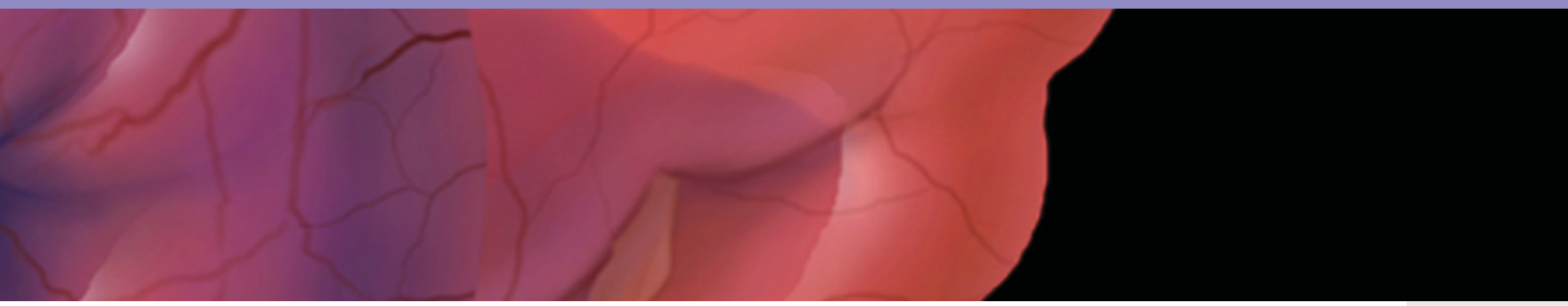

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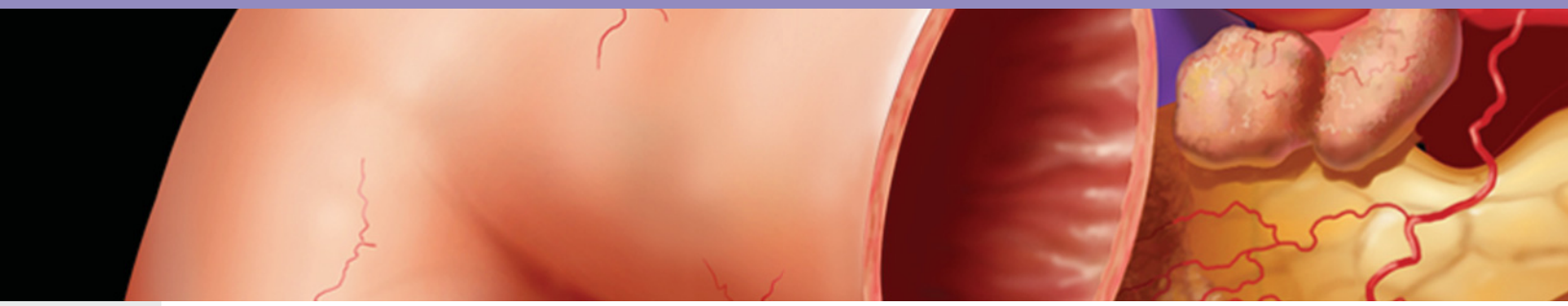
Preface



The second edition of *Diagnostic Imaging: Abdomen* was a major expansion of the first edition, containing over 150 additional diagnoses. In planning this, the third edition, we soon realized that comprehensive coverage of all of the advances in imaging and management of abdominal disorders was no longer possible in a single volume text. Therefore, we elected to separate diagnoses judged primarily "gastrointestinal," covered in this thoroughly updated text, from the "genitourinary" topics, to be covered in a subsequent book.

We have maintained the classic Amirsys style of bulleted text, allowing us to present factual material in less than half the space with greater clarity and readability. We have, however, also maintained and expanded the popular Introduction and Overview sections, which are written in a more informal prose style, to help readers grasp the essential anatomical issues, imaging protocols, and general approaches to the most common and important disorders affecting that organ system.

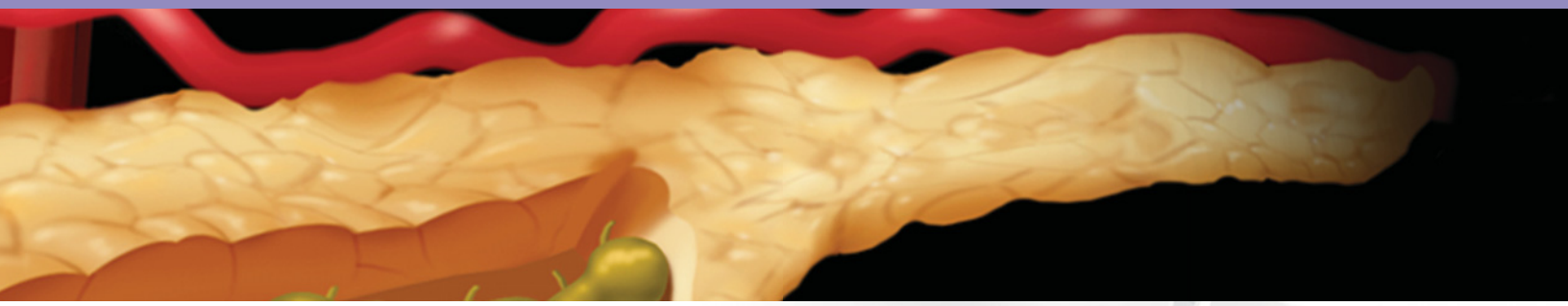
As a new feature, we have added lists of the most important differential diagnoses to each Introduction and Overview section, helping readers to zero in, for instance, on the possible etiologies for a "cystic pancreatic mass." Reference to specific chapters on the most likely candidates will then quickly lead to a more accurate and specific diagnosis.

Fluoroscopy in the modern era (read, CT, and endoscopy) has evolved to focus primarily on pre- and postoperative evaluation of patients for surgical alterations of the GI tract. Therefore, we have de-emphasized the more esoteric aspects of fluoroscopic diagnosis of diseases in favor of more expansive coverage of the radiologist's role in evaluating patients for bariatric surgery, antireflux procedures, esophageal and bowel resections, and so forth. Additional detailed diagnostic material, images, and references are included in Elsevier's Expert Consult, an eBook that accompanies the print version of *Diagnostic Imaging: Gastrointestinal, Third Edition*.

We have updated and replaced most images from the second edition, maintaining only those judged to be so classic that newer examples would not be an improvement. All references and text have been updated as well, with all material being current to within a few months of the publication date of this book.

The rapid preparation of this book was made possible in part by limiting the primary authorship to two experienced and highly motivated authors, who took responsibility for writing and illustrating all 200 plus chapters.

We hope that this new edition of *Diagnostic Imaging: Gastrointestinal* will be a welcome addition to your library, but only after you have read it!



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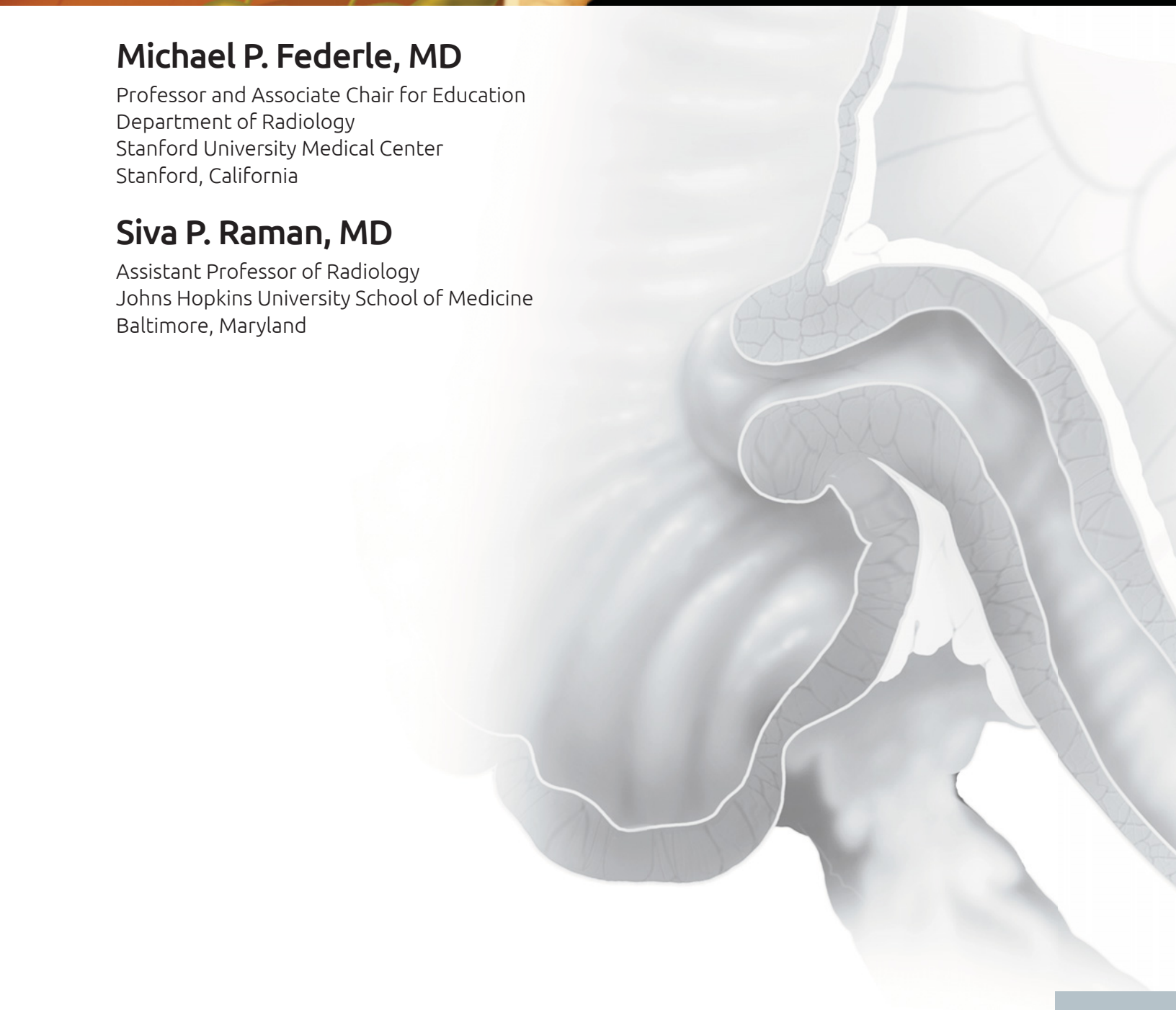

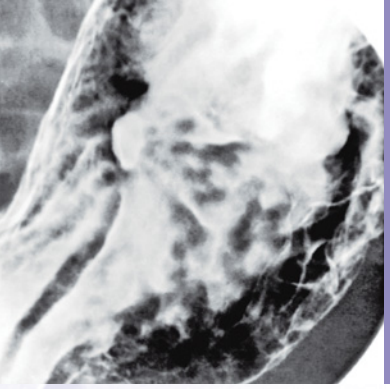

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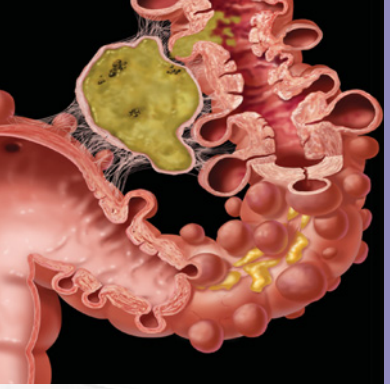

Sections

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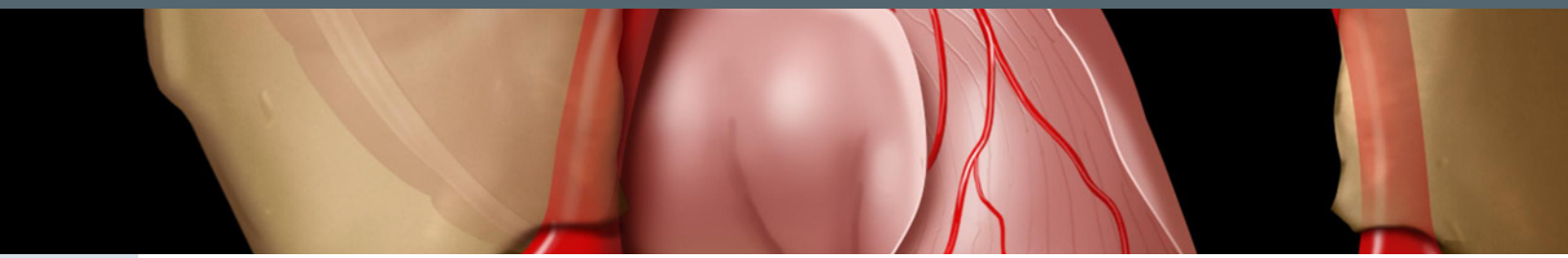

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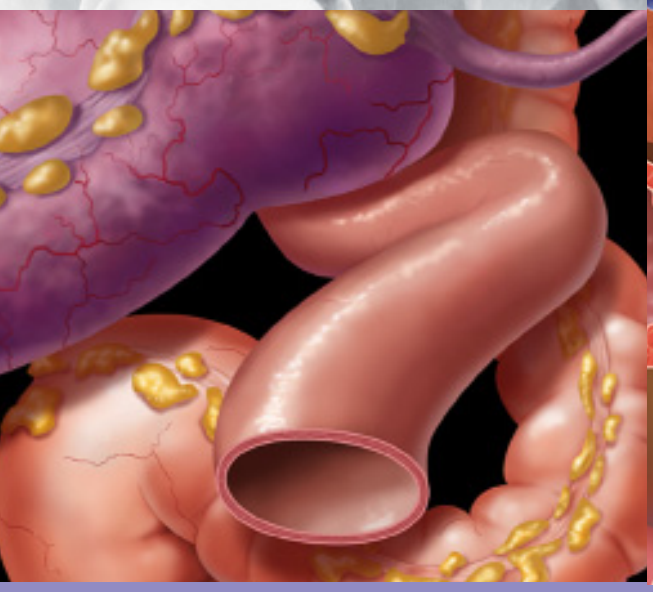

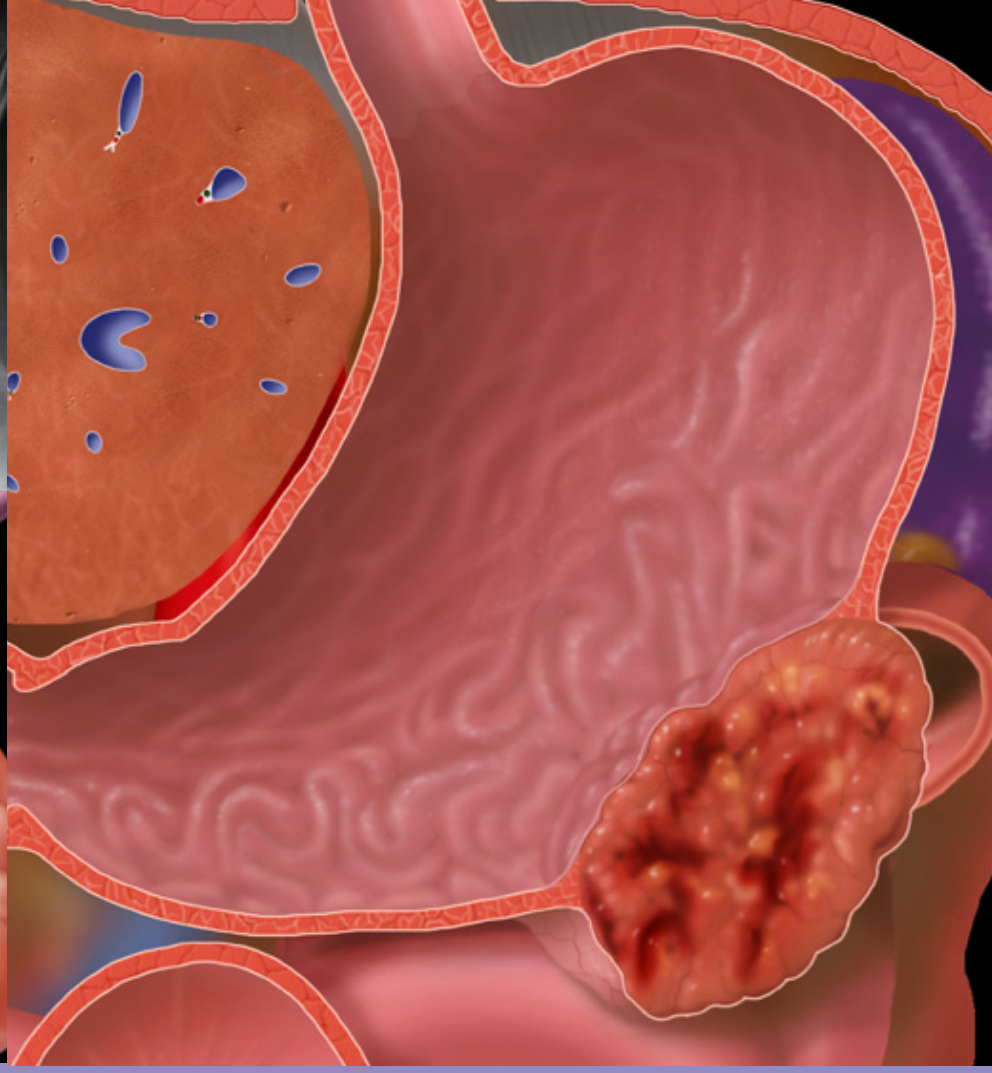

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SECTION 1

Abdominal Manifestations of Systemic Conditions

Organizational Approach to Abdominal Diseases

Most information about imaging abdominal disorders, including the gastrointestinal and genitourinary systems, fits neatly into an organ-by-organ framework. However, this approach makes it difficult to discuss diseases or conditions with manifestations throughout the abdomen and beyond. For this reason, some conditions are best discussed from a systemic perspective. Doing so provides a more accurate portrayal of these entities, and avoids unwanted redundancy.

Because many systemic disorders affect lymph node groups, neural structures, or major vessels throughout the abdomen, medical illustrations provide a helpful reminder of important anatomical considerations.

Systemic infections (including AIDS, tuberculosis, and mononucleosis) are discussed, along with important clues to help identify the infectious and neoplastic diseases they may cause or simulate.

Degenerative conditions, such as sarcoidosis and vascular disorders, are rarely limited to a single organ. These are presented in all their guises, along with tips as to how to address differential diagnoses.

Foreign bodies may be encountered throughout the gastrointestinal and genitourinary system and are well-known to be found repeatedly in certain individuals. Keys to recognition on imaging and avoiding common pitfalls are covered here.

Many malignant neoplasms are, by their very nature, systemic processes, such as lymphoma, leukemia, and malignant melanoma. Therefore, taking a systemic approach to such diagnoses gives us the opportunity to bring together some general principles about the presentation, diagnosis, and management of these important diseases.

Finally, while some conditions, such as systemic hypotension or hypervolemia, do not represent disease per se, they can result in important clinical and imaging abnormalities that must be recognized to avoid misguided patient management.

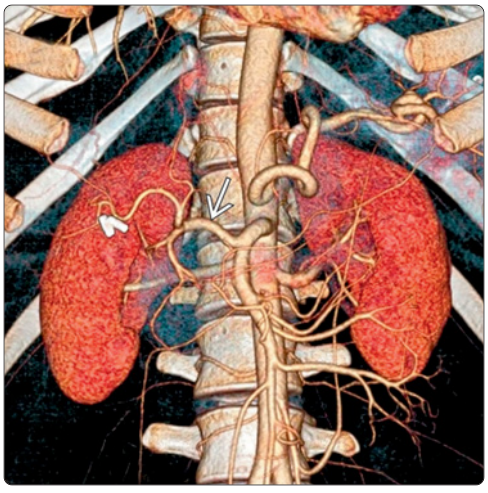
Imaging Modalities

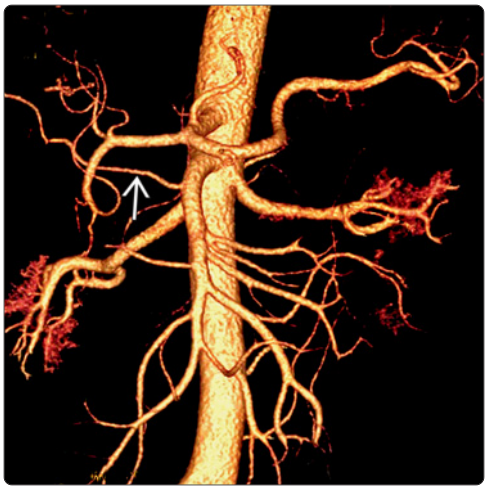
Plain radiography maintains an important role for surveillance of some generalized disease processes, such as the osseous and visceral manifestations of sickle cell anemia or cystic fibrosis.

Ultrasound is an important imaging tool for the evaluation of biliary, vascular, gynecologic, and scrotal pathology, but it lacks both sensitivity and specificity in evaluating other processes, especially bowel pathology.

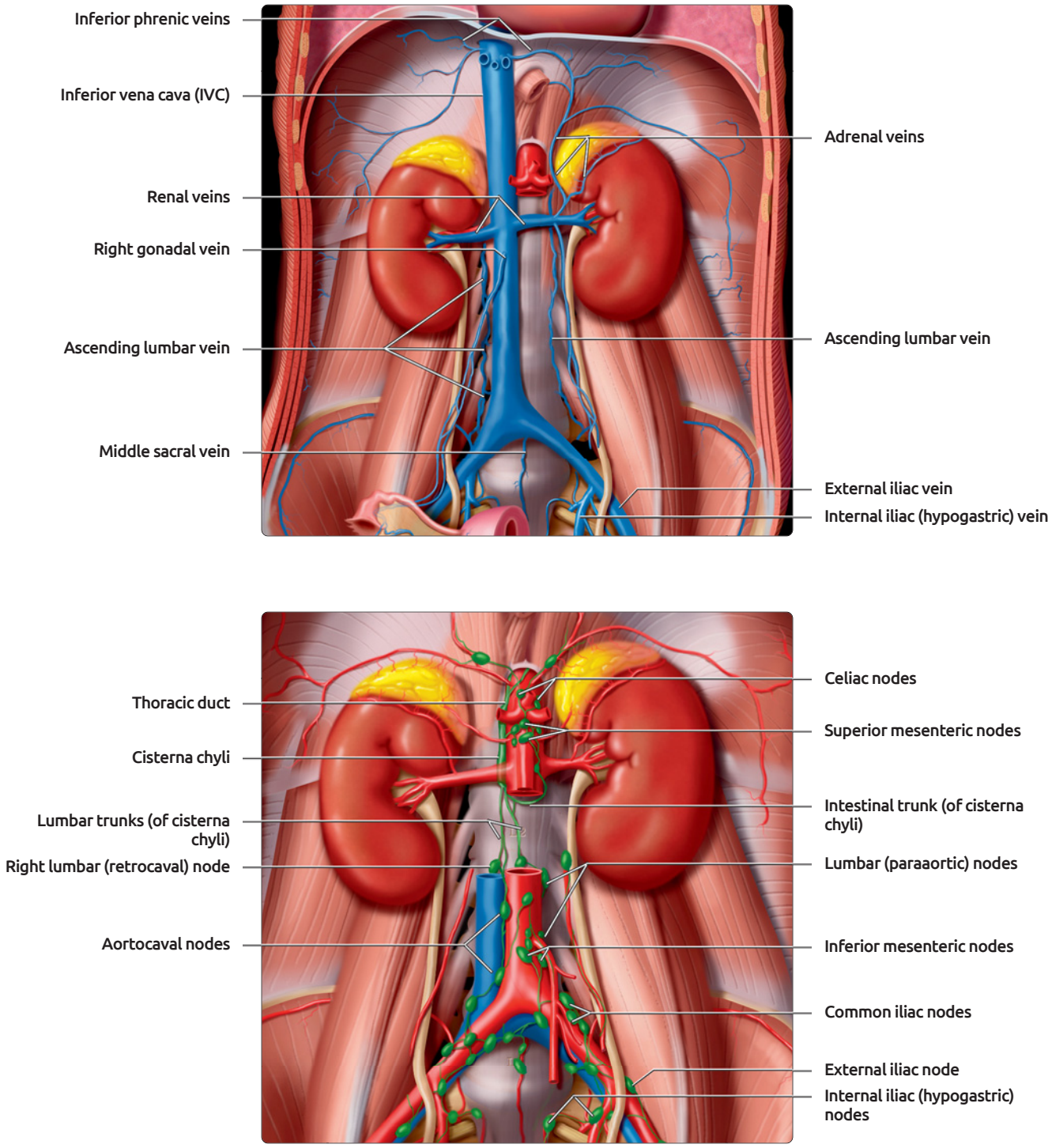
Computed tomography (CT) has become the essential tool for the comprehensive evaluation of most traumatic, inflammatory, and neoplastic abdominal processes. In patients with cancer, for instance, the ability to quickly and accurately examine different anatomic areas (thorax, abdomen, and pelvis), organs, and structures of different composition (e.g., lung, liver, and bone) is a tremendous advantage. Thus, there is continued growth and popularity of CT even in this era of powerful "competing" modalities, such as positron emission tomography (PET) and magnetic resonance (MR) imaging. PET and MR imaging do serve an important role as problem-solving tools for evaluating abdominal pathology. MR, with its excellent soft tissue characterization, is particularly helpful in evaluating masses within solid abdominal organs.

Catheter angiography remains the most accurate means of identifying certain vascular disorders and often results in catheter-based therapies in the same setting. For vasculitides, which routinely affect vessels throughout the body, angiography maintains an essential diagnostic and therapeutic role.



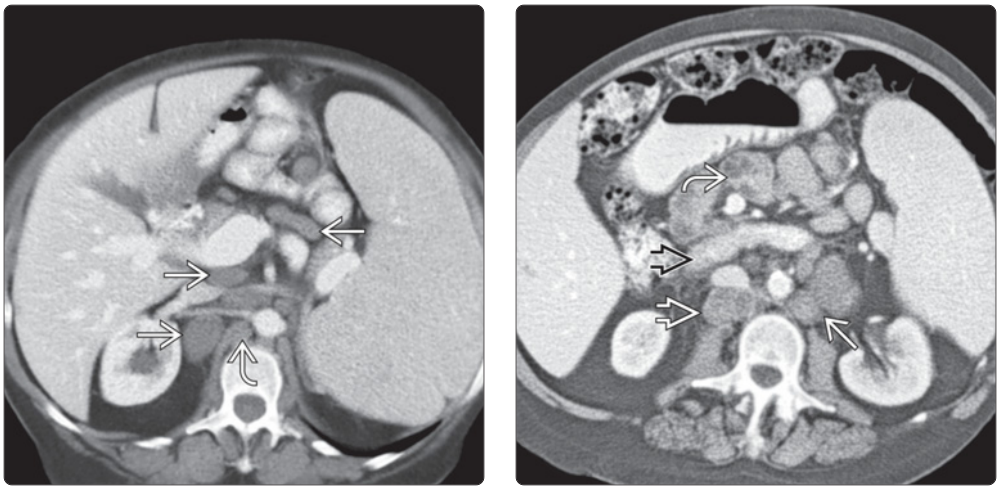


(Left) *Coronal volume-rendered CTA shows the entire common hepatic artery ➔ arising from the superior mesenteric artery. The left gastric artery also has a separate origin from the aorta, though difficult to perceive on this image. The "celiac trunk" in this patient consists only of the splenic artery. Congenital variations of vascular anatomy are very common.* **(Right)** *Oblique view of CTA clearly shows the origin of the accessory right hepatic artery ➔ from the superior mesenteric artery.*

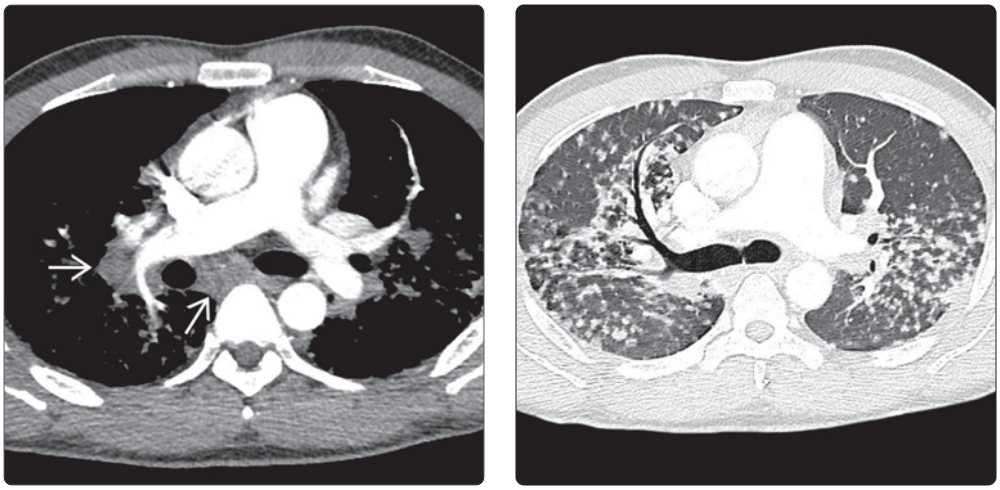


(Top) *The inferior vena cava (IVC) is formed by the confluence of the common iliac veins, which are formed by the confluence of the internal and external iliac veins. Note the ascending lumbar veins, which anastomose freely between the IVC and azygous, hemiazygos, and renal veins. These form a pathway for collateral flow in the event of IVC obstruction and play an important role in the systemic spread of pelvic tumors and infection.* **(Bottom)** *The major lymphatics and lymph nodes of the abdomen are located along, and share the same name as, the major blood vessels.*

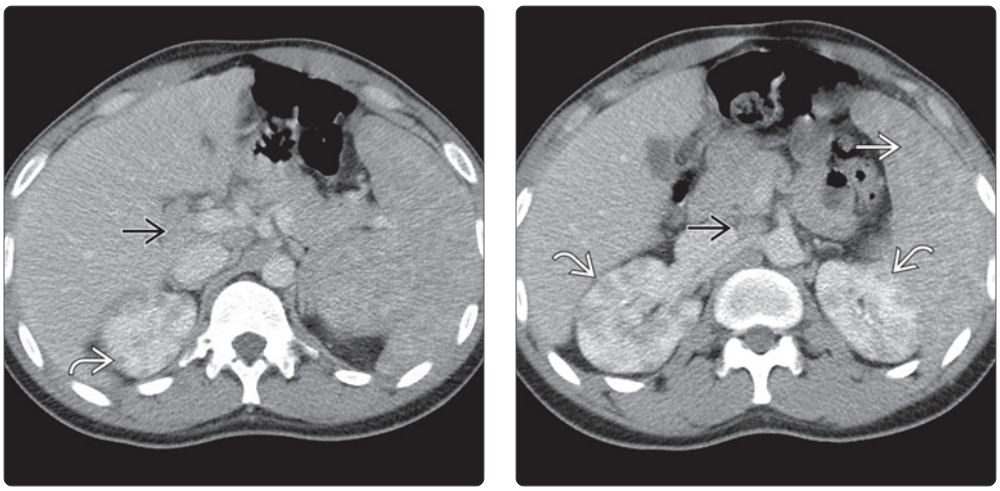
(Left) *Axial CT in a 50-year-old woman with non-Hodgkin lymphoma (NHL) shows splenomegaly and marked enlargement of multiple upper abdominal ➡ and retrocrural ➡ lymph nodes.* **(Right)** *On this CT section in the same case, the duodenum ⇨ is displaced by large retroperitoneal nodes; the mesenteric vessels are surrounded or "sandwiched" by mesenteric nodes ➡. The lumbar nodes are often referred to as para- or retroaortic ➡ (or -caval) ⇨, indicating their position relative to the great vessels.*

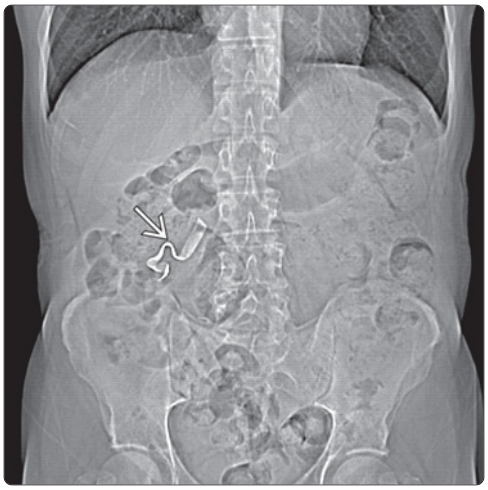


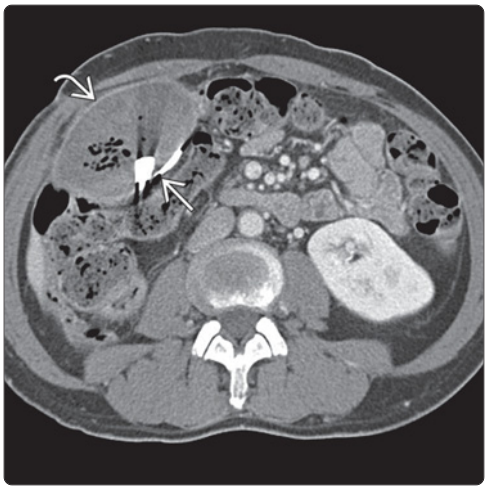
(Left) *This 33-year-old African American woman presented with dyspnea and general weakness. CT shows bilateral hilar and subcarinal lymphadenopathy ➡.* **(Right)** *CT at lung windows in the same patient shows diffuse pulmonary nodules predominantly in a peribronchial distribution.*



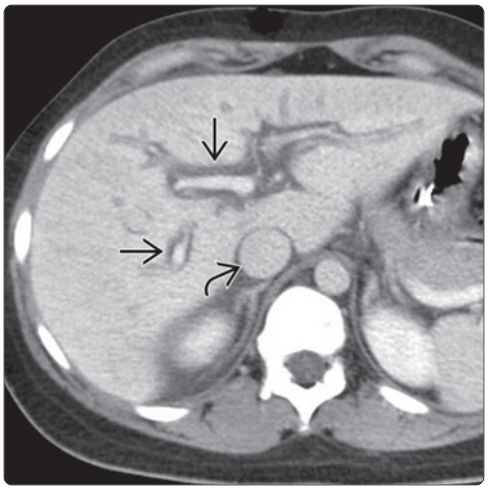
(Left) *CT in the same patient shows massive splenomegaly with innumerable small, poorly defined, hypodense nodules. Similar lesions were present in the liver, better seen on narrow window-width images (not shown). There are innumerable focal hypodense nodules ➡ in both kidneys, as well as upper abdominal lymphadenopathy ⇨.* **(Right)** *CT in the same patient shows more of the splenic ➡, renal ➡, and nodal ⇨ disease. All lesions were found to represent sarcoidosis and responded to steroid medication.*

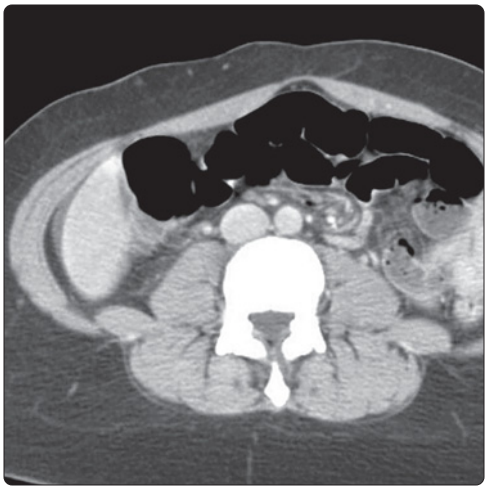




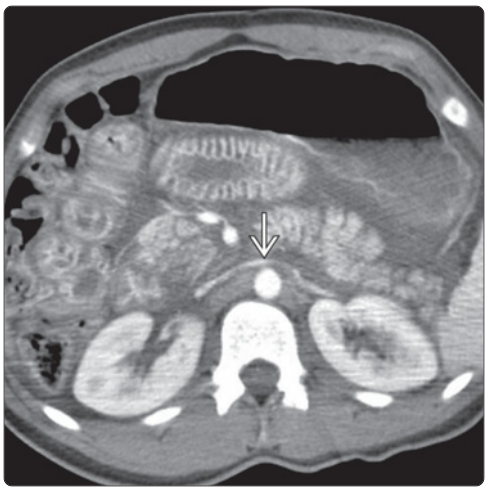


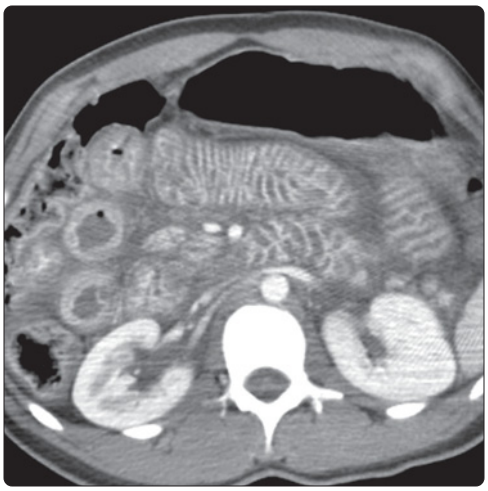
(Left) *This woman had abdominal pain for several months following laparoscopic right nephrectomy. A digital radiograph shows a curvilinear radiopaque stripe within the right side of the abdomen.* **(Right)** *CT in the same patient shows an encapsulated collection of fluid and gas density with an adjacent thin, radiopaque structure that corresponds to the stripe seen on the radiograph. This is a classic gossypiboma, a retained surgical sponge that has resulted in a chronic abscess or foreign body reaction.*





(Left) *This young man was injured in a motor vehicle crash (MVC). CT shows a distended IVC and periportal edema, which might be mistaken for dilated bile ducts or hepatic injury.* **(Right)** *CT in the same patient shows water density ascites in the Morison pouch. There was no hemoperitoneum nor visceral injury. The findings were due to aggressive IV hydration of the patient and resolved by the following morning.*





(Left) *This young man was injured in an MVC. CT shows diffuse infiltration of the peripancreatic and mesenteric fat planes. The IVC and renal veins appear flattened.* **(Right)** *CT in the same patient shows the classic "shock bowel" appearance of intense mucosal enhancement and submucosal edema. All of these findings are explainable by severe hypotension alone. There was no abdominal visceral or bowel injury, and a repeat CT scan the next morning was completely normal.*

HIV/AIDS

KEY FACTS

TERMINOLOGY

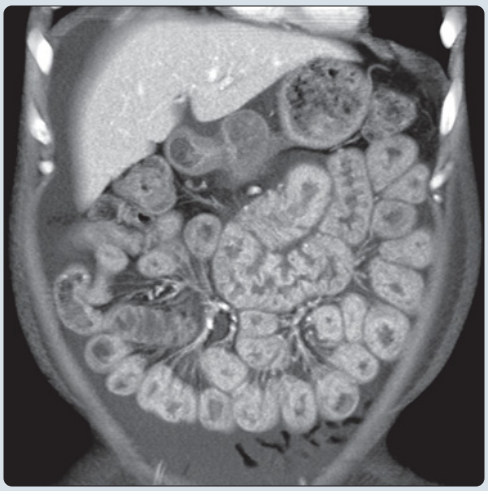
- Abdominal opportunistic infections and neoplasms resulting from HIV/AIDS-related immunodeficiency

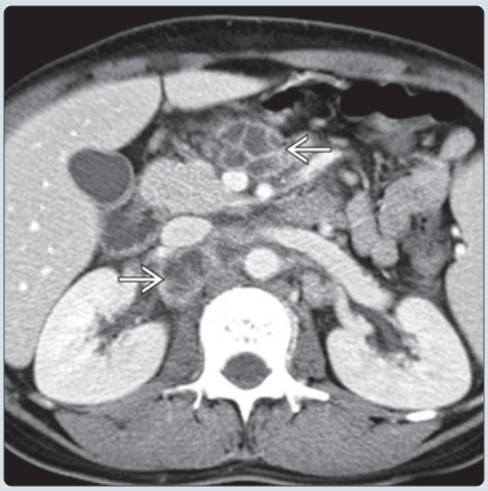
IMAGING

- **Liver and spleen**
  - Small hypodense nodules may be microabscesses
  - Larger hypodense lesions might be infectious, but AIDS-related lymphoma should be considered
  - *Pneumocystis* may result in tiny calcifications
- **Biliary tree**
  - Cholangitis or acalculous cholecystitis caused by opportunistic infections
- **Stomach, small bowel, and large bowel**
  - Wall thickening raises concern for opportunistic infection, which can involve any segment of GI tract
  - Mural thickening of esophagus suggests esophagitis, often due to candidiasis, CMV, or HSV
  - Proctitis in homosexual men related to sexual activity may be due to *Neisseria gonorrhoeae*, *Chlamydia*, or HSV
  - Focal mass-like wall thickening in GI tract should raise concern for malignancy (lymphoma, Kaposi sarcoma)
- **Lymph nodes**
  - Mild generalized lymphadenopathy (usually < 1.5 cm) is typically reactive and may be 1st clue to HIV infection
  - More significant adenopathy (> 1.5 cm) suggests opportunistic infection or AIDS-related lymphoma
- **Kidney**
  - Bilateral large kidneys (↑ echogenicity on US) with urothelial thickening due to HIV nephropathy

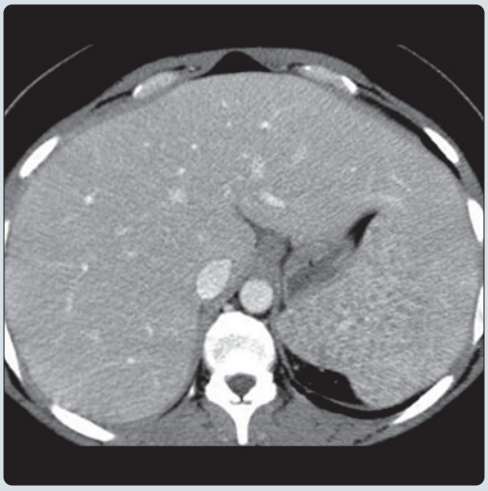
PATHOLOGY

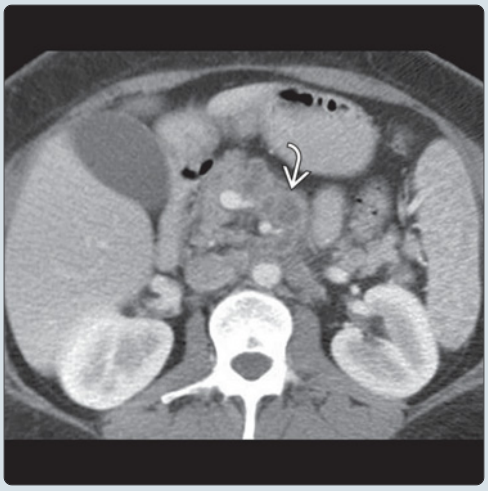
- Infections more common in HIV patients even with CD4 > 200, although risk ↑ substantially with lower CD4 counts
- Incidence of AIDS-defining malignancies (AIDS-related non-Hodgkin lymphoma, Kaposi sarcoma) has dramatically ↓ with antiretroviral therapy





(Left) *Coronal volume-rendered CECT in an AIDS patient with low CD4 count demonstrates diffuse thickening of the small bowel with surrounding ascites. The bowel appeared similar on several subsequent studies, and this was found to be infection with MAI.* **(Right)** *Axial CECT in an HIV-positive patient presenting with 3 weeks of fever, diarrhea, and weight loss shows multiple sites of low-attenuation lymphadenopathy ➡ involving retroperitoneal and mesenteric nodes. Biopsy confirmed MAI.*





(Left) *Axial CECT shows innumerable small hypodense foci in the spleen and, more subtly, in the liver. Both the liver and spleen are enlarged.* **(Right)** *Axial CECT in the same patient demonstrates multiple low-density enlarged lymph nodes ➡. This constellation of findings was found to represent disseminated mycobacterial infection.*

TERMINOLOGY

Abbreviations

- Acquired immune deficiency syndrome (AIDS)
- Human immunodeficiency virus (HIV)

Definitions

- Abdominal opportunistic infections and neoplasms resulting from HIV/AIDS-related immunodeficiency

IMAGING

General Features

- Location
  - Can affect visceral organs, gastrointestinal tract, genitourinary tract, and lymph nodes
- Size
  - Variable: Ranges from microabscesses (< 1 cm) to large masses due to lymphoma or Kaposi sarcoma

Imaging Recommendations

- Best imaging tool
  - CECT

CT Findings

- **Liver**
  - Liver may appear nodular and cirrhotic due to strong demographic overlap of HIV and chronic viral hepatitis
  - Small hypodense nodules scattered throughout liver suggests microabscesses (often due to *Mycobacterium avium-intracellulare* [MAI], tuberculosis, histoplasmosis, *Candida*, *Pneumocystis*, etc.)
  - Liver may appear globally enlarged without focal lesions due to infiltrative infections (e.g., MAI)
  - *Pneumocystis* (and rarely CMV or MAI) can result in multiple tiny calcifications throughout liver
    - Calcifications do not signify inactive disease
  - Liver involved in up to 1/4 of patients with AIDS-related lymphoma with hypodense nodules of variable size
- **Biliary tree**
  - Cholangitis caused by opportunistic infections
    - Intrahepatic and extrahepatic biliary strictures with papillary stenosis: Bile ducts may appear thickened and enhancing
    - Bile ducts may have beaded appearance very similar to primary sclerosing cholangitis
  - Acalculous cholecystitis due to opportunistic infections (e.g., CMV, *Cryptosporidium*)
    - Thickened gallbladder with pericholecystic fluid and stranding
- **Spleen**
  - Splenomegaly in up to 3/4 of AIDS patients without infection or tumor
  - Small tiny hypodense foci (microabscesses) usually due to disseminated infection (e.g., *Candida*, MAI, tuberculosis, coccidioidomycosis, *Pneumocystis*, etc.)
  - Larger hypodense lesions might still be infectious, but AIDS-related lymphoma should also be considered
  - Small calcifications (similar to liver) from *Pneumocystis*
- **Stomach, small bowel, and large bowel**
  - Bowel wall thickening, mucosal hyperemia, and fat stranding surrounding bowel should always raise concern for infection (including opportunistic infections)
    - CMV-related ulcerations of bowel may lead to GI tract perforation (one of the most common reasons for emergent abdominal surgery in AIDS patients)
  - Most opportunistic infections can involve any segment of GI tract (*Cryptosporidium*, CMV, MAI, tuberculosis, microsporidium, *Clostridium difficile*, amebiasis, etc.)
    - Difficult to predict pathogen based on distribution, but some organisms have predisposition for certain locations
      - CMV and TB tend to involve ileum
      - *Giardia*, microsporidium tend to involve proximal small bowel
      - Colon infections often due to CMV, *C. difficile*, *Campylobacter*, amebiasis, *Salmonella*, and *Shigella*
  - Mural thickening of esophagus suggests esophagitis, often due to candidiasis, CMV, or herpes simplex
  - Proctitis in homosexual men due to sexual activity may be due to *Neisseria gonorrhoeae*, chlamydia, or HSV
  - Focal mass-like wall thickening anywhere in GI tract should raise concern for malignancy (AIDS-related lymphoma, Kaposi sarcoma)
    - Lymphoma associated with intussusceptions
- **Lymph nodes**
  - Mild generalized lymphadenopathy (< 1.5 cm) is usually reactive and may be 1st clue to HIV infection
    - May persist for years in absence of symptoms (i.e., persistent generalized lymphadenopathy)
  - More significant adenopathy (> 1.5 cm) suggests opportunistic infection (MAI, tuberculosis) or AIDS-related lymphoma/Kaposi sarcoma
    - Necrotic mesenteric nodes from MAI or tuberculosis
    - Hyperenhancing lymph nodes in Kaposi sarcoma
  - AIDS-related lymphoma may be associated with discrete lesions in liver/spleen or focal mass in GI tract
    - GI tract most common extranodal site of involvement (75%), most often involving colon, ileum, and stomach
- **Kidney**
  - Bilateral large kidneys with urothelial thickening due to HIV nephropathy
  - Focal hypodense lesions could reflect infection (tuberculosis, MAI, fungus) or AIDS-related lymphoma
  - Calcifications may be present in setting of *Pneumocystis* (similar to liver and spleen) or rarely MAI/CMV
- **Pancreas**
  - Opportunistic infections can cause acute pancreatitis and pancreatic duct strictures (e.g., CMV, *Cryptococcus*, etc.)

Ultrasonographic Findings

- **Kidney**
  - HIV nephropathy: Normal sized or enlarged kidneys with increased echogenicity (kidney > liver)
    - May be associated with urothelial thickening in pelvis/intrarenal collecting system
    - Parenchymal heterogeneity and loss of corticomedullary differentiation
  - Hyperechoic foci or calcifications without posterior acoustic shadowing due to *Pneumocystis*, MAI, or CMV
- **Gallbladder**

- GB wall thickening may be reactive due to hepatitis or secondary to opportunistic acute acalculous cholecystitis
- Wall thickening and dilation of extrahepatic &/or intrahepatic bile ducts due to AIDS cholangiopathy

- **Liver**
  - Opportunistic infections present as small hypoechoic nodules (microabscesses) scattered throughout liver
  - *Pneumocystis* may result in small hypoechoic nodules or tiny echogenic foci
- **Lymph nodes**
  - Necrotic nodes most often due to MAI or tuberculosis

DIFFERENTIAL DIAGNOSIS

Lymphoma Unrelated to HIV/AIDS

- Nodal involvement more common, unlike AIDS, where extranodal involvement is disproportionately common
- AIDS-related lymphoma often aggressive with widespread dissemination, whereas non-AIDS related lymphoma may present with early stage disease confined to nodes

Biliary Hamartomas

- Multiple small cystic lesions scattered throughout liver
- May mimic hepatic microabscesses, but patients are asymptomatic without signs of infection

Sarcoidosis

- May present with multiple small hypodense lesions in liver and spleen (mimicking microabscesses)
- Upper abdominal adenopathy frequently present, and may be mistaken for HIV-related adenopathy
- Mediastinal and hilar lymphadenopathy, characteristic lung disease, and lack of symptoms may allow distinction

PATHOLOGY

General Features

- Etiology
  - HIV results in immunodeficiency through infection and lysis of CD4(+) T cells
  - HIV-infected patients have an increased risk of developing malignancies, particularly when coinfected by Epstein-Barr virus, herpesvirus, or papillomavirus
    - Incidence of AIDS-defining malignancies (AIDS-related non-Hodgkin lymphoma, Kaposi sarcoma) has dramatically ↓ with antiretroviral therapy
    - Risk of other malignancies, which are often atypically aggressive and occur at younger ages than normal, still higher in HIV patients
    - **Non-Hodgkin lymphoma**
      - AIDS-defining malignancy (usually CD4 count < 100) that includes several types of lymphoma, including diffuse large B-cell and Burkitt lymphoma
      - Strong tendency to arise in extranodal sites (especially GI tract), involve unusual locations, and present with advanced disease
    - **Kaposi sarcoma**
      - Low-grade soft tissue sarcoma of vascular origin associated with HHV-8 infection
  - Infections more common in HIV patients even with CD4 counts > 200, although risk increases substantially with lower CD4 counts
    - Many different AIDS-defining infections, including disseminated MAI, tuberculosis, *Pneumocystis* infection, recurrent bacterial pneumonias, persistent *Cryptosporidium* infection, chronic HSV, etc.
      - Most occur when CD4 count < 200, but can rarely occur at higher CD4 counts

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Acute HIV infection may resemble mononucleosis, with fever, headaches, and body aches
  - Many patients with chronic HIV infection asymptomatic when effectively treated with antiretrovirals
    - Skin abnormalities and mild constitutional symptoms possible even without immunosuppression
  - Patients with advanced HIV/AIDS and immunosuppression may experience symptoms related to opportunistic infections (diarrhea, cough/shortness of breath, abdominal pain, etc.)
    - Some patients experience wasting syndrome with profound weight loss and chronic diarrhea
- Other signs/symptoms
  - Patients with low CD4 counts frequently pancytopenic (anemia, thrombocytopenia, and lymphopenia)
  - Generalized lymphadenopathy and splenomegaly common even in absence of active infection
- Clinical profile
  - Clinical profile varies from country to country
    - HIV in developing world spread primarily by vaginal sex (small proportions due to IV drug abuse and perinatal transmission)
    - HIV in USA disproportionately associated with IV drug abuse and homosexual sexual contact

Demographics

- Age
  - Primarily adults, but perinatal transmission possible
- Gender
  - Worldwide most cases in heterosexuals, with F > M
- Epidemiology
  - > 35 million affected worldwide

Natural History & Prognosis

- Multiple opportunistic infections and AIDS-related tumors unless antiretroviral drugs used to suppress HIV
- AIDS defined as CD4 < 200 or development of AIDS-defining illness (either infection or malignancy)

Treatment

- Antiretroviral drugs to preserve immune status
- Antibiotics for bacterial infections and antiviral drugs for CMV infection

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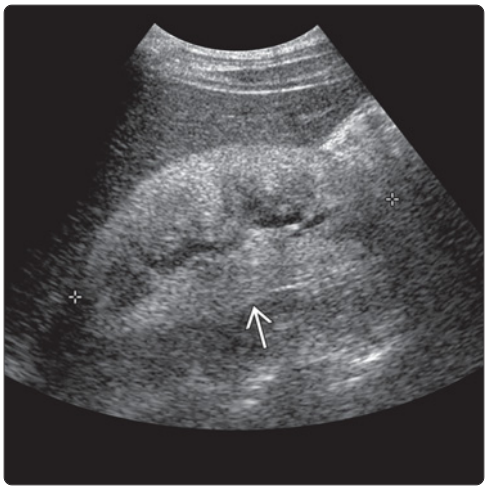
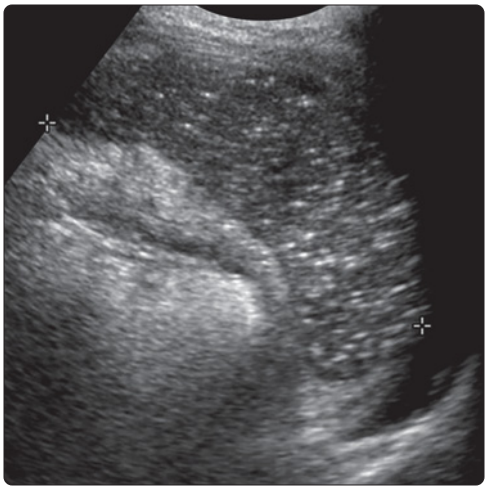
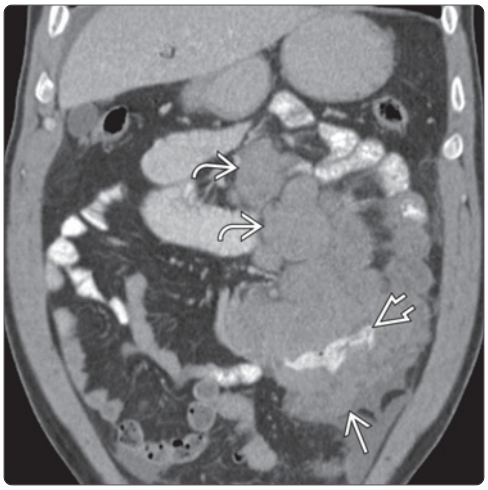
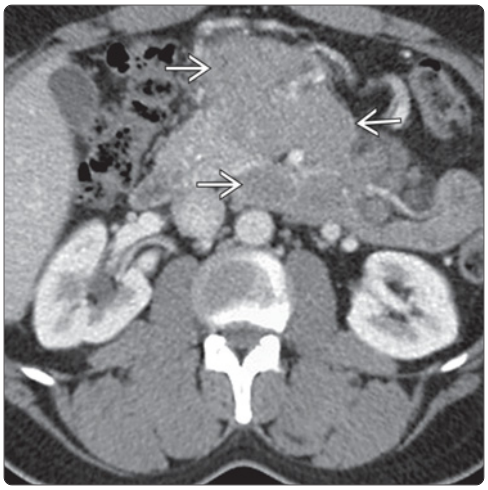


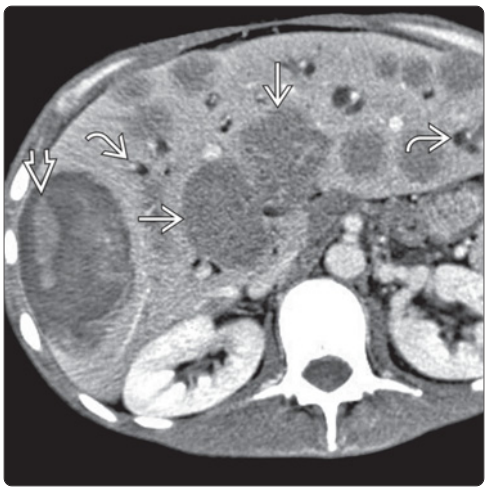


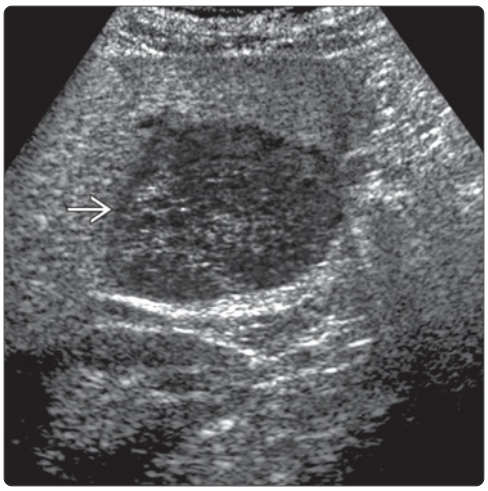
(Left) *Sagittal ultrasound demonstrates a normal-sized right kidney, which is markedly echogenic, compatible with the patient's known HIV nephropathy. Unlike other forms of chronic renal failure, the kidneys in HIV nephropathy are often normal in size or enlarged.* **(Right)** *Transverse ultrasound demonstrates innumerable tiny calcifications in the spleen of an HIV patient, representing the sequelae of the patient's known prior Pneumocystis infection.*





(Left) *Coronal CECT in an AIDS patient demonstrates diffuse mass-like wall thickening and aneurysmal dilatation of a loop of small bowel in the left lower quadrant with internal enteric contrast. Note the extensive lymphadenopathy more superiorly. These findings are compatible with the patient's biopsy-proven AIDS-related non-Hodgkin lymphoma.* **(Right)** *Axial CECT in an AIDS patient demonstrates extensive mesenteric lymphadenopathy found to represent AIDS-related non-Hodgkin lymphoma.*





(Left) *Axial CECT in an AIDS patient illustrates multiple hepatic masses, including a mass with internal hemorrhage, which were proven to be non-Hodgkin lymphoma. An unusual feature in this case is the mild obstruction of the intrahepatic bile ducts.* **(Right)** *Longitudinal ultrasound in a patient with AIDS demonstrates a large hypoechoic mass. Biopsy revealed this to represent AIDS-related B-cell non-Hodgkin lymphoma.*

Tuberculosis

KEY FACTS

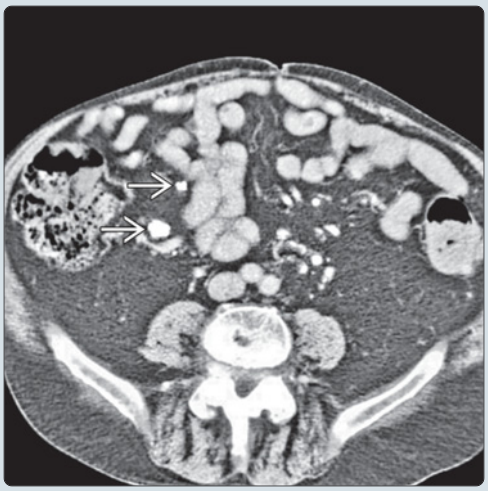
IMAGING

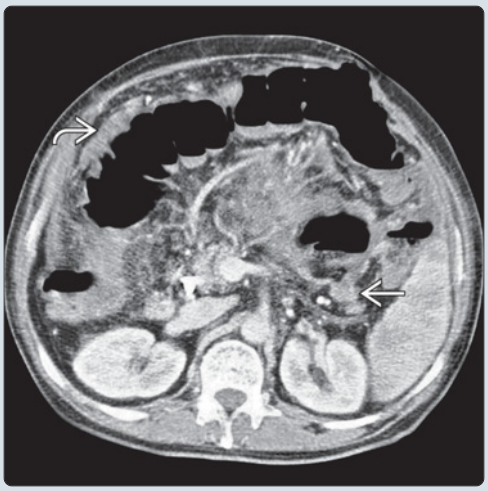
- Most common sites of involvement in abdomen are lymph nodes, GU tract, peritoneum, and GI tract
  - Abdominal lymphadenopathy is most common
- **Lymphadenopathy** (tuberculous lymphadenitis)
  - Enlarged, centrally necrotic nodes with hypoattenuating centers and hyperattenuating enhancing rims
  - Nodes often calcify after healing
- **Tuberculosis peritonitis**
  - Variables amounts of free or loculated complex ascites with infiltration of omentum ± discrete masses
- **Gastrointestinal tuberculosis**
  - Ileocecal region affected in 90% of cases
  - Asymmetric wall thickening of ileocecal valve and medial cecum
- **Adrenal tuberculosis**
  - Acute: Enlarged adrenals (often appears as discrete, centrally necrotic adrenal mass)
  - Chronic: Small adrenals with dots of calcification and low signal on all MR sequences
- **Renal tuberculosis**
  - Most common CT finding is renal calcification (50%)
  - Papillary necrosis is a very common early finding
  - Focal wedge-shaped hypodense areas, small hypodense nodules, or discrete renal abscess
  - Urothelial thickening, caseous debris, and strictures of calyces and infundibuli may lead to hydronephrosis
- **Hepatosplenic tuberculosis**
  - Hepatosplenomegaly with hypodense nodules of variable size

CLINICAL ISSUES

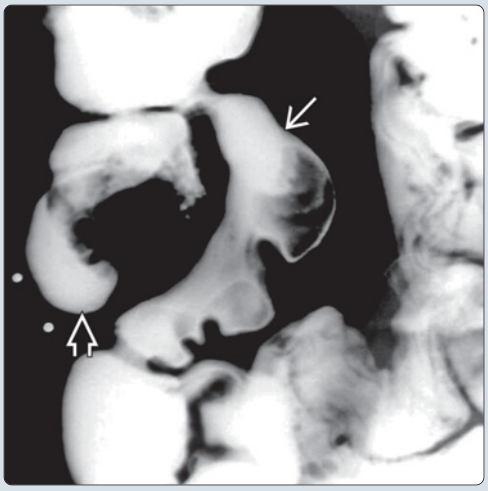
- Often presents with fever, weight loss, and abdominal pain
- May or may not have evidence of pulmonary TB
  - Negative chest radiograph or negative tuberculin skin test does not exclude extrapulmonary TB

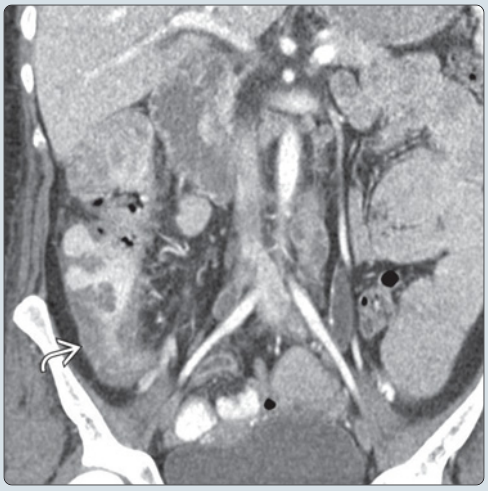
(Left) *Axial CECT in an asymptomatic elderly man shows calcification of mesenteric nodes ⇒ usually seen in elderly individuals who have had exposure to enteric mycobacteria, often from drinking unpasteurized milk.* **(Right)** *Axial CECT in a liver transplant recipient shows marked thickening of the omentum ⇒, peritoneum, and mesentery, with enlargement of mesenteric nodes ⇒. Loculated ascites was also present (not shown). This patient's reactivated TB with TB peritonitis was first acquired in his native country.*





(Left) *Spot film from a small bowel follow-through in a 25-year-old immigrant from India shows deformity of the terminal ileum ⇒ and cecum ⇒, with asymmetric thickening and stiffening of the bowel walls, ultimately found to represent TB.* **(Right)** *Coronal CECT in an immigrant patient demonstrates asymmetric thickening ⇒ of the cecum, which has a cone-shaped appearance in a patient with tuberculous colitis.*





TERMINOLOGY

Abbreviations

- Tuberculosis (TB)

Definitions

- Infection by *Mycobacterium tuberculosis*

IMAGING

General Features

- Best diagnostic clue
  - Most common sites of involvement in abdomen are lymph nodes, GU tract, peritoneum, and GI tract
    - Abdominal lymphadenopathy most common (2/3 cases)
    - GU tract is most common organ system involved

Imaging Recommendations

- Best imaging tool
  - CECT

Radiographic Findings

- Often no evidence of lung disease (CXR or CT can be normal)
- **Lymphadenopathy** (tuberculous lymphadenitis)
  - Can range from increased number of normal-sized nodes to massively enlarged conglomerate nodal masses
    - Mesenteric and peripancreatic lymph nodes most commonly involved
    - Multiple groups often affected simultaneously
  - Enlarged, necrotic nodes with hypoattenuating centers and hyperattenuating enhancing rims on CT (40-60%)
    - Characteristic of caseous necrosis
    - Mixed attenuation nodes are also possible
  - Nodes calcify with healing: TB probably most common cause of mesenteric nodal calcification
- **Tuberculosis peritonitis**
  - 3 imaging patterns: Wet, dry, and fibrotic fixed
    - Wet type: Large amount of free or loculated ascites
      - Higher than water density due to protein/cellular content
      - Complex ascites with septations or fibrinous strands
    - Dry type: Mesenteric and omental thickening, fibrous adhesions, and caseous nodules
    - Fibrotic fixed: Discrete masses in omentum with matted loops of bowel ± loculated ascites
  - CT is ~ 69% sensitive for TB peritonitis
    - Difficult to distinguish from carcinomatosis
    - Carcinomatosis more likely to demonstrate discrete implants or omental caking
- **Gastrointestinal tuberculosis**
  - Ileocecal region affected in 90% of cases
    - Common site due to presence of lymph tissue and stasis of bowel contents in that location
    - Cecum and terminal ileum are usually contracted (cone-shaped cecum) with asymmetric wall thickening of ileocecal valve and medial cecum
    - Regional lymphadenopathy with central caseation
  - Involvement of stomach and proximal small bowel is rare
    - Stomach: Affects antrum and distal body, often simulating peptic ulcer disease
    - Duodenum: Wall thickening and luminal narrowing
- **Hepatosplenic tuberculosis**
  - Micronodular pattern
    - Innumerable 0.5–2.0 mm nodules may or may not be discretely visualized (most often hypodense on CT and hyperechoic on US)
    - May simply appear as hepatomegaly on CT
  - Macronodular pattern
    - CT
      - Acute: Hypoattenuating nodules with ill-defined enhancing margins
      - Chronic: Tuberculomas often calcify
      - TB and histoplasmosis are most common causes of calcified granulomas
    - MR
      - T1WI: Hypointense, minimally enhancing, honeycomb lesions
      - T2WI: Hyperintense with less intense rim relative to surrounding liver
      - Rim enhancement on post-gadolinium images
- **Adrenal tuberculosis**
  - Unilateral (10%) or bilateral (90%)
  - Acute: Enlarged adrenals (often appear as discrete centrally necrotic adrenal masses)
  - Chronic: Small adrenals with dots of calcification and low signal on all MR sequences
  - May cause adrenal insufficiency (most common cause in developing countries)
- **Renal tuberculosis**
  - 75% unilateral
  - CT findings
    - Most common CT finding is renal calcification (50%)
      - Affected part of kidney often nonfunctional; global nonfunction and calcification = "putty" kidney
    - Papillary necrosis early finding (usually upper pole)
    - Focal wedge-shaped areas of low attenuation, multiple small hypodense nodules, or discrete renal abscess
    - Urothelial thickening, caseous debris, and strictures of calyces and infundibuli may lead to hydronephrosis
  - Intravenous urography: "Moth-eaten" calyx due to erosions and progression to papillary necrosis
    - Strictures of renal pelvis and infundibula
    - Caliectasis and hydronephrosis with irregular margins and filling defects due to caseous debris
    - Irregular pools of contrast due to parenchymal cavitation
- **Ureteral tuberculosis**
  - Usually secondary to renal TB
  - Thickened wall of ureter with strictures most common in distal 1/3 of ureter
  - Corkscrew/beaded ureter due to chronic fibrotic strictures
- **Bladder tuberculosis**
  - Decreased bladder volume with wall thickening, ulceration, and filling defects
  - Severe: Scarring → small, irregular, calcified bladder
- **Female genital tuberculosis**

- Most commonly involves fallopian tubes (in 94% of cases)
  - Bilateral salpingitis with strictures ± occlusion
- Can involve endometrium resulting in deformed, irregular endometrium on US

- **Male genital tuberculosis**
  - Affects seminal vesicles or prostate gland, rarely testes
  - Can resemble a pyogenic abscess ± calcification
- **Pancreatic tuberculosis**
  - Appears as mass mimicking cancer (caseated peripancreatic nodes involving pancreas)
    - US: Well-defined hypoechoic lesions
    - CT: Hypodense mass (usually pancreatic head) typically without pancreatic duct dilatation or vascular invasion

DIFFERENTIAL DIAGNOSIS

Peritonitis

- Nontuberculous peritonitis
- Peritoneal metastases and lymphoma
- Mesothelioma

Miliary Hepatic Lesions

- Hepatic metastases and lymphoma
- Hepatic opportunistic infection
- Sarcoidosis

Macronodular Hepatic Lesions

- Hepatic metastases and lymphoma
- Hepatic pyogenic abscess
- Primary hepatic malignancy

Ileocecal Lesions

- Amebiasis
- Crohn disease
- Primary cecal malignancy

Lymphadenitis

- Metastases or lymphoma
- Whipple disease
- *Mycobacterium avium-intracellulare* infection

Renal Lesions

- Renal papillary necrosis
- Renal transitional cell carcinoma
- Other infections
  - (e.g., pyelonephritis, xanthogranulomatous pyelonephritis)

Adrenal Lesions

- Adrenal metastases and lymphoma
- Primary adrenal neoplasm
- Adrenal hemorrhage

Bladder Lesions

- Bladder schistosomiasis
- Cytoxan cystitis
- Radiation-induced bladder calcification
- Calcified bladder carcinoma

PATHOLOGY

General Features

- Etiology
  - Primary infection from *M. tuberculosis*
  - Abdominal TB is usually secondary to pulmonary TB
    - CXR normal in 2/3 of patients with abdominal TB
    - Only 15% have active pulmonary disease
  - Other sources of abdominal infection with TB
    - Swallowing infected material
    - Hematogenous spread from active or latent infection
    - Direct extension from infected tissues

Microscopic Features

- Caseating granulomas are characteristic
- Microscopy and culture for mycobacteria

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Abdominal TB often presents with fever, weight loss, and abdominal pain
  - Negative chest radiograph or negative tuberculin skin test does not exclude extrapulmonary TB
    - May or may not have evidence of pulmonary TB
    - May or may not have positive tuberculin test
      - Possibly negative in immunosuppressed, malnourished, or severe disseminated disease
- Other signs/symptoms
  - Adrenal tuberculosis
    - Addisonian presentation (adrenal insufficiency, hypotension, and electrolyte disturbances)
  - Gastrointestinal TB
    - Usually few or no symptoms (partial obstruction)

Demographics

- Epidemiology
  - Resurgence of TB
    - ↑ in immunocompromised patients (especially those with AIDS)
    - Drug-resistant strains of *M. tuberculosis*
    - Estimated 1/3 of world population infected with TB
- Risk factors for TB
  - Immunocompromise (AIDS, transplant recipients, immunosuppressive drugs)
  - Poverty, homelessness, alcoholism, immigration from developing country, imprisonment

Treatment

- Surgery for emergent presentations
- 6-9 month course of multidrug antituberculous chemotherapy
  - Most commonly used drugs include rifampin, isoniazid, pyrazinamide, and ethambutol
  - Exact drug regimen may vary based on resistance patterns

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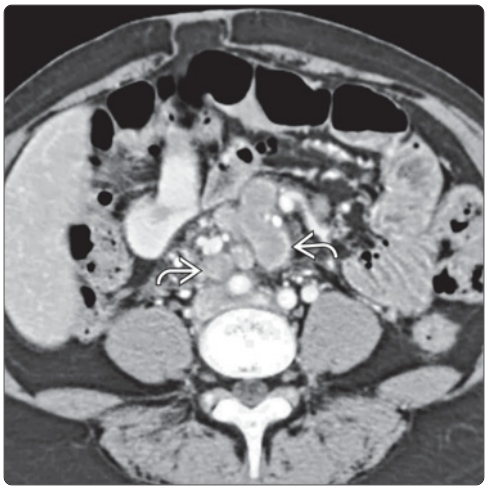
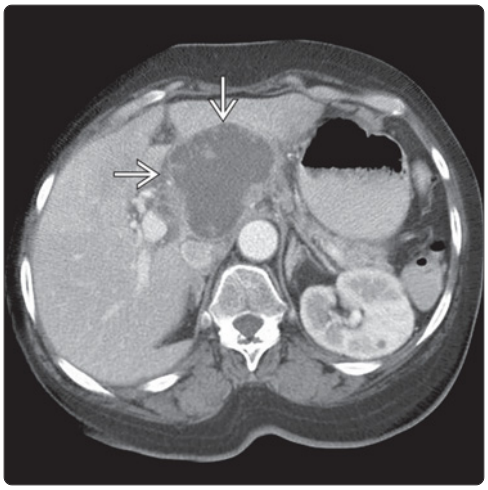
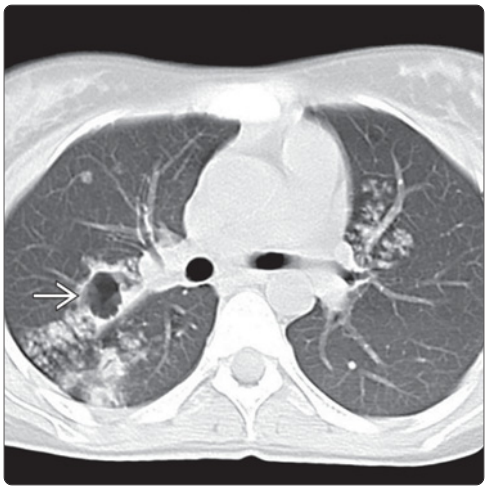
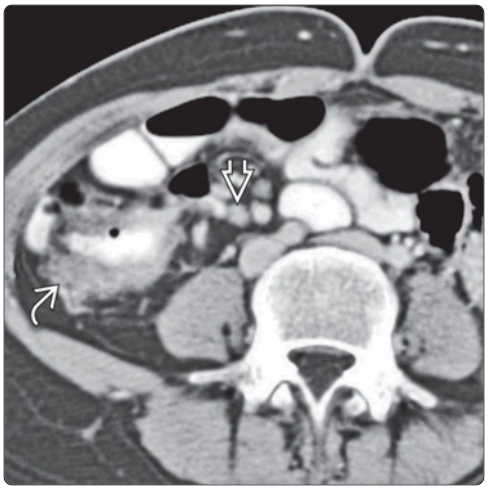


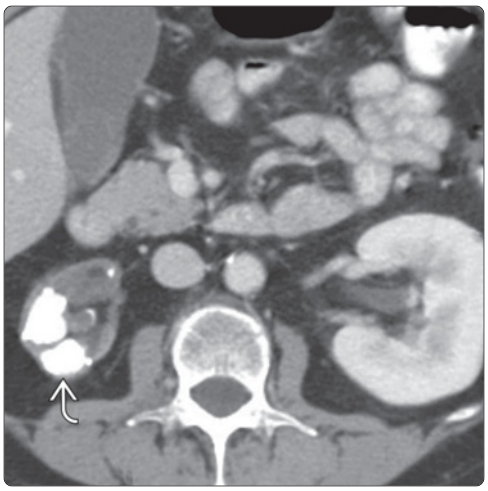


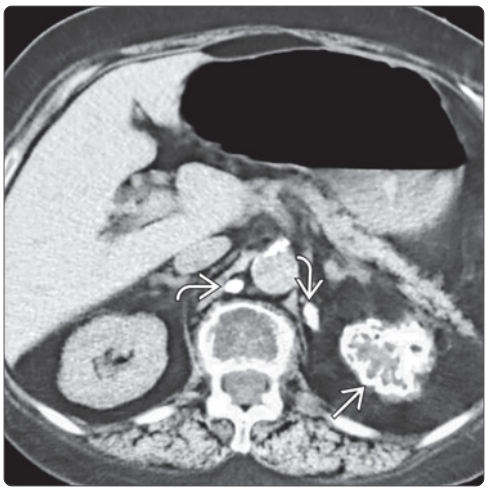
(Left) *Axial CECT in a young woman with AIDS demonstrates mesenteric and retroperitoneal lymphadenopathy. Some of the enlarged nodes have a caseated or low-density centrally necrotic appearance ➪ characteristic of mycobacterial infection.* **(Right)** *Axial CECT shows a large, complex cystic mass ➪ in the porta hepatis and pancreatic head region, representing conglomerate caseated, enlarged nodes due to Mycobacterium tuberculosis infection.*





(Left) *Axial CECT shows cavitary ➪ and multilobar bronchoalveolar infection of the lungs, typical of active tuberculosis. This patient was a young female college exchange student from Asia.* **(Right)** *Axial CECT in the same patient shows mural thickening of the cecum ➪, along with regional mesenteric lymphadenopathy ➪ typical of intestinal and nodal involvement by TB.*





(Left) *Axial CECT show a small, nonfunctional, and partially calcified "putty" kidney ➪, typical of chronic TB infection of the kidney. The patient had a known history of pulmonary TB.* **(Right)** *Axial NECT shows calcification from healed TB granulomas within retroperitoneal and retrocrural nodes ➪. The left kidney ➪ is totally calcified and nonfunctional, an autonephrectomy or "putty" kidney due to chronic renal TB. Small focal calcifications were also present in the adrenals.*

Mononucleosis

KEY FACTS

TERMINOLOGY

- Illness due to infection with Epstein-Barr virus (EBV)

IMAGING

- Splenomegaly in 60% of patients
  - Splenic rupture: Perisplenic and splenic subcapsular hematoma (sentinel clot)
  - Splenic infarct: Rare in mononucleosis, but may be due to transient thrombophilia predisposing to arterial thrombosis
- Hepatomegaly ± parenchymal heterogeneity and periportal edema
  - ± gallbladder wall thickening: May be reactive due to EBV hepatitis
- Generalized or upper abdominal lymphadenopathy

TOP DIFFERENTIAL DIAGNOSES

- Long differential, including other neoplastic, inflammatory, infectious, infiltrative, and hematologic diseases

PATHOLOGY

- EBV infection (human herpesvirus 4)
  - Replicates mainly in B lymphocytes but also in epithelial cells of pharynx and parotid duct
- Splenomegaly and lymphadenopathy
  - Due to congestion with activated T lymphocytes

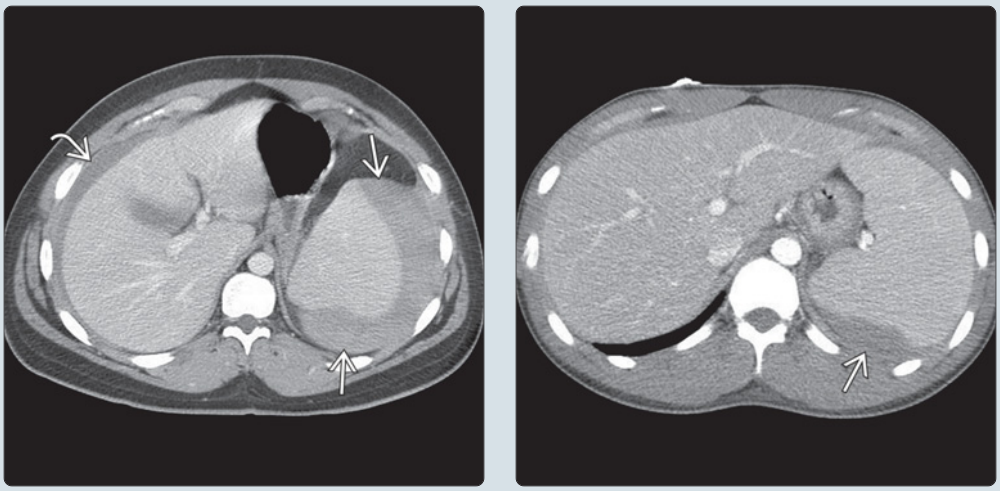
CLINICAL ISSUES

- Adolescents and young adults are most often affected
  - Uncommon in adults due to prior exposure/immunity
- Acute symptoms (e.g., sore throat, fever, headache) typically resolve in 1 month
  - Fatigue/myalgias may persist for several months
- Laboratory findings: Lymphocytosis, positive monospot test
- Treatment is typically supportive

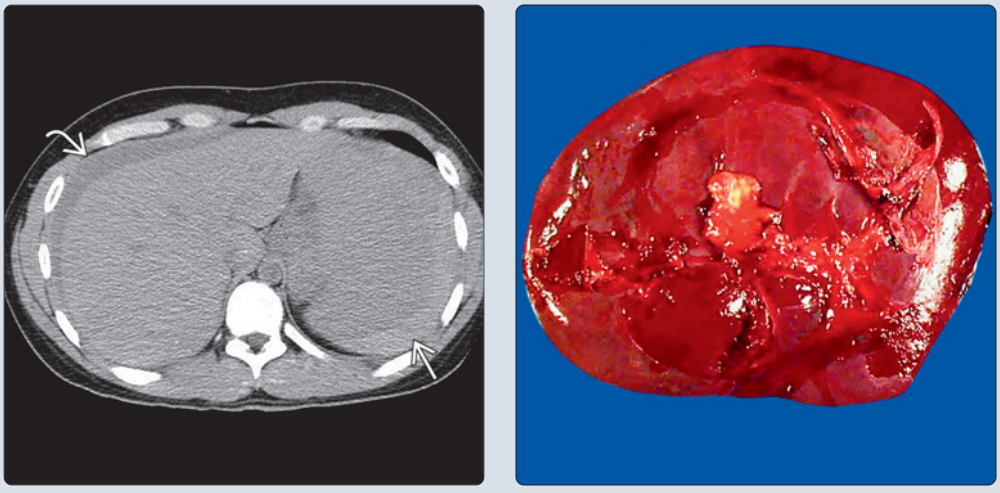
DIAGNOSTIC CHECKLIST

- Consider mononucleosis in previously healthy adolescent or young adult with splenomegaly and lymphadenopathy

(Left) *Axial CECT in a patient with mononucleosis shows clotted blood (sentinel clot) ➡ around an enlarged spleen and lower density free intraperitoneal hemorrhage ➡. This spontaneous splenic rupture resolved with nonoperative management.* **(Right)** *Axial CECT in a patient with mononucleosis shows an enlarged spleen with presplenic hematoma ➡ as a result of splenic rupture. This patient recovered without surgery.*



(Left) *Axial NECT shows an enlarged spleen with high-density adjacent sentinel clot ➡, and lower density free intraperitoneal blood ➡.* **(Right)** *Surgical specimen from the same patient shows a ruptured spleen, which was enlarged due to mononucleosis. The spleen was almost 20 cm in length, and a histologic exam showed that it was congested with activated T lymphocytes.*



TERMINOLOGY

Abbreviations

- Infectious mononucleosis (IM)

Definitions

- Illness due to infection with Epstein-Barr virus (EBV)

IMAGING

General Features

- Best diagnostic clue
  - Splenomegaly and abdominal lymphadenopathy in previously healthy adolescent or young adult

Imaging Recommendations

- Protocol advice
  - Imaging not needed unless complications are suspected
  - Contrast-enhanced CT for complications

Radiographic Findings

- Spleen
  - Splenomegaly
    - Common (60% of patients), even if spleen is not palpable on physical exam
  - Splenic rupture
    - Perisplenic and splenic subcapsular hematoma (sentinel clot sign on CT)
    - Enlarged spleen with areas of hypodensity on CT
  - Splenic infarct
    - Rare, but may be due to transient thrombophilia predisposing to arterial thrombosis
    - Wedge-shaped areas of splenic hypodensity on CT
- Liver and biliary tract
  - Hepatomegaly ± parenchymal heterogeneity and periportal edema
  - Thickened gallbladder may be reactive to EBV hepatitis
- Generalized or upper abdominal lymphadenopathy
- Focal lesions (nodular proliferation of EBV-infected cells or lymphomatoid granulomatosis) very rarely in spleen and liver

DIFFERENTIAL DIAGNOSIS

Splenomegaly and Lymphadenopathy

- Long differential, including neoplastic, inflammatory, infectious, infiltrative, and hematologic diseases
  - Always consider leukemia and lymphoma

PATHOLOGY

General Features

- Etiology
  - Infection with EBV (a type of herpesvirus)
    - Replicates mainly in B lymphocytes but also in epithelial cells of pharynx and parotid duct
    - Spread by saliva ("kissing disease" among adolescents)
  - Splenomegaly and lymphadenopathy
    - Due to congestion with activated T lymphocytes

CLINICAL ISSUES

Presentation

- Most common signs/symptoms
  - Fever, pharyngitis, adenopathy, malaise, palpable lymphadenopathy (often cervical)
  - Rare
    - Abdominal pain or falling hematocrit with splenic rupture, neurologic syndromes (e.g., Guillain-Barre, meningitis, or transverse myelitis)
  - Complications
    - Splenic rupture (often associated with sports injury)
      - Typically occurs in 1st through 4th week of disease
      - Most common cause of death in mononucleosis
    - Hepatomegaly/jaundice with severe EBV hepatitis
- Other signs/symptoms
  - Lab findings: Lymphocytosis (± atypical lymphocytes); positive "monospot" test (rapid latex agglutination)

Demographics

- Age
  - Adolescents and young adults
    - EBV infection in children is often asymptomatic
    - Symptomatic infection is much more common in adolescents
    - Uncommon in adults due to prior exposure/immunity

Natural History & Prognosis

- Acute symptoms (e.g., sore throat, fever, headache) typically resolve in 1 month
- Fatigue/myalgias may persist for several months
- Rare associations
  - Acute interstitial nephritis, hemolytic anemia, myocarditis/conduction abnormalities, thrombocytopenia, upper airway obstruction

Treatment

- Supportive care
  - Adequate hydration, analgesics, etc.
- Corticosteroids, acyclovir (Zovirax), and antihistamines
  - Not recommended for routine treatment
  - Corticosteroids helpful for respiratory compromise
  - No definitive benefit for antivirals (such as acyclovir)
- Avoid contact sports for minimum of 3-4 weeks due to risk of splenic rupture

DIAGNOSTIC CHECKLIST

Consider

- Consider mononucleosis in previously healthy adolescent or young adult with splenomegaly and lymphadenopathy
- Imaging findings may mimic lymphoma or leukemia, but diagnosis usually confirmed easily by monospot test

Image Interpretation Pearls

- Imaging mostly to evaluate complications, not for diagnosis

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1. Hedgire SS et al: Mono-belly and beyond: spectrum of imaging manifestations of EBV infection in the abdomen. Clin Imaging. 37(4):711-7, 2013

KEY FACTS

TERMINOLOGY

- Recessively inherited disorder of epithelial chloride transport caused by mutation of *CFTR* gene
- Cystic fibrosis (CF) increasingly seen to affect GI tract due to improving life expectancy

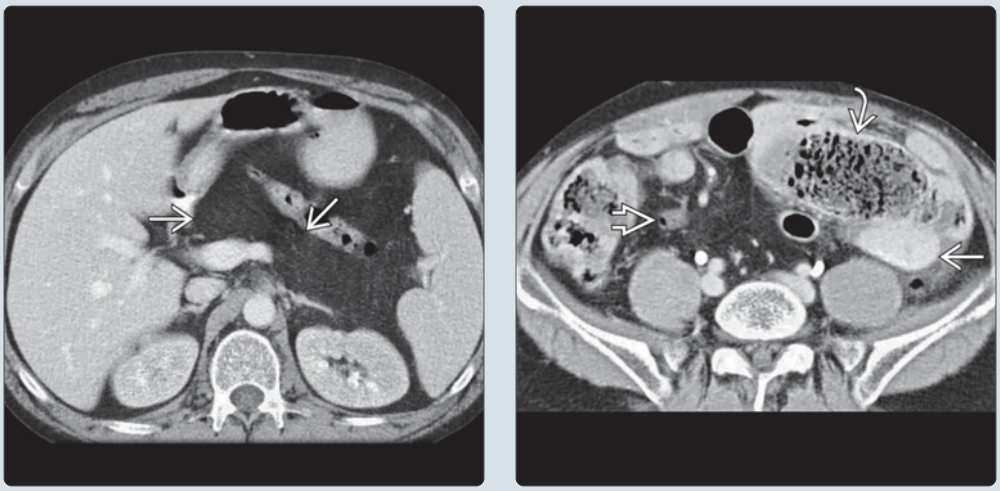
IMAGING

- Most common sites of involvement are lungs, pancreas, bowel, liver, and exocrine glands
- Pancreas
  - Complete fatty infiltration and replacement of parenchyma (often by end of teenage years)
  - Pancreatic cysts: Usually small (< 3 mm), but can be larger and can completely replace pancreas (cystosis)
  - Repeated episodes of acute pancreatitis with development of chronic pancreatitis
- Liver
  - 30-50% develop hepatic steatosis ± hepatomegaly
  - Can develop multinodular cirrhosis in severe cases
- Biliary
  - Biliary abnormalities similar to primary sclerosing cholangitis (PSC)
- Bowel
  - Inspissated fecal material resulting in proximal obstruction, most often in infants (meconium ileus)
  - Obstruction can also occur in adults: Distal intestinal obstruction syndrome (DIOS)
  - Increased risk for intussusception
  - Chronically distended appendix may be difficult to distinguish from acute appendicitis

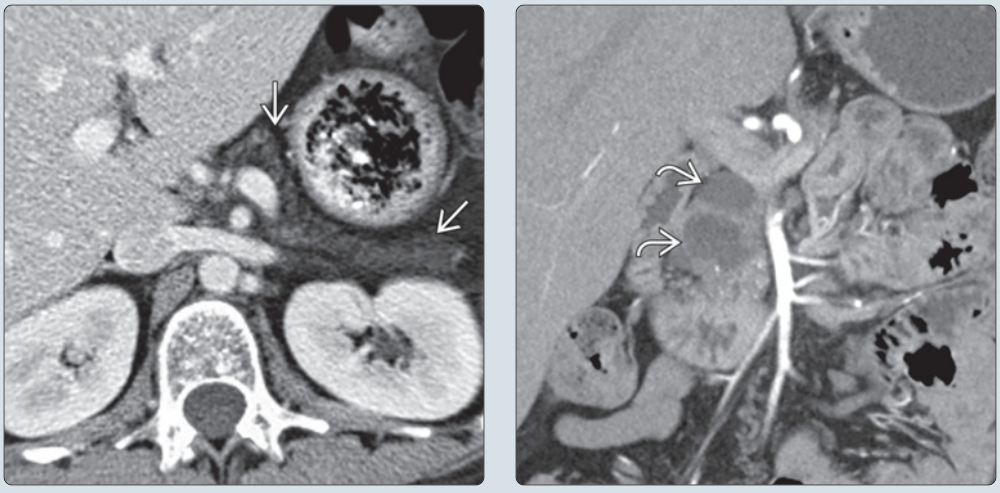
CLINICAL ISSUES

- Overall prognosis for CF has dramatically improved, with average life expectancy now 35-40 years
- Respiratory failure most common cause of mortality, with liver disease 2nd leading cause of death
- Pancreatic insufficiency most common (~ 85%) GI manifestation of CF

(Left) *Axial CECT shows the classic lipomatous replacement and pseudohypertrophy of the pancreas in a young adult patient with cystic fibrosis (CF).* **(Right)** *Axial CECT in the same patient shows dilated proximal small bowel and collapsed distal small bowel. Just proximal to the point of transition is the classic "small bowel feces sign" associated with mechanical small-bowel obstruction, with the obstruction caused by inspissated enteric contents (distal intestinal obstruction syndrome or DIOS).*



(Left) *Axial CECT demonstrates heterogeneous lipomatous replacement of the pancreatic parenchyma but less pseudohypertrophy. This 29-year-old woman had longstanding pancreatic exocrine dysfunction due to CF.* **(Right)** *Coronal CECT demonstrates simple-appearing cysts in the pancreatic head in a young patient with CF. While pancreatic cysts are often very small in CF patients, they can rarely be larger, as in this case.*



TERMINOLOGY

Abbreviations

- Cystic fibrosis (CF)

Definitions

- Recessively inherited disorder of epithelial chloride transport caused by mutation of CF transmembrane conductance regulator (*CFTR*) gene
  - Pulmonary manifestations are primary cause of morbidity and mortality
  - CF increasingly seen to affect GI tract due to improved life expectancy

IMAGING

General Features

- Best diagnostic clue
  - Diffuse fatty replacement of pancreas is most common abdominal imaging finding
- Location
  - Most common sites of involvement are lungs, pancreas, bowel, liver, and exocrine glands

CT Findings

- Pancreatic manifestations
  - Early childhood: Heterogeneous attenuation of pancreas
  - Later childhood: Complete fatty replacement of parenchyma (often by teenage years)
  - Pancreatic cysts: Usually simple in appearance
    - Related to inspissated secretions which lead to ductal obstruction
    - Cysts are usually small (< 3 mm), but can be larger
    - Cysts may diffusely replace pancreas (cystosis)
    - Usually occur in patients in their 20s, and may be symptomatic as result of cyst hemorrhage
  - Repeated episodes of acute pancreatitis with development of chronic pancreatitis
    - Pancreatic ductal strictures, dilatation, beading, etc.
  - May have scattered calcifications (< 10% of patients)
  - Increased risk of pancreatic cancer (very rare)

MR Findings

- Enlarged pancreas with diffuse fatty infiltration appears hyperintense on T1WI
- Pancreas can also appear abnormally hypointense on T1WI due to chronic pancreatitis and fibrosis
- Pancreatic cysts well demonstrated on T2WI and MRCP (hyperintense)

Ultrasonographic Findings

- Pancreas may be barely identifiable as result of diffuse fibrofatty replacement

Other Abdominal Findings

- Liver
  - 30-50% develop hepatic steatosis ± hepatomegaly
    - Can be visualized with US, CT, or chemical shift MR
    - Fatty liver usually asymptomatic and does not correlate with development of CF-related cirrhosis
  - Cirrhosis in 5-15% of all CF patients with portal hypertension in 1-8%
- Biliary
  - Gallstones in up to 1/4 of patients with CF, often with nonspecific gallbladder wall thickening and sludge
  - Microgallbladder in 30% of CF patients at autopsy
    - Probably due to chronic stenosis or atresia of cystic duct (does not usually cause symptoms)
  - May develop bile duct abnormalities very similar to primary sclerosing cholangitis (PSC), including strictures, bile duct wall thickening, biliary obstruction, etc.
    - May develop focal biliary fibrosis (> 3/4 of patients with CF older than 24 years)
      - Periductal thickening and fibrosis due to thick secretions within duct and reactive inflammation
      - Focal thickening of bile duct with changes in adjacent liver parenchymal density/signal
      - US: Hyperechoic periportal thickening and diffuse hepatic hyperechogenicity
- Esophagus
  - Gastroesophageal (GE) reflux seen in 27% of patients younger than 5 years and increases with age
- Bowel
  - ↑ frequency of peptic ulcer disease (gastric or duodenal)
  - Obstruction
    - Bubbly fecal mass within bowel causing proximal obstruction, most often in infants (meconium ileus)
    - Obstruction can also occur in adults: Distal intestinal obstruction syndrome (DIOS) (previously known as meconium ileus equivalent)
      - Obstruction results from thickened intestinal secretions, poor motility, and formation of mass-like fecal material which obstructs bowel
      - Usually obstructs at distal ileum or right colon (rarely at distal rectum)
      - Water-soluble contrast enema might be able to reduce obstruction
  - Increased risk for intussusception
    - More common in older patients
    - Most often ileocolic intussusception, usually due to inspissated fecal material acting as lead point
    - US: "Doughnut" or pseudokidney appearance
    - CT: "Target" sign of edematous bowel and intermixed mesenteric fat
    - Water-soluble contrast enema can be used for reduction, but high rates of recurrence
  - Chronically distended appendix common with internal inspissated high-density material
    - Differentiating acute appendicitis from chronic distention is difficult with imaging
  - Abnormalities of colon can include proximal colonic wall thickening, proliferation of fat surrounding the colon, and pericolonic fat stranding
    - Benign pneumatosis due to air dissecting below diaphragm into bowel wall due to lung disease
    - Increased susceptibility to pseudomembranous colitis
    - Increased risk of colon cancer
    - Rectal mucosal prolapse usually in younger patients
- Renal
  - Nephrolithiasis in 3-6% CF patients
  - Interstitial nephritis due to antibiotics and amyloidosis will likely become more common as patients live longer

DIFFERENTIAL DIAGNOSIS

Pancreatic Lipomatous Pseudohypertrophy

- Diffuse enlargement and fatty replacement of pancreas
- Associated with chronic liver disease; no signs of CF

Shwachman-Diamond Syndrome

- Pancreatic lipomatosis associated with short stature
- Appearance of pancreas is identical to CF

Normal Fatty Lobulation

- Fatty replacement of pancreatic parenchyma with mild glandular atrophy
- Common in elderly, obese, and diabetic patients

Chronic Pancreatitis

- Pancreatic atrophy, parenchymal and ductal calcifications, and pancreatic ductal beading, irregularity, and dilatation

Pancreatic Cystic Neoplasms (IPMN, etc.)

- May be indistinguishable from CF-related pancreatic cysts without clinical history

PATHOLOGY

General Features

- Etiology
  - Mutations of *CFTR* gene lead to multisystem pathology
  - Disruption of chloride ion, bicarbonate, and water transport in duct cells
  - Primary ductal cell chloride channel abnormality results in dehydrated protein-rich secretions obstructing proximal ducts
    - Leads to acinar cell destruction, fibrosis, and exocrine insufficiency
    - Also causes ductal obstruction leading to lung infections, biliary obstruction, etc.
- Genetics
  - Mutations in *CFTR* gene
  - Autosomal recessive gene on chromosome 7

Lab

- Positive sweat test
- Genotyping has proved useful in identifying gene carriers; antenatal diagnosis and treatment

CLINICAL ISSUES

Presentation

- Clinical profile
  - Usually diagnosed in infants with meconium ileus, lung infections, etc.
    - Until recently, patients died in childhood of pulmonary infections and respiratory failure
    - Longer survival now reveals other multisystem manifestations of cystic fibrosis
  - 7% of CF patients do not present until adulthood
    - Of these, 26% present with GI symptoms and 4% with pancreatitis
  - Symptoms of pancreatic dysfunction
    - Steatorrhea, malabsorption, fat intolerance
      - Pancreatic insufficiency usually present from birth
    - Repetitive acute pancreatitis leading to chronic pancreatitis
      - Pancreatitis in 10% of CF patients
    - Endocrine (diabetes) and exocrine dysfunction
  - Symptoms of hepatobiliary dysfunction
    - Inspissated secretions obstruct bile ducts and caustic bile acts on adjacent parenchyma
    - Liver disease can vary from asymptomatic elevation in liver function tests to end-stage liver disease and cirrhosis
    - 10-20% of patients develop chronic liver disease (including cirrhosis and portal hypertension)
    - Cholelithiasis in 10% of patients
  - Intestinal manifestations
    - Intestinal obstruction
      - Usually in infancy (meconium ileus), but may occur in adults as well (DIOS or meconium ileus equivalent)
    - Intussusceptions (1% of patients)
  - Infertility in males (azoospermia, turbid semen)

Demographics

- Age
  - Children most affected
  - Patients reaching adulthood represent rapidly growing percentage of CF population
- Ethnicity
  - More common in Caucasians
    - Most common inherited fatal disease in Caucasians
- Epidemiology
  - CF prevalence: 1 in 3,500
  - 85–90% of CF patients have pancreatic exocrine gland insufficiency
    - Cystic fibrosis is major cause of pancreatic exocrine failure in childhood
  - 30-50% of CF patients have pancreatic endocrine gland dysfunction

Natural History & Prognosis

- Overall prognosis for CF has dramatically improved due to screening and better treatment, with average life expectancy now 35-40 years
- Respiratory failure most common cause of mortality
  - Liver disease is 2nd leading cause of death
- Pancreatic insufficiency most common (~ 85%) GI manifestation of CF
  - Varies depending on degree of ductal obstruction by mucus, which leads to exocrine gland atrophy, progressive fibrosis, and cyst formation
  - CF may predispose to pancreatic cancer

Treatment

- Aggressive nutritional and pancreatic enzyme therapy
- DIOS usually treatable with hydration, laxatives, or water soluble contrast enema although severe cases may require surgery

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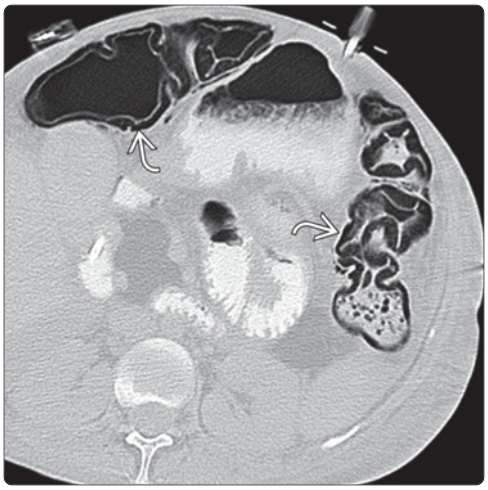
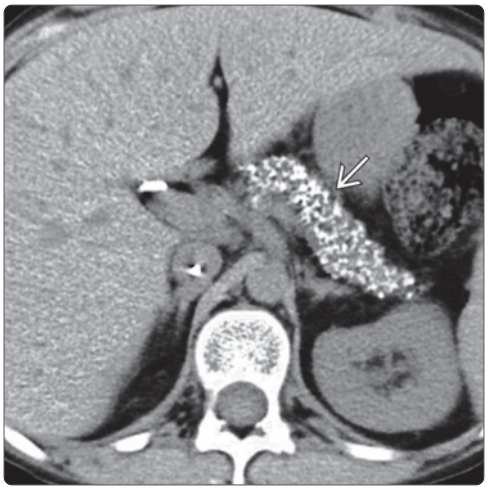
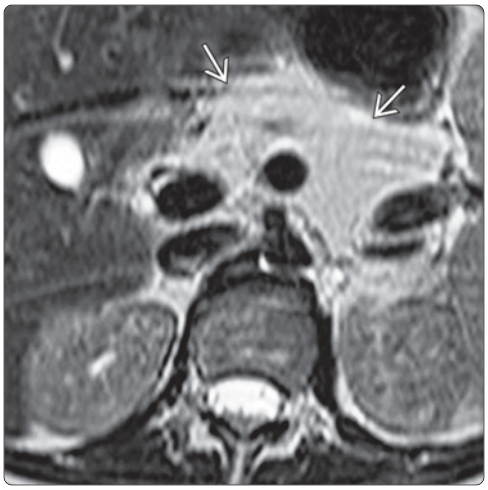
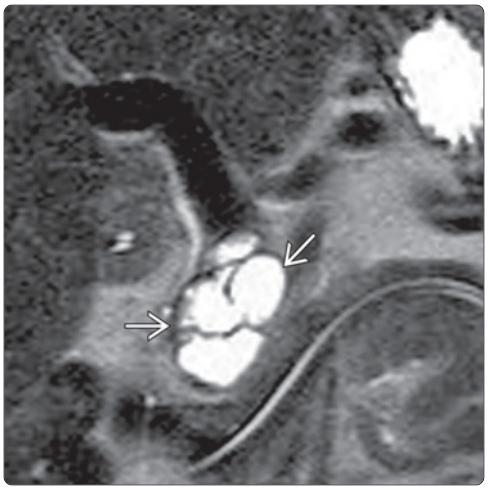
1. Keyzer C et al: Cystic fibrosis: unenhanced CT description of the appendix in asymptomatic adults. AJR Am J Roentgenol. 202(4):759-64, 2014



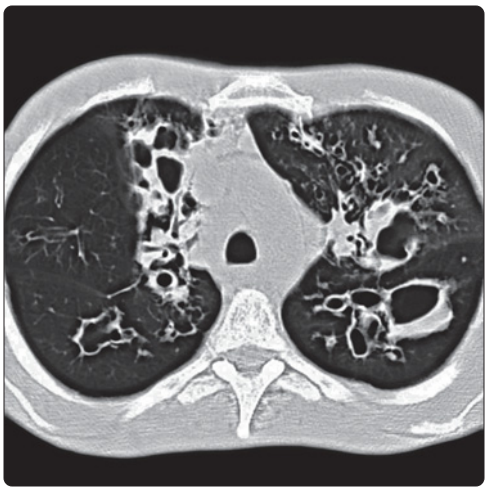


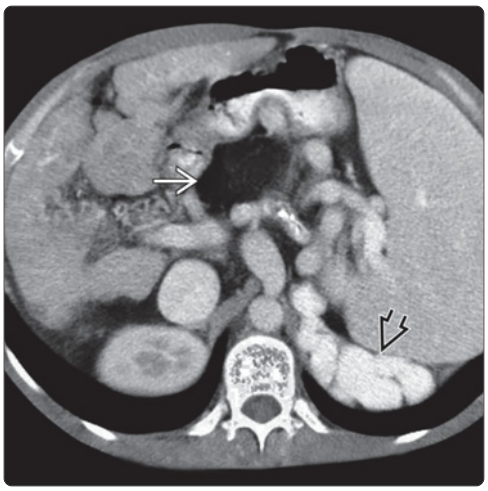
(Left) *Axial CT image shows diffuse pneumatosis ➡ of the transverse and left colon. The patient was completely asymptomatic, and this was felt to be benign pneumatosis due to gas dissecting from the chest into the bowel wall as a result of the patient's lung disease.* **(Right)** *Axial NECT in a 24-year-old woman with CF shows an unusually severe degree of pancreatic calcification ➡. Scattered, small calcifications are a more common finding in this disease.*





(Left) *Axial T2WI in a 16-year-old girl with CF demonstrates complete fatty replacement ➡ of the body and tail of the pancreas. Note that the pancreas consequently shows high T1WI signal.* **(Right)** *Coronal MRCP in the same patient shows a septate cystic mass in the pancreatic head ➡ that mimics a cystic neoplasm. However, pancreatic cysts of variable size are commonly encountered in patients with CF.*





(Left) *Axial CECT in a 31-year-old man shows classic cystic bronchiectasis in the lungs. Many patients are being kept alive longer with better medical care and even lung transplantation, resulting in an increased prevalence of extrapulmonary manifestations of CF.* **(Right)** *Axial CECT in the same patient shows that the liver is small and cirrhotic with obvious signs of portal hypertension, including splenomegaly and large varices ⇨. The pancreas shows fatty replacement ➡.*

Sickle Cell Anemia

KEY FACTS

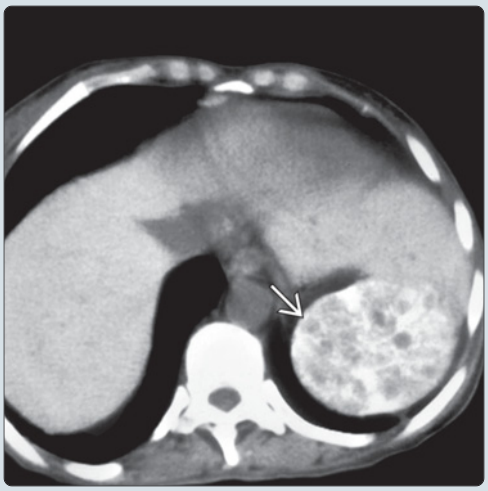
TERMINOLOGY

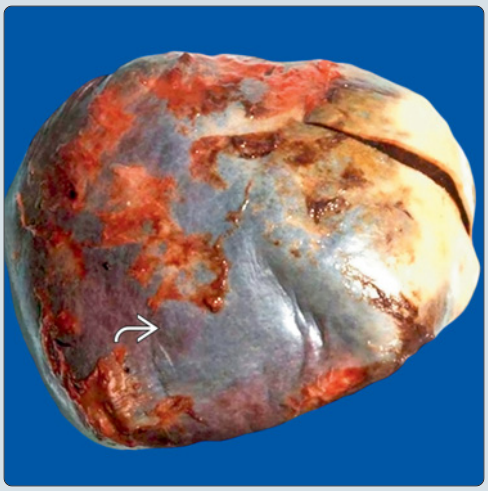
- Inherited hemolytic anemia arising due to abnormal hemoglobin, resulting in deformation of red blood cells and leading to microvascular occlusions and infarcts

IMAGING

- Spleen
  - Splenic autoinfarction: Absent or small calcified spleen
  - Massive splenic infarction: Rare complication defined as when > 50% of spleen is infarcted
  - Splenic sequestration: Massive splenomegaly
  - Splenic abscess: Usually due to prior infarcts
- Gallbladder: Gallstones in young patients
- Extramedullary hematopoiesis: Most commonly paravertebral soft tissue masses of homogeneous density
- Kidneys
  - Papillary necrosis on CT urography
  - Large kidneys in early phase of disease; gradual atrophy with development of chronic renal failure
- Bones
  - Widened medullary spaces, thinning of cortex, coarsening of trabecular pattern, and osteopenia
  - Osteonecrosis and multiple bone infarcts
    - High T2WI MR signal in affected areas with serpiginous low T2 signal outline (double-line sign)
  - H-shaped or "Lincoln log" vertebrae
    - Cortical thinning leads to endplate deformities
  - Osteomyelitis
  - Persistence of red (cellular) marrow in bones with low-signal marrow on T1WI
    - Demand for increased production of RBCs prevents normal conversion of red to yellow marrow
- Findings of iron deposition due to repetitive transfusions
  - Hyperdense liver on NECT
  - Liver, spleen, and bone marrow abnormally low signal on all MR pulse sequences (particularly on T2WI)
  - Involved organs demonstrate characteristic signal loss on in-phase GRE images

(Left) *Axial NECT in a patient who presented with sickle cell anemia and severe left upper quadrant pain demonstrates a heavily calcified and heterogeneous spleen, indicating chronic and possibly acute infarction.* **(Right)** *Splenectomy specimen from the same patient illustrates a mottled spleen with capsular discoloration that was heavily calcified on microscopy.*





(Left) *Axial NECT in a patient with homozygous sickle cell anemia demonstrates a small and heavily calcified spleen, also known as autosplenectomy.* **(Right)** *Axial CECT in a patient with sickle cell anemia demonstrates multiple wedge-shaped hypodense areas in the spleen, representing massive splenic infarction, an uncommon complication that rarely results in the formation of a splenic abscess.*