The USMLE Step 2 CK BIBLE

The Ultimate Step 2 CK Preparation Guide

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The USMLE Step 2 CK BIBLE

2nd Edition

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CONTENTS

FOREWORD

The USMLE Step 2 CK BIBLE is the culmination of over four months of my own intense personal Step 2 CK preparation. This document contains and all of the notes I made, all of the charts, graphs, and images I put together to create the ultimate study guide, and I guarantee it is more than enough to help you pass, and if used properly can help you achieve a top score on the Clinical Knowledge exam.

I used five different study guides as well as all of the notes I took from working in the wards to put together this in-depth study guide. This preparation guide contains the most up-to-date as well as the most commonly asked clinical information, which will help you score high on the Step 2 CK exam.

When I put this preparation guide together, I did so with my own score in mind, and I made it so that I would have to study from one source, and that's exactly what you have here. If you study hard and use the CK BIBLE, you will not only pass, you will do very well.

Best of luck on the Step 2 CK exam...

Chapter 1

Surgery

TRAUMA

Trauma patients are managed using the ABCDE's in the primary survey after a traumatic incident.

A – Airway

- Ensure patient is immobilized and maintain airway with jaw thrust
- If airway cannot be established, insert 2 large bore needles into the cricothyroid membrane
- Never perform tracheotomy in the field
- If patient is unconscious or you cannot establish an airway otherwise, intubate the patient.

B - Breathing

- Look for chest movement
- Listen for breathing sounds
- Observe the respiratory rate
- Look for life-threatening injuries (tension pneumothorax, flail chest, open pneumothorax)

C - Circulation

- Placement of 2 large-bore IV's in the upper extremities
- If patient is in shock, place a central line in the patient
- Keep blood on stand-by in case of hemorrhage

D - Disability

- Assess the neurological status with the Glasgow coma scale
- Check all lab tests (blood, ETOH, electrolytes)
- Loss of consciousness

A loss of consciousness can be assessed with the mnemonic AEIOU TIPS

Alcohol, **E**pilepsy, **I**nsulin, **O**verdose, **U**remia, **T**rauma, **I**nfection, **P**sychogenic, **S**troke

E – Exposure

- Examine the skin (must remove all clothes)

In the secondary survey, perform the following:

- Check the Glasgow coma scale
- Check all orifices for trauma and/or injuries (bleeding)
- Perform checks using ultrasound, XRAY, CT
- Check for compartment syndrome

GLASGOW COMA SCALE

STATUS/FINDING	POINTS
Eye Opening	
Spontaneous	4
To Voice	3
To Stimulation	2
No Response	1
Verbal Response	
Oriented	5
Confused	4
Incoherent	3
Incomprehensible	2
No Response	1
Motor Response	
To Command	6
Localizes	5
Withdraws	4
Abnormal Flexion	3
Extension	2
No Response	1

A coma scale below 8 indicates severe neurologic injury

SHOCK

The type of shock can be diagnosed by checking the cardiac output (CO), the pulmonary capillary wedge pressure (PCWP), and the peripheral vascular resistance (PVR).

Differential Diagnosis of Shock

	HYPOVOLEMIC	CARDIOGENIC	SEPTIC
CO	⇓	⇓	⇑
PCWP	⇓	⇑	⇓
PVR	⇑	⇑	⇓

How to correct the different types of shock

	Problem	**Initial Treatment**
Hypovolemic Shock	**Decreased Preload**	**2 Large bore IV's, replace fluids**
Cardiogenic Shock	**Cardiac Failure**	**02, dopamine and/or NE**
Septic Shock	**Decreased PVR**	**02, NE, IV antibiotics**

Recognizing Shock

In Chest trauma:

- The most common type of shock resulting from chest trauma is ***hypovolemic***
- Patient will be pale, cold, and diaphoretic
- This patient is likely losing large amounts of blood, thus searching for source of bleeding is imperative
- Pericardial tamponade can be a result of thoracic trauma, look for distended neck veins
- In suspected pericardial tamponade, look for an enlarged heart on CXR, perform cardiocentesis, look for electrical alternans on EKG

***Management of shock*:**

- Control the site of bleeding
- Give fluids
- Prepare for an emergency laparotomy

**** If a laparotomy isn't warranted, simply resuscitate with fluids**

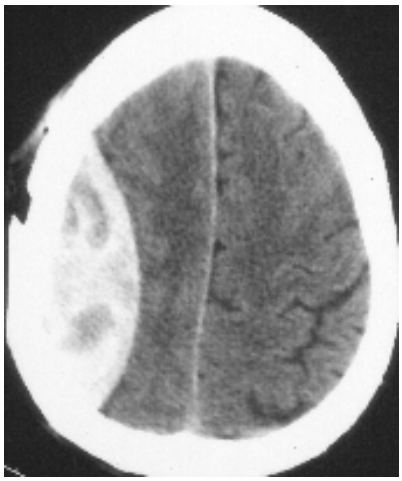
HEAD TRAUMA

Epidural Hematoma

- There will be a history of trauma
- Sudden loss of consciousness followed by a lucid interval, then followed by rapid deterioration
- Most commonly bleed is from the middle meningeal artery

Diagnosis:

- With a CT, looking for a lens-shaped hematoma



Management:

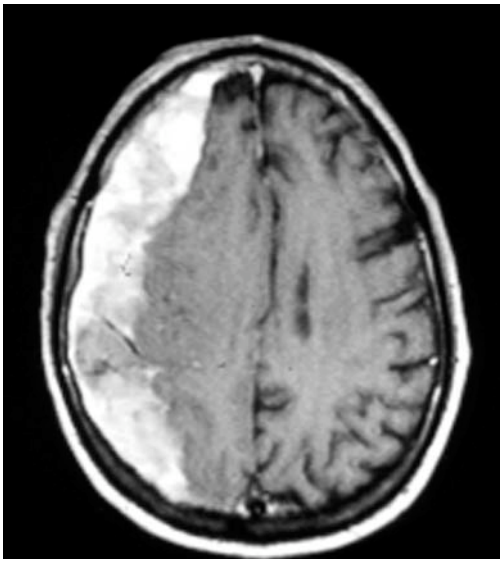
- Emergency craniotomy essential because this is a deadly case within a few hours

Subdural Hematoma

- Is a low-pressure bleed coming from the bridging veins
- There is usually a history of head trauma with fluctuating consciousness

Diagnosis:

- CT showing crescent-shaped bleed



Management:

- If there is midline displacement and signs of mass-effect then do an emergency craniotomy
- If symptoms are less severe, conservative management includes steroids

Diffuse Axonal Injury

- This type of injury occurs after an acceleration-deceleration injury to the head
- Patient is usually unconscious
- There is a terrible prognosis associated with this injury

Management:

- Lower ICP and prevent further injury

Basal Skull Fracture

- This presents with ecchymosis around eyes, behind the ears, or with CSF leak from the nose

Diagnosis:

- CT scan of head and neck

Management:

- CSF rhinorrhea will stop on its own
- If facial palsy is present, give steroids

BURNS

1st degree and Second-degree burns:

- Epidermis and superficial dermis
- Skin is painful, red, and blistered
- Treatment with ointments and/or pain relievers

Third and Fourth degree burns:

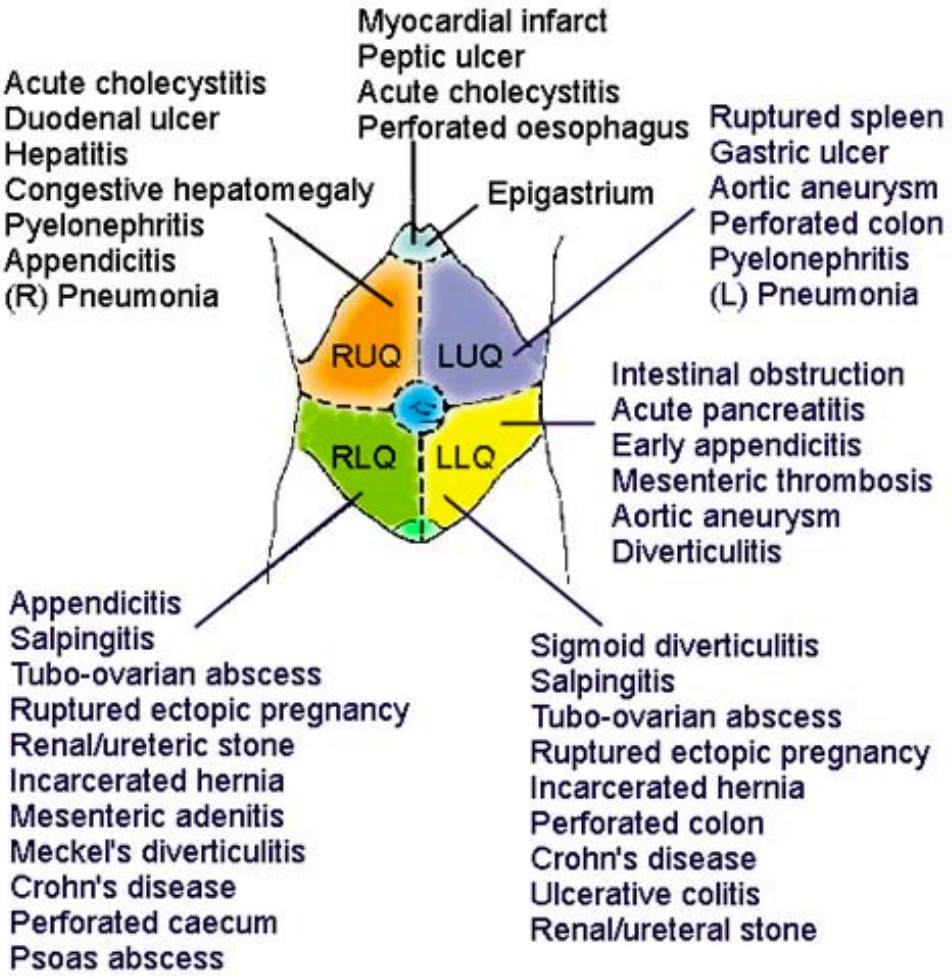
- Affects all layers + subcutaneous tissues
- Painless, dry, charred, and cracked skin
- Burns affecting all layers of the skin require surgical intervention

Treatment:

- Depending on severity, resuscitate with fluids
- Removal of eschars
- Do a CXR to rule out lung injuries
- Topical antibiotics after eschar removal
- Burns that cover more than 20% of the body require admission to a burn center

SURGICAL ABDOMEN

The following illustrates the differential diagnosis for abdominal pain in the right upper quadrant, right lower quadrant, left upper quadrant, left lower quadrant.



Right Upper Quadrant Conditions - Differentiating

Hepatitis

- Presents with RUQ pain and/or tenderness
- Jaundice is most likely present
- Fever is present
- Perform an ***ultrasound*** to rule out other causes of pain

Cholecystitis

- RUQ pain and tenderness
- (+) Murphy's sign (inspiratory arrest during palpation)
- Perform an ultrasound to detect gallstones, a thickened gallbladder wall, or pericholecystic fluid

Choledocholithiasis

- RUQ pain that is worsened with the ingestion of fatty foods
- Jaundice is often present
- Perform an ultrasound to detect dilatation of the common bile duct

Biliary Cholic

- Constant epigastric and RUQ pain
- Perform an ultrasound to detect the presence of gallstones without any other gallbladder-related findings

Cholangitis

- A life-threatening condition
- Presence of Charcot's triad: Fever + Jaundice + RUQ pain
- If there is also hypotension and mental status changes, this qualifies as Reynold's pentad
- Perform and ultrasound and a CT to detect biliary duct dilatation due to gallstone obstruction
- Confirm diagnosis with ERCP

Pneumonia

- Presence of pleuritic chest pain
- Perform a CXR, which will show pulmonary infiltrates

Fitz-Hugh-Curtis Syndrome

- RUQ pain, fever
- There is going to be a history of salpingitis
- Caused by ascending Chlamydia or gonorrhea-related salpingitis
- Perform an ultrasound which will show a normal gallbladder and biliary tree with fluid around the liver and gallbladder

Right Lower Quadrant Conditions – Differentiating

Appendicitis

- Diffuse abdominal pain that localizes to the RLQ at McBurney's point (2/3 distance from umbilicus to ASIS)
- Fever and diarrhea often present
- Abdominal xray or CT to solidify diagnosis
- Decision to remove is based on clinical presentation

Ectopic Pregnancy

- Presents with constant lower abdominal pain, crampy in nature
- Vaginal bleeding
- Tender adnexal mass
- Labs will show ⇑ hCG

Salpingitis

- Lower abdominal pain
- Purulent vaginal discharge
- Cervical motion tenderness
- Perform an ultrasound to detect the abscess, and a CT to rule out other conditions

Meckel's Diverticulitis

- Follows the 1-10-100 rule
- 1%-2% prevalence
- 1-10cm in length
- 50-100 cm proximal to ileocecal valve
- Presents with GI bleed, small bowel obstruction (SBO)
- Technetium pertechnetate scan to detect

Yersinia Enterocolitis

- Presents similarly to appendicitis (fever, diarrhea, severe RLQ pain)
- XRAY will be negative
- Treat with aggressive antibiotic therapy

Ovarian Torsion

- Patient develops an acute onset of severe, unilateral pain
- Pain changes with movement
- Presence of a tender adnexal mass
- Ultrasound is done first
- Confirm with a laparoscopy

Pyelonephritis

- Classically presents with CVA tenderness, high fever, and shaking chills
- Best initial diagnostic test is a UA and Urine culture

Intussusception

- Seen most commonly in infants between 5 and 10 months of age
- Presence of currant jelly stool (mix of blood and mucus)
- Vomiting, intense crying
- Infants will often pull legs into the abdomen to relieve some pain
- Barium enema is used for both diagnosis and treatment

Left Upper Quadrant Conditions – Differentiating

Myocardial Infarction

- Crushing chest pain that radiates to the jaw, neck, left arm
- Nausea, diaphoresis is present
- Diagnosed by EKG, cardiac enzymes (CKMB, trop I)

Peptic Ulcer

- Presents as epigastric pain that is relieved by foods and/or antacids
- Perforations presents with acute and severe epigastric pain, may radiate to shoulders (Phrenic nerve involvement)
- Diagnose with an upper GI endoscopy

Ruptured Spleen

- Usually a history of trauma
- Presence of Kehr's sign (LUQ pain that radiates to the left shoulder)
- Diagnose with an abdominal CT

Left Lower Quadrant Conditions – Differentiating

- Similar to the RLQ conditions are: Ovarian torsion, Ectopic pregnancy, and Salpingitis

Diverticulitis

- Patient has LLQ pain, fever, and urinary urgency
- Diagnose with a CT scan, which shows thickening of the large intestine wall

Sigmoid Volvulus

- Most commonly seen in an older patient
- Presents with constipation, distended abdomen, and abdominal pain

- Contrast enema to diagnose, will see the classic "bird's beak"

Pyelonephritis

Classically presents with CVA tenderness, high fever, and shaking chills

Differential Diagnoses for Midline Conditions

GERD

- Epigastric/substernal burning pain
- Degree of pain changes with different positions (worse when patient is supine)
- Diagnosis made with either a barium swallow, pH testing, or upper GI endoscopy

Abdominal Aortic Aneurysm

- Asymptomatic usually until it ruptures
- If rupture occurs, patient experiences abdominal pain + shock
- There is usually a palpable pulsatile periumbilical mass
- Ultrasound done first (least invasive), but can visualize with an xray or CT of the abdomen

Pancreatitis

- Epigastric pain that radiates to the back
- Nausea and vomiting are usually present
- Patient often has a history of alcoholism

Pancreatic Pseudocyst

- Is a result of pancreatitis
- Consider this if patient had pancreatitis that recurred and/or did not resolve
- Ultrasound will show a pseudocyst

Surgical Conditions of the Esophagus

Achalasia

- A condition where the lower esophageal sphincter fails to relax

Signs and Symptoms:

- Dysphagia to BOTH solid and liquid
- Regurgitation of food

Diagnosis:

- Best initial test its the Barium Swallow, which demonstrates narrowing of the distal esophagus
- Most accurate test is esophageal manometry, which will demonstrate the lack of peristalsis

Treatment:

- The best initial therapy is pneumatic dilation
- If pneumatic dilation is not successful, surgery should be performed
- If patient does not want surgery, can attempt to relax the LES with injection of botulinum toxin

Esophageal Diverticula (Zenker's diverticulum)

- Most common presentation is a patient with dyphagia that is accompanied by terrible breath
- Pathology is related to the posterior pharyngeal constrictor muscles with dilate, causing the diverticulum

Signs and Symptoms:

- Dysphagia
- Halitosis

Diagnosis:

- The best initial test is the barium swallow

Treatment:

- Surgical resection of the diverticula is the best initial treatment option

Cancer of the Esophagus

There are Squamous Cell Carcinoma and Adenocarcinoma

Common symptoms to both:

- Dysphagia to solids 1st, then to liquids 2nd
- Weight loss
- Heme (+) stool
- Anemia
- Hoarseness

Squamous Cell Carcinoma

- Is the 2nd MCC of esophageal cancer
- Related to chronic use of alcohol and tobacco
- Most commonly seen in the 6th decade of life and later

Adenocarcinoma

- Occurs in patients who have chronic GERD
- Chronic GERD leads to Barrett's esophagus, which then leads to Adenocarcinoma

Diagnosing:

- The best initial diagnostic test is an endoscopy

Treatment:

- The best initial treatment is surgical resection as long as there is no metastasis
- Surgery should be followed with 5-FU

Diffuse Esophageal Spasms

- Patient presents with severe chest pain
- Often times, they don't fit the criteria for an MI, but should get the cardiac enzymes and do EKG to rule out an MI
- Often comes after having a cold drink

Diagnosis:

- Manometry is the most accurate diagnostic test

Treatment:

- Calcium channel blockers and nitrates are the treatment option of choice

Mallory Weiss Tear

- Violent retching and/or vomiting causes sudden bleeding
- Most cases resolve spontaneously, if they don't though give epinephrine to constrict the blood vessels and stop the bleeding

Cancer of the Stomach

- Most cancers of the stomach are found to be malignant
- There is a link of stomach cancers to blood group A, which may indicate a genetic predisposition to the condition
- Linitis plastica is a diffuse cancer that is fatal within months, and is the most deadly form of gastric cancer

Signs and Symptoms for all:

- GI discomfort and/or pain
- Weight loss
- Anemia
- Anorexia

There is an ⇑ risk when there is:

- Low fiber consumption
- Excess nitrosamines in the diet (due to smoked meats)
- Excess salt intake in the diet
- Chronic gastritis

There are some classic findings in metastatic gastric cancer, they include:

Virchow's node:

- Left supraclavicular node is hard

Krukenberg Tumor:

- The metastasis of gastric cancer bilaterally to the ovaries
- Ovaries are palpable in this case
- They are signet-ring cells

Sister Mary Joseph sign:

- Hard nodule at the umbilicus due to metastasis
- Indicative of a very poor prognosis

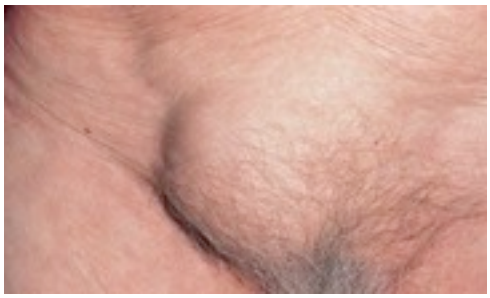
Treatments:

- Surgery + chemotherapy
- Palliative care is often the only choice if too advanced

Hernias

Inguinal Hernias

- Is the most common type of hernia
- Men > women



Direct Inguinal Hernia:

- Protrudes directly through Hasselbach's triangle (inferior epigastric artery, rectus sheath, and inguinal ligament), medial to the inferior epigastric artery

Indirect Inguinal Hernia:

- More common than the direct hernia, passes laterally to the inferior epigastric artery into the spermatic cord

Signs and Symptoms:

- Groin mass (intermittent) that protrudes with valsalva-type maneuvers

Diagnosis:

- Must differentiate from a femoral hernia, which will herniated below the inguinal ligament
- Diagnosis is based on clinical examination

Treatment:

- Surgical repair

Femoral Hernias

- Women > Men
- Have a greater risk of incarceration due to the way they herniated
- Diagnose clinically
- Surgical correction (do not delay due to risk of incarceration and subsequent strangulation)

Visceral Hernias

- This type of hernia causes intestinal obstruction

Signs and Symptoms:

- Abdominal pain
- Obstipation (no flatulence)

Diagnosis:

- XRAY will show air-fluid levels, no gas in rectum
- Differentiate from adhesions

Treatment:

- Surgical repair

Surgical Conditions of the Gallbladder

Gallstones (Cholelithiasis)

Seen mostly in women with the 4 F's

1. Female
2. Fat
3. Forty
4. Fertile

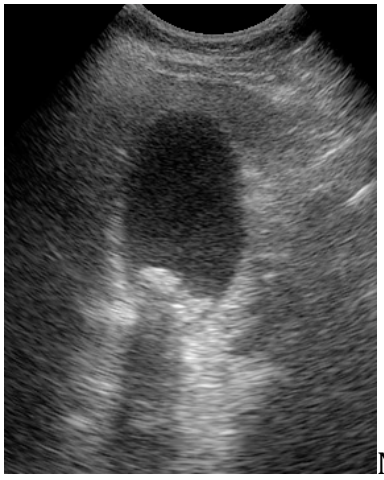
- It isn't the presence of gallstones that warrants intervention, but the possible complications associated with them
- Ultrasound is the test of choice for identifying gallstones

Treatment:

- Asymptomatic gallstones require no intervention
- Chronic pain may require a cholecystectomy
- With an increased risk of cancer, such as in the case of a calcified gallbladder wall, cholecystectomy may be warranted

Cholecystitis

- Is an infection of the gallbladder that is a result of an obstruction
- Common causes are: E. Coli, Enterobacter, Enterococcus, and Klebsiella

 Note the shadow from the impacted stone.

Signs and Symptoms:

- Acute onset of right upper quadrant pain that is non-remitting
- (+) Murphy's sign – arrest of inspiration upon palpation

Diagnosis:

- Ultrasound to detect stones, a thickened wall, or fluid surrounding the GB
- Confirm with HIDA scan
- Labs show WBC's >20,000, ⇑ Bilirubin, ⇑ AST/ALT

Treatment:

- Keep patient NPO, give IV fluids, and give antibiotics to cover gram (-) rods and anaerobes
- Do not give morphine for pain because it causes a spasm of the sphincter of oddi
- If improvements are not seen, cholecystectomy may be warranted

Ascending Cholangitis

- Obstructed bile flow from an obstructed common bile duct leads to an infection
- Presence of Charcot's triad: RUQ, fever, jaundice is commonly seen

Diagnosis:

- Ultrasound to detect dilation
- An ERCP can be used after the preliminary US diagnosis

Treatment:

- NPO
- IV fluids
- Gram (-) antibiotics
- ERCP for decompression of the biliary tree and for removal of the stones

Choledocholithiasis

- An obstruction of the common bile duct

Signs and Symptoms:

- Jaundice (obstructive)
- ⇑ Alkaline phosphatase
- ⇑ Conjugated bilirubin

Diagnosis:

- Ultrasound to detect CBD obstruction

Treatment:

- Cholecystectomy

Cancer of the GB

- Is a rare cancer that is associated with a history of gallstones
- Occurs later in life
- The MC primary tumor of the gallbladder is the adenocarcinoma
- Associated with Clonorchis sinensis infestation
- Has a grave prognosis, with most patients dying within 1yr of diagnosis

Signs and Symptoms:

- Sharp, colicky pain

Diagnosis:

- US or CT to detect the tumor

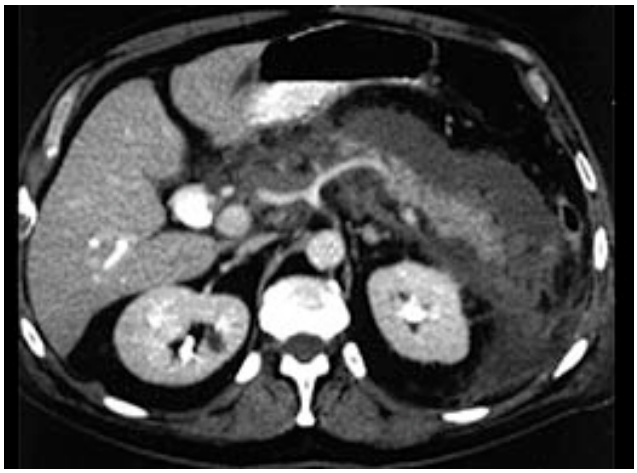
Treatment:

- Placement of bile duct stents
- Surgery as a palliative option, but is not curative

Surgical Conditions of the Pancreas

Pancreatitis

- Autodigestion of the pancreas by it's own enzymes
- MCC is alcohol and gallstones



Signs and Symptoms:

- Severe epigastric pain that radiates to the back
- ⇑ Serum amylase and lipase

Diagnosis:

- Clinical suspicion + abdominal CT
- There may be discoloration of the flank (Grey Turner's sign) and Cullen's sign (bluish discoloration of the periumbilicus)

Treatment:

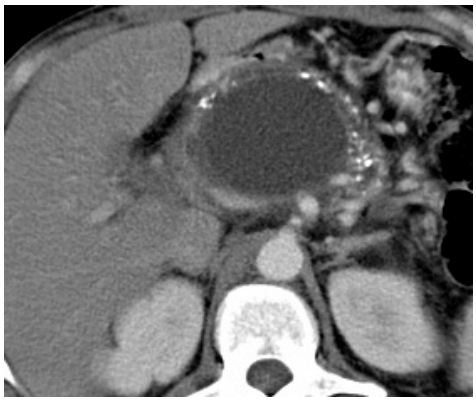
- NPO, IV fluids, and Demerol for pain relief
- Be aware of the potential for alcohol withdrawal

Complication:

- There is a risk for abscesses, renal failure, duodenal obstruction, and pancreatic pseudocysts

Pancreatic Pseudocyst

- Is a complication of chronic pancreatitis
- Results in a fluid collection within the pancreas that is encapsulated by a fibrous capsule



Diagnosis:

- Ultrasound
- Abdominal CT

Treatment:

- Surgical drainage
- Creation of a fistula draining the cyst into the stomach

Complications:

- Infection followed by rupture can cause peritonitis

Endocrine Pancreas

- β-cell hyperplasia causes an insulinoma
- α-cell tumor causes hyperglucagonemia

Cancer of the Pancreas

- More common in African Americans/males/smokers
- May be more common in diabetics
- 90% are adenocarcinomas
- 60% arise from the head of the pancreas

Signs and Symptoms:

- Weight loss
- Painless jaundice

Diagnosis:

- ⇑ bilirubin, ⇑ alkaline phosphatase, ⇑CA19-9
- CT scan

Treatment:

- Although usually a terminal diagnosis, can do a resection of the pancreas, or Whipple's procedure
- The 5yr survival rate is only 5%

Surgical Conditions of the Small Bowel

Small Bowel Obstruction

- SBO can be caused by a number of conditions

Causes:

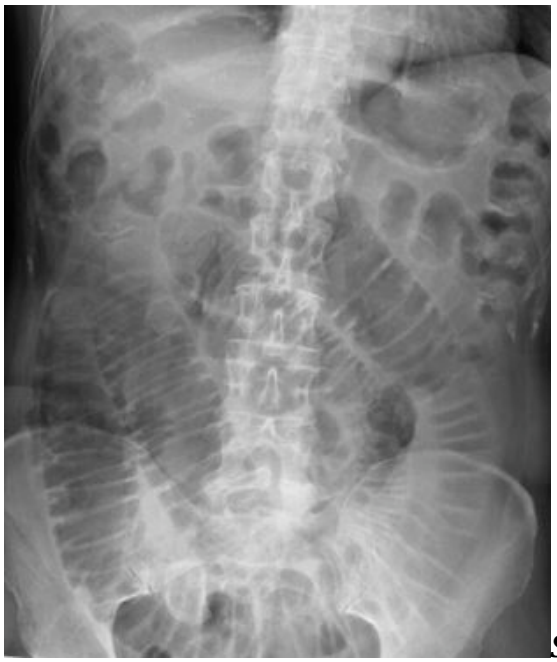
- Peritoneal adhesions
- Hernias
- Crohn's disease
- Meckel's
- Gallstone ileus
- Abdominal inflammation

Signs and Symptoms:

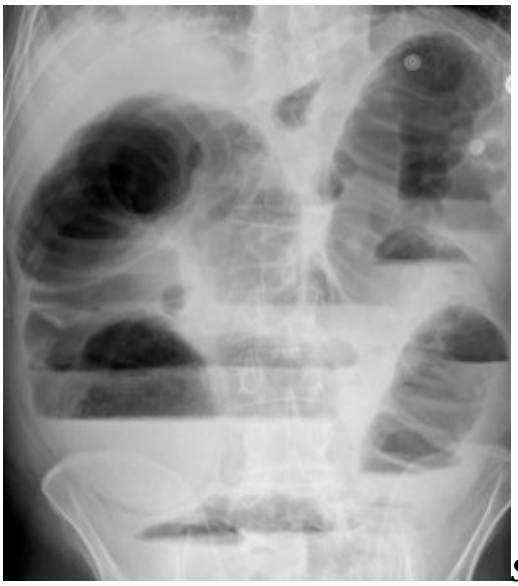
- Nausea/vomiting
- Abdominal pain
- Abdominal cramps
- Tenderness/distention
- Hyperactive and high-pitched bowel sounds

Diagnosis:

- Abdominal xray
- Air-fluid levels on upright film



Small Bowel Obstruction (Supine View)

 ***Small Bowel Obstruction (Upright View)***

Treatment:

- NG tube decompression
- NPO
- IV fluids
- If only partially obstructed may be able to treat without surgery
- If surgery is required, must remove both obstruction and dead bowel

<u>Neoplasm of the Small Bowel</u>

- Most commonly is a leiomyoma, second MC is a carcinoid tumor (benign types)
- Most common malignant types are: adenocarcinoma, carcinoid, lymphoma, and sarcoma
- Biopsy required for diagnosis
- Treatment involves surgical resection along with LN's and metastases

Surgical Conditions of the Large Bowel

Polyps

- Are neoplastic, hamartomas, or inflammatory
- Neoplastic polyps are MC adenomas

Adenomas can be classified as:

- Tubular (these have the smallest potential for malignancy)
- Tubulovillous
- Villous (these have the highest risk of malignancy)

Signs and Symptoms:

- MC presents with intermittent rectal bleeding

Diagnosis:

- Colonoscopy or sigmoidoscopy

Treatment:

- Polypectomy

Diverticular Disease

General Information:

- Up to half of the population has diverticula
- The risk increases after 50yr of age
- Only 1/10 people are symptomatic when diverticula are present
- A TRUE diverticula is rare, and includes full bowel wall herniation
- A FALSE diverticula is most common, and involves only a herniation of the mucosa
- The MCC is a low-fiber diet which causes an increased intramural pressure (this is hypothesis)

Diverticulosis

- This is the presence of multiple false diverticula

Signs and Symptoms:

- Most people are asymptomatic, with diverticula found only on colonoscopy or other visual procedures
- May have recurrent bouts of LLQ abdominal pain
- Changes in bowel habits is common
- Rarely, patient may present with lower GI hemorrhage

Diagnosis:

- Colonoscopy
- Barium enema can also be used for diagnosis

Treatment:

- If patient is asymptomatic, the only therapy should be to increase fiber and decrease fat in the diet
- If patient has GI hemorrhage, circulatory therapy is warranted (IV fluids, maintenance of hemodynamic stability)

Diverticulitis

- Inflammation of the diverticula due to infection
- There are many possible complications, such as abscess, extension into other tissues, or peritonitis

Signs and Symptoms:

- LLQ pain
- Constipation OR diarrhea
- Bleeding
- Fever
- Anorexia

Diagnosis:

- CT demonstrating edema of the large intestine
- DO NOT perform a colonoscopy or barium enema in an acute case, this might aggravate the problem

Complications:

- Perforation
- Abscesses
- Fistula formation
- Obstructions

Treatment:

- If there is an abscess, percutaneous drainage is required
- Most patients are managed well with fluids and antibiotics
- For perforation or obstruction, surgery is required

Obstruction of the Large Intestine

- Most common site of colon obstruction is the sigmoid colon

Common causes include:

- Adhesions
- Adenocarcinoma
- Volvulus
- Fecal impaction

Signs and Symptoms:

- Nausea/vomiting
- Abdominal pain with cramps
- Abdominal distention

Diagnosis:

- XRAY – showing a distended proximal colon, air-fluid levels, and an absence of gas in the rectum

Treatment:

- If there is severe pain, sepsis, free air, or signs of peritonitis there must be an urgent laparotomy

- Laparotomy if cecal diameter is >12cm

Volvulus

- Twisting and rotation of the large intestine
- Can cause ischemia, gangrene, perforation
- The MC site is the sigmoid colon
- Occurs most commonly in older patients

Signs and Symptoms:

- High-pitched bowel sounds
- Distention
- Tympany

Diagnosis:

- XRAY – "kidney bean" appearance (ie. Dilated loops of bowel with loss of haustra)
- Barium enema showing a "bird's beak" appearance – points to the site of rotation of the bowel

Treatment:

- Sigmoidoscopy or colonoscopy acts as diagnosis and treatment
- If this doesn't work, laparotomy is warranted

Cancer of the Colon

- Colon cancer is the 2nd MCC of cancer deaths
- Believed that a low-fiber, high-fat diet increases the risk
- There are many genetic factors that contribute to colon cancer, such as Lynch syndrome and HNPCC

Lynch Syndrome:

- LS 1 is an autosomal dominant predisposition to colon cancer that is usually right-sided
- LS2 is the same as LS 1 with the addition of cancers outside the colon, such as in the endometrium, stomach, pancreas, small bowel, and ovaries

Screening:

- Screening should start at 40yr in people with no risk factors
- If a family member has had cancer of the colon, screening should start 10yr prior to when they were diagnosed (assuming this is less than 40yr)
- Should have yearly stool occult tests
- Colonoscopy every 10yr
- And a sigmoidoscopy every 3-5yrs

Diagnosis:

- Obtain preoperative CEA (allows you to follow the progression or recession of the disease)
- Endoscopy + barium enema

Treatment:

- Surgical resection + LN dissection
- If disease is metastatic, add 5-FU to the post-operative regimen

Follow-up:

- CEA levels every 3 months for 3 years
- Perform a colonoscopy at 6 and 12 months, then yearly for 5 years
- If a recurrence is suspected, a CT should be performed

Surgical Conditions of the Rectum and Anus

Hemorrhoids

- Varicosities of the hemorrhoidal plexus
- Often related to strenuous bowel movements

Signs and Symptoms:

- Bright red blood per rectum
- Itching
- Burning
- Palpable anal mass
- Internal hemorrhoids are NOT painful, while external hemorrhoids ARE painful

Treatment:

- Usually self-limiting
- Sitz bath
- Hemorrhoidal cream
- Stool softeners to relieve pain

Thrombosed Hemorrhoids

- These are not a true hemorrhoid, but are external hemorrhoidal veins of the anal canal
- They are a painful bluish elevation that lie beneath the skin

Classifications:

- 1° hemorrhoids involve no prolapse
- 2° hemorrhoids classically prolapse with defecation but return without manual reduction
- 3° hemorrhoids prolapse with either straining or defecation and require manual reduction
- 4° hemorrhoids are not capable of being reduced

Treatment:

- Conservative therapies
- Sclerotherapy, rubber band ligation, and surgical hemorrhoidectomy

Anal Fissure

- A crack or tear in the anal canal
- Usually occurs after the passage of diarrhea or constipation

Signs and Symptoms:

- The most common presentation is the passage of a painful bowel movement that is accompanied by bright red blood

Diagnosis:

- Perform an anoscopy to diagnose

Treatment:

- Bulking agents and stool softeners are usually all that is needed
- If fissures persist despite conservative measurements, a lateral internal sphincterotomy may be required

Anal and Rectal Cancer

Anal Cancer:

- The most common form is squamous cell carcinoma

Signs and Symptoms:

- Anal bleeding, pain, and mucus upon evacuation

Diagnose:

- Biopsy

Treatment:

- Chemotherapy + Radiation

Rectal Cancer:

- Seen in males > females

Signs and Symptoms:

- Rectal bleeding, altered bowel habits, tenesmus, obstruction

Diagnosis:

- Colonoscopy

Treatment:

- Surgery that spares the sphincter
- If metastasis involved, addition of 5-FU chemotherapy + radiation

NEUROSURGERY

Tumors of the Brain

- Often presents as a ***headache*** that is severe enough to awaken the patient during the night
- ***Increased intracranial pressure*** causing nausea, vomiting, and Cushing's triad (Bradycardia, hypertension, and Cheyne-Stokes respiration)
- Presence of focal deficits
- Often presents with a fixed, dilated pupil

Diagnosing a brain lesion:

- The most accurate diagnosis comes from biopsy
- With clinical suspicion a CT and/or MRI can often help make the diagnosis

Treatment:

- Excision is the best treatment for all tumors (except prolactinoma and lymphoma)
- PROLACTINOMA – give bromocriptine to shrink it, then surgery if this doesn't work
- LYMPHOMA – radiation is the treatment of choice
- If there is metastasis of brain tumors, adjunct therapy is radiation

Differentiating between the different types of brain tumors

Glioblastoma Multiforme:

- The most common 1° CNS neoplasm
- Is large and irregular with a ring-enhancing appearance

Meningioma:

- The 2nd MC 1° CNS neoplasm
- Grows slowly
- Benign

Retinoblastoma:

- Occurs in children and is often bilateral
- 40% of cases are familial while the rest are sporatic cases

Medulloblastoma:

- Common in children
- Found in the cerebellum/4th ventricle

Prolactinoma:

- Is the MC pituitary tumor
- Presents with many endocrine disturbances such as amenorrhea, impotence, galactorrhea, and gynecomastia.
- The MC presenting symptoms is visual disturbance (bitemporal hemianopsia)

Lymphoma:

- MC CNS tumor in AIDS patients
- An MRI shows a ring-enhanced lesion
- Often confused with toxoplasmosis

Schwannoma:

- A tumor that affects the 8th cranial nerve
- Presents with tinnitus, loss of hearing, and increased intracranial pressure

Hydrocephalus

- An increase in CSF causes an enlargement of the ventricles

Signs and Symptoms:

- ⇑ ICP, ⇓ cognition
- Headache
- Focal neurological deficits

Diagnosis:

- A CT or MRI can show the dilation of ventricles
- A lumbar puncture can help determine the type of hydrocephalus
- If ICP is normal, it is a communicating hydrocephalus (presents with urinary incontinence, dementia, and ataxia)
- If ICP is ⇑, it may be either communicating or non-communicating (Pseudotumor cerebri, congenital)

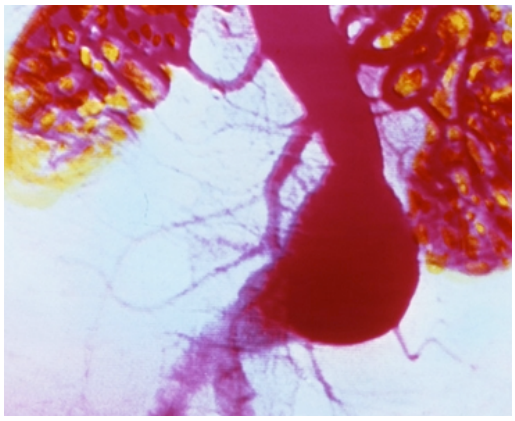
Treatment:

- If possible, treat the underlying cause
- If not possible, a shunt should be placed (usually drained into peritoneum)

Surgical Conditions of the Vascular System

Aneurysms

- Is a dilatation of an artery to greater than two times its normal diameter
- "True" aneurysms involve all 3 layers of the vessel, and are caused most commonly by atherosclerosis and congenital disorders
- "False" aneurysms are covered only by the adventitia of the vessel, and are most commonly caused by trauma



Signs and Symptoms:

- Gastric/epigastric discomfort
- Back pain
- Commonly in the abdomen (abdominal aorta aneurysms)
- Also commonly in the peripheral vessels

Complications:

- A rupture of an abdominal aneurysm is an emergency
- Presents with abdominal pain, a pulsatile abdominal mass, and severe hypotension

Diagnosis:

- Ultrasound can help detect aneurysms
- CT is the best test to determine size
- The most accurate test is the aortogram

Treatment:

- Control blood pressure
- Reduce risk factors
- Surgery recommended if aneurysms are >5cm

Peripheral Vascular Disease (PVD)

- Due to atherosclerosis

Signs and Symptoms:

- Presents with claudication
- Patient may have smooth and shiny skin with a loss of hair in the affected area

Diagnosis:

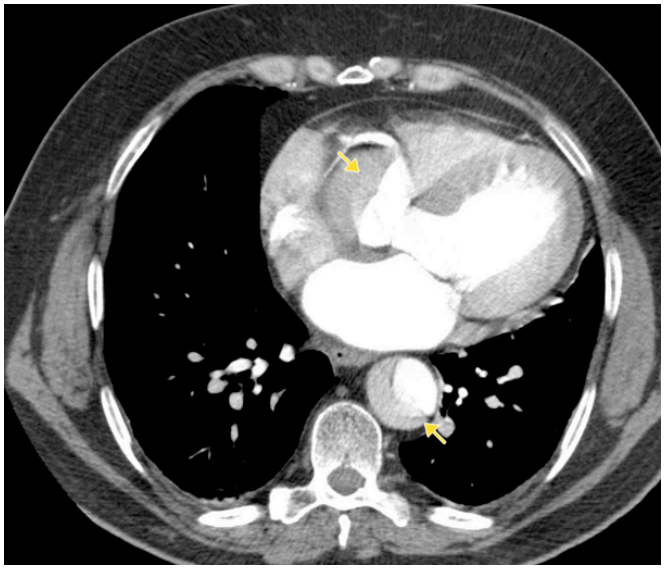
- Ankle:Brachial Index (ABI) is the best initial test – normal test is ≥ 0.9
- The most accurate test is an angiography

Treatment:

- Lifestyle modifications such as cessation of smoking and incorporation of exercise
- Control lipids with an LDL <100
- Control blood pressure
- Daily aspirin
- Surgery is required if there is pain at rest, necrosis, intractable claudication, and/or a non-healing infection

Aortic Dissection

- Is a dissection of the thoracic aorta
- Presents with intense "tearing" pain that radiates to the back
- There is a difference in blood pressures between the right and left arm



Diagnosis:

- The best initial test is a CXR – showing a widening of the mediastinum
- The most accurate test is the CT angiography

Treatment:

- Urgent blood pressure control with β-blockers followed by nitroprusside to maintain a decreased blood pressure
- Urgent EKG and CXR
- Then get a TEE or CT
- Surgical correction is necessary, otherwise this is rapidly fatal.

Subclavian Steal Syndrome

- An occlusion of the subclavian artery leads to a decreased blood flow distal to the obstruction
- The vertebral artery "steals" the blood due to retrograde flow
- Patient experiences claudication of the arm, nausea, syncope, and supraclavicular bruit

Diagnose:

- Angiography
- Doppler ultrasound
- MRI

Treatment:

- Carotid-subclavian bypass

Carotid Vascular Disease

- Is an atherosclerotic plaque in the carotid arteries

Signs and Symptoms:

- Patient may present with a TIA
- Amaurosis fugax (blindness in one eye)
- Carotid bruit

Diagnosis:

- Angiography

Treatment:

- Decrease the modifiable risk factors
- Aspirin
- Other anticoagulation medications
- Surgery is warranted if there is stenosis >70%, if patient has recurring TIA's, or if they have suffered from a previous cerebrovascular accident

Surgical Conditions of the Urinary System

Testicular Torsion

- Usually occurs in a younger patient
- Acute edema and severe testicular pain
- Patient usually experiences nausea and vomiting due to the degree of the pain
- ABSENCE of the cremasteric reflex
- Presence of scrotal swelling
- Testicle may have a horizontal lie

Diagnosis:

- Ultrasound to assess arterial patency
- Upon elevation of the teste, the pain is not alleviated

Treatment:

- 1st step is to secure the circulation
- 2nd step is to evaluate the need for excision of the testicle if it is dead

Epididymitis

- Unilateral pain of the testicle
- Dysuria
- Painful and swollen epididymus
- Less common in prepubertal children as opposed to torsion

Diagnosis:

- Swab for Chlamydia and Gonorrhea

Treatment:

- NSAIDs and antibiotics

Prostate Cancer

- Obstructive symptoms
- Rock-hard nodule in the prostate

Diagnosis:

- PSA

- Serum phosphatase
- Azotemia
- Transrectal ultrasound

Treatment:

- The only surgical requirement is a radical prostatectomy in very severe cases – risk of incontinence and/or impotence

Orthopedic conditions requiring surgery

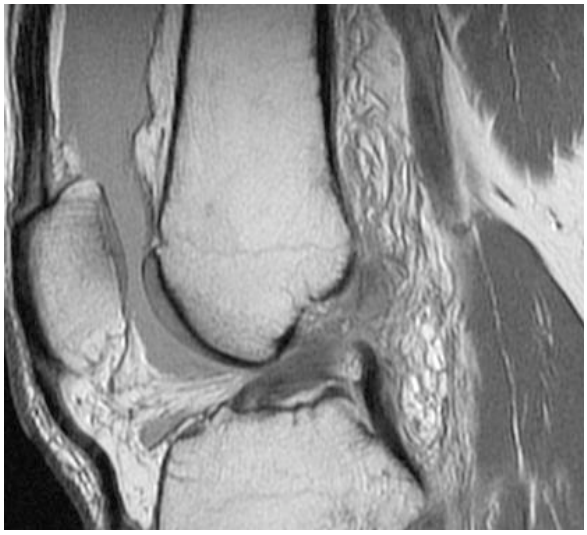
Knee Injuries

Include:

- Anterior cruciate ligament tears
- Posterior cruciate ligament tears
- Collateral ligament tears
- Meniscus tears

Anterior Cruciate Ligament tears:

- Injury history usually reveals a "pop" sound during the trauma
- The Lachman test (anterior drawer test) is used in the field to make a diagnosis
- MRI is the test of choice to determine the severity of the injury
- Treatment is either with conservative measures, or if severe with arthroscopic repair

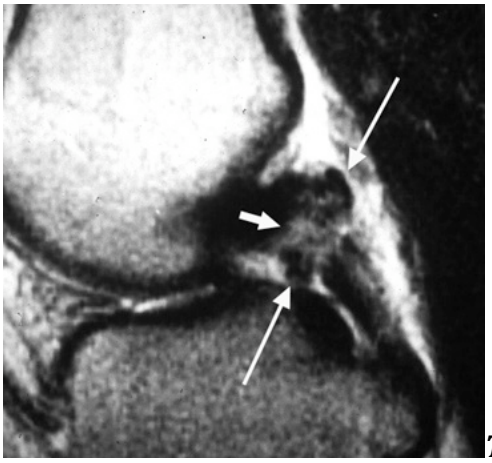


Tear of the ACL with associated joint effusion

Posterior Cruciate Ligament tears:

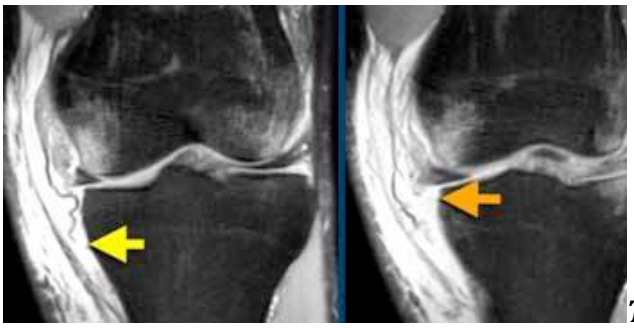
- Injury usually occurs when the knee is flexed
- "Posterior dresser drawer sign"
- MRI is the test of choice to determine severity of the injury

- Treatment is either with conservative measures, or if severe with arthroscopic repair


Tear of the PCL

Collateral Ligament tears:

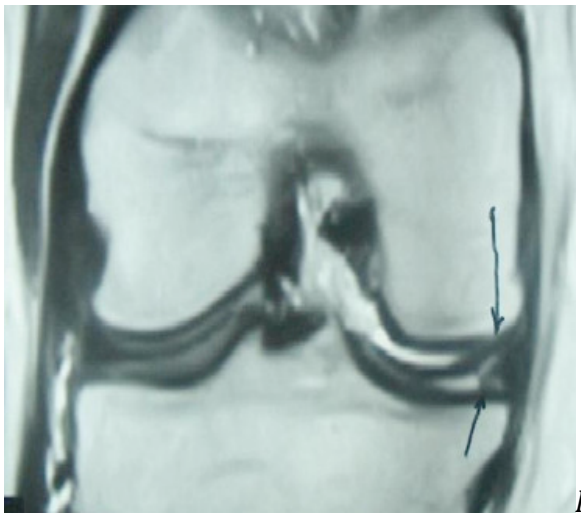
- The MCL is the most commonly injured ligament
- Seen with a direct blow to the lateral knee
- Is commonly injured in conjunction with the injury to the ACL or PCL
- MRI to determine severity of the injury
- Knee brace


Tear of the MCL

Meniscus tears:

- Often seen in older patients and is due to degeneration
- Injury is most commonly seen in the medial meniscus and is much more common in men

- Diagnose with McMurray's test
- MRI to determine the severity of the injury
- Treatment is usually rest alone, if severe can treat with arthroscopic surgery



Medial meniscus tear

<u>Shoulder Injuries</u>

Rotator Cuff Injury

- Can range from mild tendonitis to severe tears
- Involve the Supraspinatus, Infraspinatus, Teres Minor, Subscapularis

Signs and Symptoms:

- Pain and tenderness of the deltoid with movement
- Pain over the anterior aspect of the humeral head
- Neer's sign (+) – pain elicited when the arm is forcefully elevated forward

Diagnosis:

- Clinical suspicion
- MRI is used for confirmation

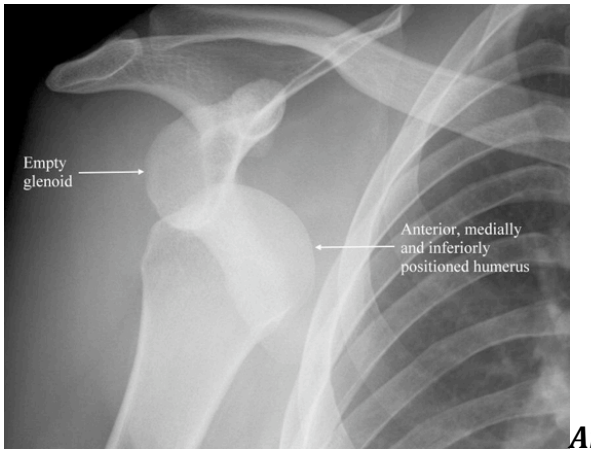
Treatment:

- NSAIDs
- Steroids injections

- For severe diseases that are not successfully treated with steroids, arthroscopic surgery is helpful

Dislocation of the Shoulder

- Is most commonly an anterior dislocation
- Posterior dislocation seen when patient is electrocuted and/or experiences a status epilepticus seizure



Anterior dislocation of the humerus

Signs and Symptoms:

- Immobility
- Extreme pain

Treatment:

- Traction-countertraction techniques to put the bone back in the socket
- Immobilization period (2-6 weeks)

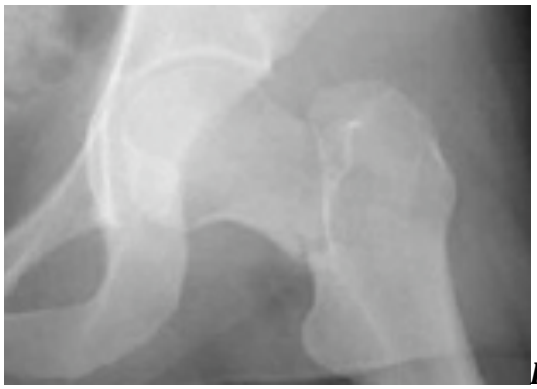
Hip and Thigh Injuries

Dislocations

- Dislocations require emergency reduction under sedation
- Risk of injury to sciatic nerve
- Avascular necrosis is a severe complication

Femoral Neck Fracture

- Requires significant force for injury
- Produces severe pain of the hip/groin that is exacerbated with movement
- Leg is classically "externally" rotated
- Diagnose with xray
- Requires surgical reduction and internal fixation

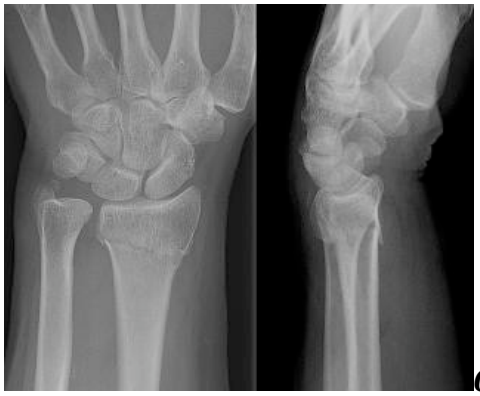


Fracture of the left femoral neck

Wrist Injuries

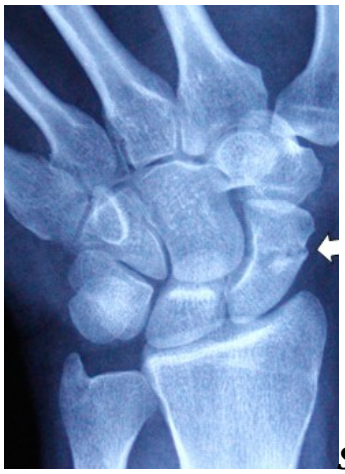
Colles' Fracture:

- This is a fracture to the distal radius
- Occurs after falling on an outstretched hand
- Diagnose with H & P and xray
- Treat with cast immobilization for 2-4 wk


Colles' fracture

Scaphoid Fracture:

- Almost always secondary to a fall
- Most commonly misdiagnosed as a sprained wrist
- Diagnosis is classically made when there is pain in the anatomic snuff box
- Manage with a thumb splint for 10 weeks
- Complication is avascular necrosis


Scaphoid Fracture

Carpal Tunnel Syndrome

- Presents with pain, numbness, tingling of the hands along the distribution of the median nerve

Diagnosis:

- Pathognomonic sign is "Tinel's Sign", where tapping over the palmar aspect of the wrist elicits shooting pains
- "Phalen's test" is also diagnostic

Treatment:

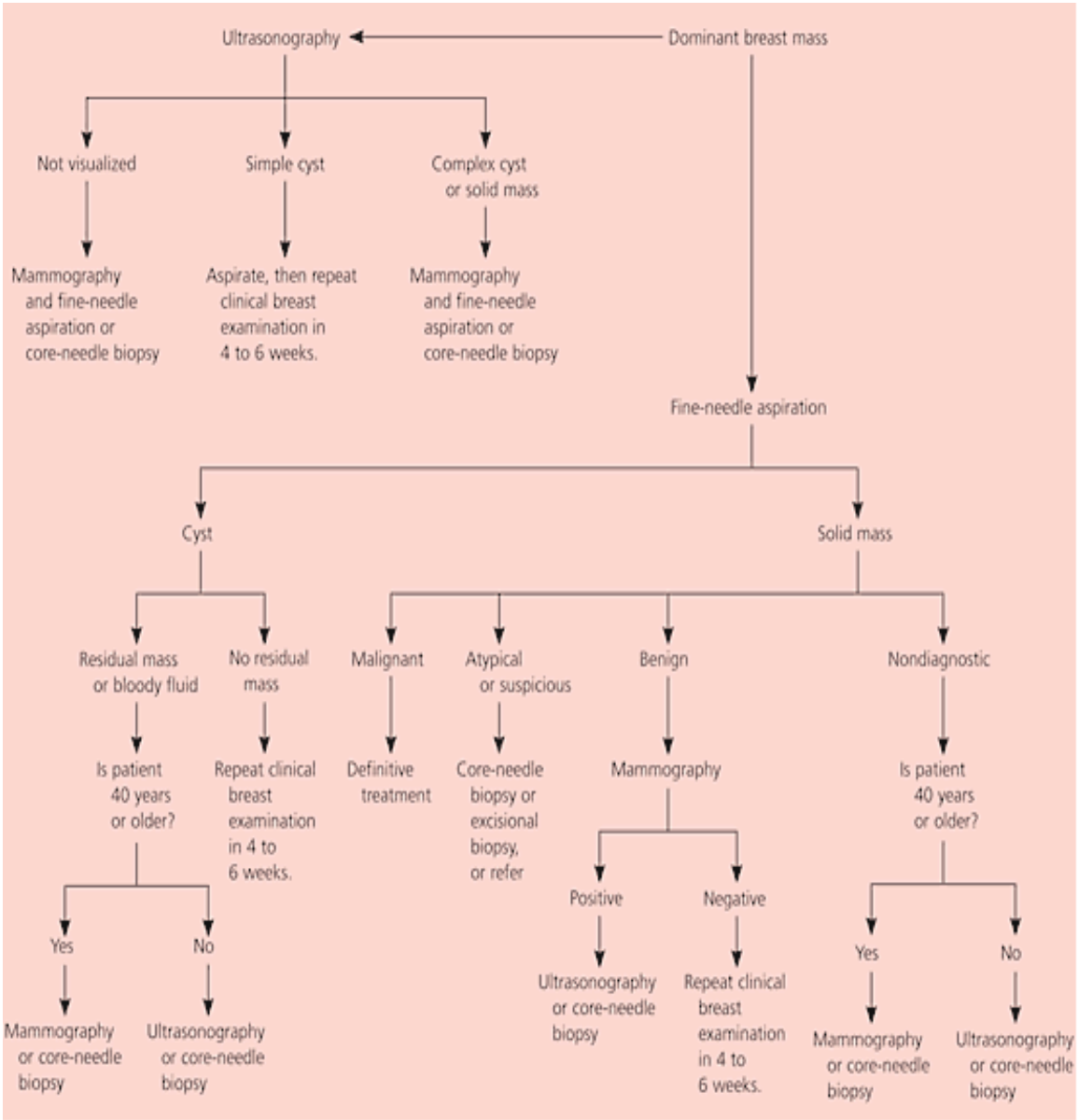
- Treat by avoidance of aggravating activity, use wrist splints which hold the wrist in ***slight extension***
- Severe cases should first be managed with steroid injection in the carpal tunnel, if no treatment surgery is performed

<u>Wrist Splints:</u>

Holds the wrist in a position of 'slight extension'

The Breast

Workup of a Breast Mass Algorithm (AAFP)



Cancer Risks

- The #1 risk factor for breast cancer is gender (Female >>> Male)
- In women, age is the #1 factor for breast cancer risk
- Late menopause increases the risk of breast cancer (after 50yr)
- If less than 11yr at menarche, the risk of breast cancer is increased
- If >30yr at first pregnancy, the risk for breast cancer increases

History of Fibroadenoma and/or Fibrocystic disease does not increase the risk of getting breast cancer

Family History and Breast Cancer:

- Only 5% of breast cancers are familial
- With a 1st degree relative being affected, the risk of cancer increases
- Autosomal dominant conditions with increased risk: BRCA-1, BRCA-2, Li-Fraumeni syndrome, Cowden's disease, Peutz-Jeghers

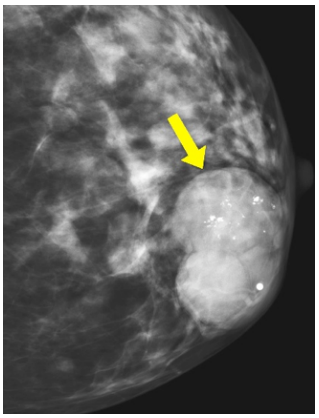
Tumors of the Breast

Mammogram

- All women >40yr (controversial as to age to start) should have yearly mammograms
- Not effective in young patients because the breast tissue is too dense

Fibroadenoma

- The classic presentation is a firm, non-tender, mobile breast nodule
- Most commonly seen in teens and younger women



Diagnosis:

- Breast exam
- FNA
- Follow-up clinical breast exam in 6 weeks

Treatment:

- Not required as this condition is not a cancer precursor and often disappears on its own

When to perform certain tests pertaining to breast masses:

A palpable mass that feels cystic always requires an ultrasound first.

A palpable mass that doesn't feel cystic requires a FNA (after an US or instead of an US).

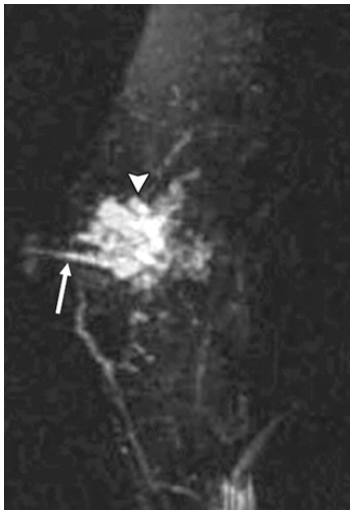
Any FNA that reveals bloody fluid requires cytology.

Always do a mammogram in patients >40yr who present with almost all pathologies of the breast.

A biopsy is required when a cyst recurs more than 2 times within 4 weeks, when there is bloody fluid, when there are signs of inflammatory breast disease, and when a mass does not disappear with FNA.

Fibrocystic Disease

- This presents with multiple/bilateral painful lumps in the breast that vary in pain with the menstrual cycle
- Is the most commonly seen breast tumor in women between 35-50yr of age



Diagnosis:

- Fine-needle aspiration to drain fluid, and it will collapse after the FNA

Treatment:

- OCP's can help prevent this from occurring

Pre-Invasive Breast Cancers

Include Ductal Carcinoma In Situ and Lobular Carcinoma In Situ

Ductal Carcinoma In Situ (DCIS)

- It's presence increases the risk of invasive breast cancers
- Usually non-palpable and seen on mammogram as irregularly shaped ductal calcifications
- Will lead to invasive ductal carcinoma

Diagnosis:

- Histology shows puched-out areas in ducts and haphazard cells along the papillae

Treatment:

- Surgical excision ensuring clean margins
- Post-operative radiation is recommended to decrease the risk of recurrence (Can give Tamoxifen in addition to radiation or instead of radiation)

Lobular Carcinoma In Situ (LCIS)

- In contrast to DCIS, this is not precancerous, it does however increase the risk of future invasive ductal carcinoma

Diagnosis:

- Hard to diagnose with mammogram
- Cannot be detected clinically
- The histology shows mucinous cells in the classic “saw-tooth” pattern

Treatment:

- Tamoxifen alone is used for treatment

Invasive Breast Cancers

Treatment for all invasive cancers:

1. If lump is <5cm, Lumpectomy + radiation, may add chemo and adjuvant therapy.
2. Perform sentinel node biopsy (preferred over an axillary node biopsy)
3. Test for estrogen/progesterone receptors and the HER2 protein
4. If tumor is >5cm, the treatment involves systemic therapy

Invasive Ductal Carcinoma

- Is the most common form of breast cancer, seen in almost 85% of all cases
- Is unilateral
- Metastasizes to the brain, liver, and bone
- Important prognosis factors are size of the tumor and the lymph node involvement

Paget's Disease of the Breast

- Presents with an erythematous and scaly lesion of the nipple that is pruritic. Nipple may be inverted
- Nipple discharge common

Inflammatory Breast Cancer

- Less common
- Rapid growth/progression
- Early metastasis
- Red, swollen, pitted, and warm breast (peau d'orange)

Lobular Carcinoma

- Multifocal and within the same breast (usually)
- 20% of cases present as bilateral multifocal lesions

BRCA1 and BRCA2

Testing for these genes should be performed if there is a history of the following:

- Family history of early-onset breast cancer

- Family history of male breast cancer
- Past history of breast and/or ovarian cancer in that patient
- Ashkenazi Jewish heritage

Chapter 2

Obstetrics

Terminologies

Gravidty = total number of pregnancies

Parity = number of births with a gestational age >24 weeks

Term delivery = delivery after 37 weeks of gestation

Premature delivery = delivery of infant between 20 and 37 weeks

The Uncomplicated Pregnancy

Diagnosing Pregnancy

The presence of amenorrhea and + urinary ß-hCG suggests pregnancy.

Confirm pregnancy with the following:

- **Presence of gestational sac** [seen with transvaginal US at 4-5 weeks. ß-hCG level approx 1500mIU/ml.]
- **Fetal heart motion** [seen by US between 5-6 weeks.]
- **Fetal heart sounds** [heard with Doppler US at 8-10 weeks.]
- **Fetal movement** [on examination after 20 weeks.]

Estimating date of confinement (EDC)

Use Nagele's rule = Last Menstrual Period (LMP) + 7 days – 3 months + 1yr.

*Calculation accuracy depends on regular 28-day cycles.

DRUG CATEGORIES DURING PREGNANCY

Category	Description
A	**Medication has not shown an increase in risk for birth defects in human studies.**
B	**Animal studies have not demonstrated a risk, and there are no adequate studies in humans, OR animal studies have shown a risk, but the risk has not been seen in humans.**
C	**Animal studies have shown adverse effects, but no studies are available in humans, OR studies in humans and animals are not available.**
D	**Medications are associated with birth defects in humans; however,**

	potential benefits in rare cases may outweigh their known risks.
X	**Medications are contraindicated in human pregnancy because of known fetal abnormalities that have been demonstrated in both human and animal studies.**

COMMON TERATOGENS IN PREGNANCY

Drug	Birth Defect
Lithium	**Ebstein's anomaly (single-chambered right side of heart).**
Carbamazepine, Valproate	**Neural tube defects.**
Retinoid Acid	**CNS defects, craniofacial defects, cardiovascular defects.**
ACE Inhibitors	**Decreased skull ossification, renal tubule dysgenesis, renal failure in neonate.**
Oral hypoglycemics	**Neonatal hypoglycemia**
Warfarin	**CNS & Skeletal defects**
NSAIDs	**Necrotizing enterocolitis, constriction of ductus arteriosis.**

At first visit upon discover of pregnancy

Perform the following:

- Complete physical exam with pelvic and Pap smear.
- Culture for gonorrhea and Chlamydia

Labs include the following:

- CBC
- Blood type with Rh status
- UA with culture
- RPR for syphilis
- Rubella titer
- TB skin test
- Offer HIV test

Additional testing:

- Genetic testing if history indicates the need
- If pt not immune to rubella, DO NOT immunize (live virus).

Recommend:

- Folic acid
- Iron
- Multi-vitamin
- 25-35 lb weight gain during pregnancy

What to do during each trimester

1st trimester:

Should see patient every 4 weeks.

Assess:

- Weight gain/loss
- Blood pressure
- Edema
- Fundal height
- Urine for glucose and protein
- Estimation of gestational age by uterine size

2nd trimester:

Continue to see the patient every 4 weeks

Assess:

- At 12 weeks use Doppler US to evaluate fetal heart beat (each visit)
- Offer triple-marker screen (ß-hCG, estriol, α-fetoprotein(AFP)) at 15-18 weeks, [AFP decreased in Down's syndrome], [AFP increased in multiple gestation, neural tube defects, and duodenal atresia].
- Document quickening (fetal movement) at 17-19 weeks and beyond.
- Amniocentesis if mother is >35yr or if history indicates (hx of miscarriages, previous child with deficits, abnormal triple-marker screen).
- Glucose screening at 24wk
- Repeat hematocrit at 25-28wk

⇑ MS-AFP:

Neural tube defect (NTD), ventral wall defect, twin pregnancy, placental bleeding, renal disease, sacrococcygeal teratoma.

⇓ MS-AFP:

Trisomy 21, Trisomy 18

*** The most common cause of inaccurate lab results is dating error.**

SECOND TRIMESTER ROUTINE TESTS

Screening	Test	Diagnostic Significance	Next Step in Mgmt
Diabetes	**1hr 50g OGTT (24-28 wks)**	**Abn if >140mg/dL**	**If +, perform 3hr 100g OGTT**
Anemia	**CBC measured between 24-28 weeks.**	**Hb <10g/dL = anemia**	**Iron supplementation**
Atypical antibodies	**Indirect Coombs test**	**Performed in Rh(-) women looking for antibodies (anti-D Ab) before giving RhoGAM**	**RhoGAM not indicated in Rh (-) women who have developed anti-D antibodies**
GBS screening	**Vaginal and rectal culture for group B strep at 35-37 weeks.**	**(+) GBS is a high risk for sepsis in newborns. [treat with intrapartum IV antibiotics].**	**IV: Pen G Clindamycin Erythromycin in PCN allergic patient.**

3rd trimester:

See patient every 4 weeks until week 32, every 2 weeks from week 32-36, then every week until delivery.

Assess:

- Inquire about preterm labor [vaginal bleeds, contractions, rupture of membranes].
- Inquire about pregnancy-induced hypertension.
- Screen for group B streptococcus at 35-37 weeks.
- Give RhoGAM at 28-30 weeks if indicated

The confirmatory test for diabetes in pregnancy is the 3hr 100g oral glucose tolerance test (OGTT).

- Plasma glucose >125mg/dL at beginning of test = DM
- Abnormal plasma glucose is >140mg/dL at 1hr, >155mg/dL at 2hr, and >180mg/dL at 3hr.
- If 1 post glucose load measurement is abnormal, impaired glucose tolerance is the diagnosis.
- If 2 or more post glucose load measurements are abnormal, gestational diabetes is the diagnosis.

The following antiemetics are safe to use during pregnancy:

- Doxylamine
- Metoclopramide
- Ondansetron
- Promethazine
- Pyridoxine

The Complicated Pregnancy

Bleeding after 20 weeks (late pregnancy)

Most common causes of late pregnancy vaginal bleeding are:

1. Abruptio placenta
2. Placenta previa
3. Vasa previa
4. Uterine rupture

* Never perform a digital or speculum exam in any patient with late vaginal bleeding until a vaginal ultrasound has ruled out placenta previa

ABRUPTIO PLACENTA

- Sudden onset of severely painful vaginal bleeding in patient with history of hypertension or trauma.
- Bleeding may be concealed, in which case there will be severe, constant pain without the presence of blood.
- DIC is a feared complication

Management:

- Emergent C-section if patient or fetus is deteriorating
- Admit and observe if bleeding has stopped, vitals and HR are stable, or fetus is <34 weeks.

PLACENTA PREVIA

- Sudden onset of painless bleeding that occurs at rest or during activity without warning.
- May include history of trauma, sexual activity, or pelvic exam before onset.
- Occurs when the placenta is implanted in lower uterine segment
- Best management is emergency C-section.

3 forms of placenta previa:

1. Accreta – does not penetrate entire thickness of endometrium

2. Increta – extends further into the myometrium
3. Percreta – placenta penetrates entire myometrium to uterine serosa

VASA PREVIA

- A condition life-threatening to the fetus.
- Occurs when vilamentous cord insertion results in umbilical vessels crossing the placental membranes over the cervix. Membrane rupture causes tearing of the fetal vessels, and blood loss is from the fetal circulation.
- Fetal bleeding and death occur rapidly.

Management:

Immediate C-section.

Classic triad of vasa previa:

1. Rupture of membranes
2. Painless vaginal bleeding
3. Fetal bradycardia

- Emergency C-section is always the first step in management

UTERINE RUPTURE

- Occurs when there's a history of uterine scar with sudden-onset of abdominal pain and vaginal bleeding.
- Associated with a loss of electronic fetal HR, uterine contractions, and recession of the fetal head.

Management:

Immediate surgery and delivery

Comparing Placenta Previa and Placental Abruption

	Placenta Previa	Placental Abruption
Abnormality	Placenta implanted over internal cervical os (completely or partially)	Premature separation of normally implanted placenta from decidua
Epidemiology	⇑ Risk grand multiparas and prior C-section	⇑ Risk preeclampsia, previous history of abruption, ROM in a pt with hydramnios, cocaine use, cigarette smoking, and trauma.
Time of onset	20-30 weeks	Any time after 20 weeks
Signs & Symptoms	Sudden, painless bleeding	Painful bleeding, can be heavy and painful, with frequent uterine contractions
Diagnosis	US → Placenta in abnormal location	Clinical, based on presentation of painful vaginal bleeding, frequent contractions, and fetal distress.
Treatment	Hemodynamic support, expectant management, delivery by C-section when fetus is mature enough	Hemodynamic support, urgent C-section or vaginal induction if patient is stable and fetus is not in distress
Complications	Associated with a two-fold increase in congenital malformations so evaluations for fetal anomalies should be undertaken at diagnosis	⇑ Risk of fetal hypoxia and/or death, DIC may occur as a result of intravascular and retroplacental coagulation.

Hypertension in Pregnancy

Hypertension in pregnancy predisposes both the mother and fetus to serious conditions. A **BP of ≥ 140/90** during pregnancy can be classified as chronic hypertension or gestational hypertension.

- Hypertension accompanied by signs and symptoms of end-organ damage or neurological sequelae is diagnosed as **preeclampsia**, **eclampsia**, or **HELLP syndrome**.
- Sustained hypertension may cause fetal growth restriction and hypoxia, and increase the risk of **abruptio placenta**.

Diagnosis:

- Elevated pregnancy before pregnancy or before 20 weeks gestation = chronic hypertension
- Development of hypertension after 20 weeks gestation that returns to normal baseline by 6 weeks post-partum = gestational hypertension
- Presence of proteinuria and/or presence of warning signs = preeclampsia

Warning Signs of Preeclampsia:

Hallmark symptoms include:

- Headache
- Epigastric pain
- Visual changes/disturbances

Signs:

- Pulmonary edema
- Oliguria

Labs:

- Thrombocytopenia
- Elevated liver enzymes

Disease	Characteristics
Preeclampsia	• HTN (>140/90 or ⇑ systolic BP >30 mmHg or diastolic BP >15 mmHg compared to previous BP). • New onset proteinuria and/or edema. • Commonly around week 20
Severe Preeclampsia	• SBP >160 mmHg or DBP >110 mmHg. • Significant proteinuria (>1g/24hr urine collection or >1+ on dip) • CNS disturbances such as headache or visual disturbance • Pulmonary edema • RUQ pain
Eclampsia	• CONVULSIONS • 25% occur before labor, 50% during labor, 25% in first 72hr post-partum

Primiparas – are at greatest risk for eclampsia.

Chronic hypertension with superimposed preeclampsia – is diagnosed when there is chronic hypertension with increasingly severe hypertension, proteinuria, and/or warning signs.

Eclampsia – is the diagnosis when there is unexplained grand mal seizures in a hypertensive and/or proteinuric in a woman in the last half of pregnancy. Seizures are due to severe diffuse cerebral vasospasm, which cause cerebral perfusion deficits and edema

HELLP syndrome – is diagnosis when there is **H**emolysis, **E**levated **L**iver enzymes, and **L**ow **P**latelets.

Treatment:

- BP goal is 140-150 mmHg (systole) and 90-100 mmHg (diastole).
- Don't treat unless BP is >160/100 mmHg

Maintenance Therapy:

- First line therapy – Methyldopa
- Second line therapy - β-blockers such as labetalol

* β-blocker use in pregnancy can cause IUGR

Acute elevation in BP:

- IV hydralazine or labetolol

Seizure management:

- Airway protection first
- IV Magnesium Sulfate bolus for seizure and infusion for continued prophylaxis
- Induce labor is fetus is ≥36 weeks with mild preeclampsia and attempt vaginal delivery with IV oxytocin if both mother and fetus are stable

HELLP Syndrome

- Occurs in up to 10% of preeclampsia patients
- Occurs in third trimester, may occur 2 days after delivery
- Risk factors include: whites, multigravids, older maternal age

Management:

- Immediate delivery at any gestational age
- IV corticosteroids if platelets are <100,000/mm^3 both antepartum and postpartum, continuing until platelets are >100,000/mm^3 and liver functions normalize
- Give platelet transfusion if platelets drop below 20,000/mm^3 or platelet count less than 50,000/mm^3 if C-section will be performed
- IV Magnesium Sulfate for seizure prophylaxis
- Steroids for fetal lung maturity.

Complications of HELLP:

- DIC
- Placental abruption
- Fetal death

- Hepatic rupture
- Ascites

Normal physiologic changes in pregnancy

There are many normal physiological changes that occur in pregnancy, these include the following.

Hematological – pregnancy creates a state of hypercoagulability

- ⇑ clotting factor levels
- Venous stasis caused by uterine pressure on lower-extremity veins

Anemia of Pregnancy:

- Between weeks 6-30, plasma volume increases approximately 50%
- Red cell mass increases to a smaller degree, anemia due to 15% dilution
- Slight leukocytosis
- Platelets decrease slightly but still remain wnl.

Cardiac –

- CO ⇑ 50% (both HR and SV increase)
- ⇑ flow causes ⇑ S2 split with inspiration, distended neck veins, systolic ejection murmur, and S3 gallop are normal findings.
- ⇓ PVR due to progesterone-mediated smooth muscle relaxation
- BP ⇓ during first 6 months, then return to normal

Pulmonary –

- Mucosal hyperemia causes nasal stuffiness and nasal hypersecretion
- Diaphragm elevates due to uterus expansion
- Tidal volume and minute ventilation ⇑ 30-40%
- Functional residual capacity and residual volume ⇓ 20%

- Hyperventilation that allows for a pressure gradient so maternal 02 can transfer to fetus
- Respiratory rate, vital capacity, inspiratory reserve all remain unchanged.

GI –

- ⇓ GI motility
- ⇓ esophageal sphincter tone (leads to GERD)
- ⇑ alkaline phosphatase
- Hemorrhoids caused by constipation and ⇑ venous pressure due to enlarging uterus compression on IVC

Renal –

- ⇓ bladder tone due to progesterone predisposes to urinary stasis and UTI's/pyelonephritis
- GFR ⇑ by 50%, thus glucose excretion without increased protein loss
- Serum creatinine and BUN decrease

Endocrine –

- ⇓ fasting glucose in mother due to fetal utilization
- ⇑ postprandial glucose in mother due to ⇑ insulin resistance
- At 9-11 weeks the fetus produces its own insulin
- ⇑ maternal thyroid-binding globulin (TBG) due to ⇑ estrogen, ⇑ total T3 and T4 due to ⇑ TBG.
- ⇑ cortisol and cortisol-binding globulin

Skin –

- Increased estrogen causes skin changes similar to those in liver disease
- Spider angiomas, palmar erythema
- Hyperpigmentation due to ⇑ estrogen and melanocyte-stimulating hormone

Medical Complications in Pregnancy

Cardiac Abnormalities

- Heart disorders account for up to 10% of maternal obstetric deaths
- Women with very high-risk disorders should be advised against pregnancy due to increased risk of death
- Cardiovascular physiological changes may unmask and worsen underlying conditions, seen maximally between 28-34 weeks gestation.

Peripartum Cardiomyopathy

- From the 8th month until 5 months post-partum, heart failure without identifiable cause is possible
- Risk factors include multiparity, age over 30, history of multiple gestations, and preeclampsia
- 5-yr mortality rate is 50%

Managing specific cardiac conditions

Heart Failure:

- Loop diuretics, nitrates, and β - blockers
- Digoxin may improve symptoms but does not improve outcome.
- Never use ACEI's in pregnancy

Arrhythmias:

- Rate control as with non-pregnant patients
- Never give warfarin or amiodarone

Endocarditis Prophylaxis:

- Same as in non-pregnant patient
- Daily prophylaxis in patient with rheumatic heart disease

Valvular Disease:

- Regurgitant lesions do not require therapy
- Stenotic lesions have increased risk of maternal and fetal mortality
- Mitral stenosis has increased risk of pulmonary edema and Afib.

Hypercoagulable States

Pulmonary Embolus:

- The leading cause of maternal death in US

When to anticoagulate:

- Anticoagulate when DVT or PE in pregnancy
- Anticoagulate when Atrial Fibrillation is present with underlying heart disease
- When patient has antiphospholipid syndrome
- When ejection fraction is <30%

The anticoagulant of choice is low molecular weight heparin (won't cross placenta)

- Patients with a history of DVT or PE in a previous pregnancy should receive prophylactic LMWH throughout the pregnancy, unfractionated heparin during labor and delivery, and warfarin for 6 weeks postpartum.

Thyroid Disorders

- Hyperthyroidism in pregnancy causes fetal growth restriction and stillbirth
- Hypothyroidism in pregnancy can cause intellectual deficits and miscarriage
- β-blockers are the DOC for symptoms due to hyperthyroidism
- Radioactive iodine never given in pregnancy

Grave's Disease:

- Propylthiouracil (PTU) is DOC in pregnancy
- Methimazole is 2nd line drug
- PTU can cross the placenta and cause goiter + hypothyroidism.
- Maternal thyroid-stimulating Ig's and thyroid-blocking Ig's can cross the placenta and cause fetal tachycardia, growth restriction, and goiter.

Diabetes in Pregnancy

- The target values for fasting blood sugar are between 90mg/dL and 120mg/dL 1hr after eating.
- Insulin-dependent DM requires insulin and is safe through pregnancy
- Gestational DM is managed solely through diet

- Oral hypoglycemics are contraindicated and should also be avoided if breastfeeding, since it can cause hypoglycemia.

Routine monitoring tests during pregnancy:

- Monthly sonograms
- Monthly biophysical profiles
- HbA1c each trimester
- Triple marker screen at 16-18 weeks assessing NTD's.
- At 32 weeks start weekly non-stress tests and amniotic fluid index
- If gestational DM, do a 2hr 75g OGTT at 6-12 weeks postpartum to see if diabetes has resolved

HbA1c > 8.5 in the first trimester is strongly associated with congenital malformations, especially neural tube defects!

Labor in the diabetic patients:

- Due to delayed fetal maturity, aim to deliver the baby at 40 weeks
- If there is poor glycemic control, induce labor at 39-40 weeks if <4,500g.
- Check L/S ratio, if >2.5 and there is presence of phosphatidyl glycerol this ensures lung maturity
- If baby is >4,500g, schedule a C-section
- Ensure maternal glucose is between 80-100mg/dL, this can be ensured with D5 (5% dextrose)
- Insulin resistance decreases rapidly after delivery, so keep an eye on insulin administration and maintain blood-glucose levels with a sliding scale

Common neonatal problems in diabetic pregnancy:

- Hypoglycemia
- Hypocalcemia
- Polycythemia
- Hyperbilirubinemia
- Respiratory distress syndrome

Liver Disease

Intrahepatic Cholestasis of Pregnancy:

- A genetic condition most commonly seen in European women

Si/Sx → Intractable nighttime pruritis of palms and soles of feet

Diagnosis → up to 100x increase in serum bile acids

Treatment → Ursodeoxycholic acid in treatment of choice. Antihistamines also helpful

Acute Fatty Liver of Pregnancy:

- A rare condition caused by disordered metabolism of fatty acids by the fetal mitochondria.
- Causes HTN, proteinuria, and edema that can mimic preeclampsia.

Diagnosis:

- ⇑ liver enzymes (ALT, AST, GGT)
- Hyperbilirubinemia
- DIC
- Hypoglycemia
- Increased serum ammonia

Treatment:

- Emergency situation requires ICU admission and aggressive IV fluids + immediate delivery

Urinary Tract Infections, Bacteriuria, and Pyelonephritis

Infection Type	Asymptomatic Bacteriuria	Acute Cystitis	Pyelonephritis
Symptoms	(+) urine culture NO burning NO frequency NO fever NO urgency	(+) urine culture BURNING FREQUENCY URGENCY NO fever	(+) urine culture BURNING FREQUENCY URGENCY FEVER + CVA TENDERNESS
Treatment	Outpatient Nitrofurantoin is the DOC	Outpatient Nitrofurantoin is the DOC	Admission, give: IV hydration IV cephalosporin's Give tocolytics
Possible Complications	Acute pyelonephritis if untreated in approximately 30%	Acute pyelonephritis if untreated in approximately 30%	Preterm labor and delivery. Severe cases may progress to sepsis, anemia, and pulmonary dysfunction

Ectopic Pregnancy/Tubal Pregnancy

The earlier the age of gestation the less there is a risk of complications.

1st trimester:

- D&C is the most common 1st trimester procedure.
- Give prophylactic antibiotics
- Perform under twilight anesthesia + paravertebral block
- Can perform medical abortion as an alternative to D&C with use of oral mifepristone (progesterone antagonist), and oral misoprostol (prostaglandin E1). * Must be performed within 63 of onset of amenorrhea.

Complications:

- Endometritis (rarely)
- Retained products of conception (POCs)

2nd trimester:

- D&C is the most common 2nd trimester procedure.
- For more advanced pregnancies, an "intact D&C" can be used.

Complications:

- Retained placenta is the most common immediate complication
- Cervical trauma/cervical insufficiency is the most common delayed complication

Abortion

- Termination of pregnancy usually ≤ 20 weeks, spontaneously in 15% of all pregnancies.
- Risk factors = ⇑ paternal age, conception within 3 months of live birth, ⇑ parity
- 50% are due to chromosomal abnormalities
- Vaginal bleeding in first half of pregnancy is presumed to be a threatened abortion until proven otherwise

Ectopic Pregnancy:

- Implantation outside of the uterine cavity
- PID increases the risk

Risk factors:

- Previous ectopic pregnancy
- History of gonorrhea, Chlamydia, PID, salpingitis
- Advanced maternal age (>35)
- > 3 prior pregnancies

Differential dx of ectopic pregnancy:

- Ovarian torsion
- Endometriosis
- Surgical abdomen
- Abortion
- Salpingitis
- Ruptured ovarian cyst

Diagnosing:

- (+) β-hCH + US to determine intra vs. extra uterine pregnancy
- Low progesterone level is suggestive of a non-viable pregnancy that may be located outside of the uterine cavity

Treatment:

- Laparoscopic removal, tubes usually heal on their own
- Methotrexate early, especially if size is <3.5cm in diameter + no cardiac activity on US
- Rh negative women should receive RhoGAM
- Serial β-hCH must be done to ensure success of termination
-

Types of Abortions

Threaten	**Inevitable**	**Completed**	**Incomplete**	**Missed**	**Recurr.**
Vaginal bleeding in first 20 wk without passage of tissues or ROM. Cervix is closed. Occurs in 25% of pregnancies. Diagnose with ultrasound. If no cardiac activity by week 9 consider a D&C.	Threatened abortion with dilated cervical os and/or rupture of membranes (ROM). Pregnancy loss is unavoidable. Treat with surgical evacuation + admin of RhoGAM if mother is Rh (-).	A documented pregnancy that spontaneously aborts all products of conception (POCs). Must do pathology report of POCs. Pt may require additional curettage if POCs are not completely evacuated. RhoGAM to Rh (-) women.	Cramping, bleeding, dilated cervix with passage of tissue into vagina or endocervical canal. Curettage often required. Rh (-) pt requires RhoGAM. Hemodynamic stabilization may be required.	POCs do not get fully expelled. No uterine growth, no fetal heart tones, cessation of pregnancy symptoms. Evacuation of uterus after confirmed fetal death. DIC is rare complication. RhoGAM is Rh (-)	≥2 consecutive or total of 3 spontaneous abortions. May be d/t chromosomal anomaly. Treat with cervical (13-16wk) circlage until labor or ROM occurs (36-37wk).

Disproportionate Fetal Growth

IUGR

- Diagnosed when fetal weight is <5-10 percentile for gestational age or <2,500g (5lb, 8oz.)
- Early sonogram is required if accurate dates are not known

Symmetric IUGR:

Fetal Causes:

- ⇓ growth potential
- US shows a ⇓ in all measurements

Etiology:

- TORCH infection
- Aneuploidy
- Structural anomalies such as: cardiac, neurological.

Ultrasound:

- ⇓ in all measurements (symmetric)

Diagnosing:

- Karyotype
- Screen for fetal infection
- Detailed sonogram

Asymmetric IUGR:

1. Maternal Causes:

- ⇓ placental perfusion

Etiology:

- Hypertension
- Malnutrition
- ETOH, tobacco, illicit drugs

Ultrasound:

- ⇓ abdomen measurements with normal head measurements

Diagnosing:

- Serial sonograms
- NST
- AFI (often ⇓, especially with severe uteroplacental insufficiency).
- Biophysical profile

2. Placental Causes:

- ⇓ placental perfusion

Etiology:

- Infarction
- Twin-twin transfusion
- Abruption
- Velamentous cord insertion

Ultrasound:

- ⇓ abdomen measurements with normal head measurements

Diagnosing:

- Serial sonograms
- NST
- AFI (often ⇓, especially with severe uteroplacental insufficiency).
- Biophysical profile

Macrosomia

- Indicated by fetal weight >90-95 percentile for gestation age or birth weight of 4000-4500g.

Risk Factors:

- Gestational Diabetes Mellitus
- Prolonged gestation
- Obesity
- Increased pregnancy weight gain
- Multiparity
- Male fetus

Complications:

Maternal → injury during birth, post-partum hemorrhage, emergency C-section

Fetus → shoulder dystocia, birth injury, asphyxia

Neonate → hypoglycemia, Erb's palsy

Management:

Elective C-section → in diabetic if >4500g, or >5000g in non-diabetic mother.

Fetal Assessment and Intrapartum Surveillance

The following must be assessed and observed closely during pregnancy:

- Fetal growth
- Fetal well-being
- Tests of fetal maturity
- Intrapartum fetal assessment
- Isoimmunization
- Genetic testing

1. Fetal Growth

- US is most reliable tool for growth assessment
- Measure by fundal height, a deviation from expected fundal height during weeks 18-36 require repeat measurement and/or US
- Early pregnancy: Gestational sac + crown-rump length correlate quite well with gestational age
- Later pregnancy: 4 measurements done due to wide deviation of normal range: 1- Biparietal diameter of skull 2- Abdominal circumference 3- Femur length 4- Cerebellar diameter

2. Fetal well-being

- ≥ 4 movements per hr usually indicates fetal well-being

Non-stress test (NST):

- Measures response of fetal heart rate to movement
- Normal NST when FHR ⇑ by 15 beats per minute for 15 sec following fetal movement (2 of these accelerations within 20min considered to be normal)
- A non-reactive NST requires further assessment
- NST has a high false-positive rate, thus a repeat within 24hr is a good idea

Biophysical profile:

Measures:

- Fetal breathing
- Fetal breathing
- Fetal tone

- Qualitative amniotic fluid volume
- Reactive fetal heart rate
- All measures from 0-2, a score of 8-10 is normal, score of 6 requires further evaluation, score of ≤ 4 usually requires immediate intervention

3. Tests of fetal maturity

- Since the respiratory system is last thing to develop in the fetus, decisions regarding delivery are usually based on the maturity of this system
- Surfactant is still low all the way up to 33 weeks
- Lack of surfactant causes neonatal respiratory distress syndrome (RDS)

Testing fetal lung maturity:

- Lecithin : sphingomyelin ratio >2.0 is the safe ratio and suggests lung maturity.
- Phosphatidylglycerol appears late in pregnancy and its presence indicates fetal lung maturity.

NOTE: Sphingomyelin remains constant throughout pregnancy, while lecithin only elevates when the fetal lungs become mature

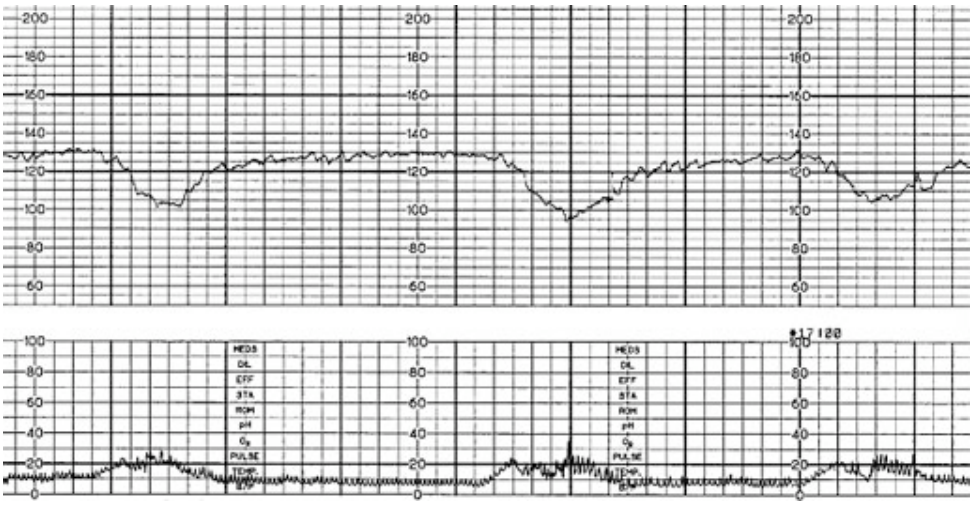
4. Intrapartum fetal assessment

FHR monitoring:

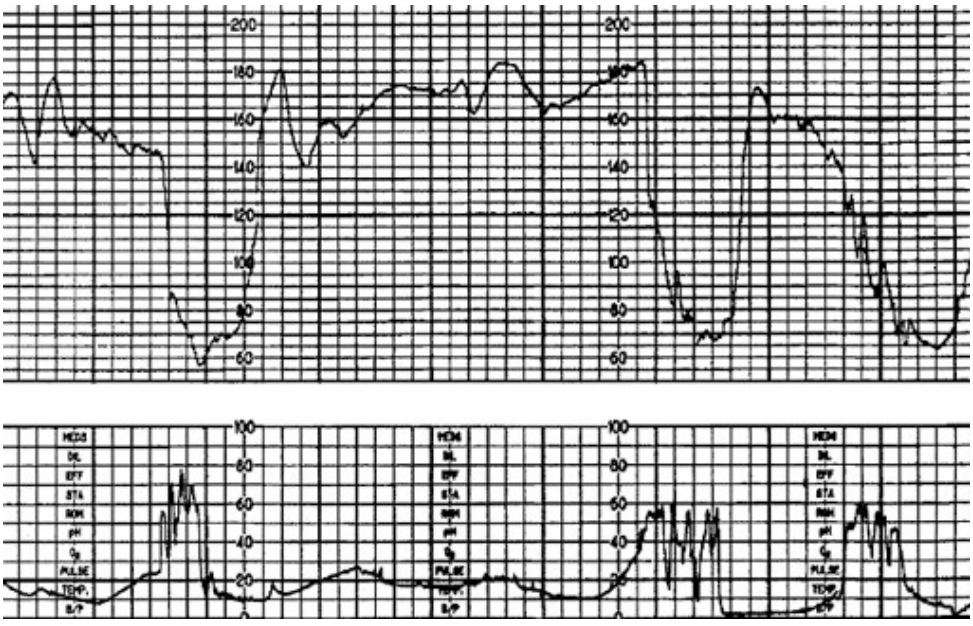
- Normal FHR is 120-160 BPM
- FHR >160 for ≥ 10 min considered tachycardia (MCC is maternal fever)
- FHR <120 for ≥ 10 min considered bradycardia (congenital heart block, fetal anoxia, maternal treatment with β-blockers
- FHR variability is a reliable indicator of fetal well-being and is suggestive of sufficient oxygenation of fetus.
- Decreased FHR variability is associated with fetal hypoxia, fetal tachycardia, depressants, prolonged uterine contractions

Accelerations:

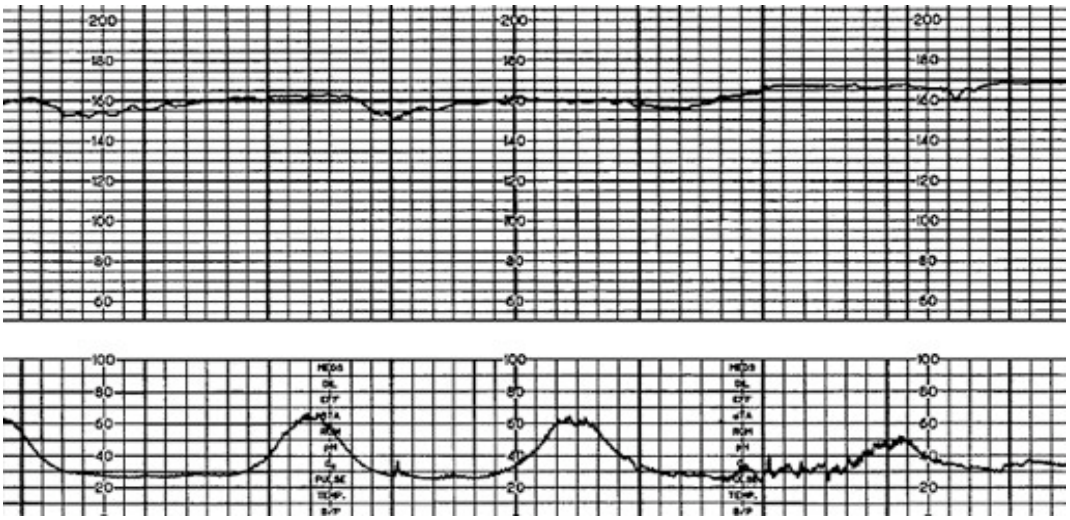
Early decelerations - ⇓ FHR mirroring contraction, due to pressure on fetal head (vagal nerve stimulation), is physiologic and not harmful to fetus.



Variable decelerations – don't always coincide with uterine contraction, characterized by rapid dip in HR followed by rapid return to baseline. Reflex-mediated and due to umbilical cord compression



Late decelerations – begin after contraction has started, dips after peak of contraction, and returns to baseline after contraction is over. Viewed as dangerous and is associated with placental insufficiency.



Causes:

- Placental abruption
- Pregnancy induced hypertension
- Maternal diabetes
- Maternal anemia
- Maternal sepsis
- Post-term pregnancy
- Hyperstimulation of uterus

** Repetitive late decelerations require intervention

5. Isoimmunization

Following exposure to fetal red blood cell antigens, the mother can develop maternal immunoglobulin antibodies (IgG). This can occur during birth or during the pregnancy depending on certain problems throughout the pregnancy where blood is mixed.

- Most commonly occurs when mother is Rh (-) and fetus is Rh (+)
- IgG crosses placenta and can affect the fetus
- A significant transfer of antibodies can result in hydrops fetalis
- Fetal ascites and edema occur (liver makes too many RBC's at the expense of proteins, causing a ⇓ in oncotic pressure, resulting in fetal ascites and/or edema.
- A maternal IgG titer ≥ 1.16 is sufficiently high to hurt the fetus

- The Kleihauer-Betke test helps to identify fetal RBC's in maternal blood.

Treatment/Avoidance:

- Give RhoGAM within 72hr of delivery to prevent an antibody response in the mother.
- Given at 28 weeks as well, can further reduce risks by 0.2%
- Important to test father for Rh status, if he is Rh (+), administer RhoGAM both at 28 weeks of gestation + at 72hr after delivery.

Other times maternal-fetal blood can mix:

- During amniocentesis
- After an abortion
- After an ectopic pregnancy treatment

6. Genetic Testing

- 50%-60% of spontaneous abortions due to chromosomal abnormalities
- 5% of stillbirths are due to chromosomal abnormalities
- 2%-3% of couples with multiple miscarriages are caused by chromosomal abnormalities
- 0.6% of live births have chromosomal abnormalities

When to do prenatal genetic testing?

- Advanced maternal age (most common) – Down's syndrome is increased 10-fold when mother is 35-45yr.
- Amniocentesis should be offered to all mothers with AMA
- When there is a prior child with chromosomal abnormalities
- When parent(s) have known chromosomal abnormalities
- When there has been an abnormal screening test

Labor and Delivery

1. Initial Presentation

- 85% of patients undergo spontaneous labor and delivery between 37 and 42 weeks
- Regular contractions every 5 minutes for at least 1hr, rupture of membranes, significant bleeding, and/or ⇓ fetal movement are all reasons for visiting the hospital

Initial Exam Upon Arrival:

- Auscultation of fetal heart tones
- Determination of fetal life (Leopold maneuver)
- Vaginal examination

2. Stages of Labor

Labor is divided into 3 stages:

Stage 1-

- Interval between the onset of labor and full cervical dilation (10cm)

This stage further subdivided into two phases:

Latent phase = cervical effacement and early dilation

Active phase = rapid cervical dilation occurs when approximately at 3-4cm

Stage 2-

- Is the interval between complete dilation and the delivery of the infant

Stage 3-

- Is the interval between delivery of the infant and delivery of the placenta

** There is a 4^{th} stage, which lasts 2hr and is the period immediately after delivery of the placenta, whereby there are many physiological changes

3. Management of Labor

During first stage:

- Continuous FHR monitoring
- Continuous monitoring of uterine activity with external tocodynamometer and intrauterine pressure catheter
- Analgesia and/or anesthetic given, most commonly when reaching the active stage of labor.

During second stage:

- At this stage maternal pushing can accelerate delivery
- Episiotomy done at this stage (if performed at all)
- Bulb and suction after delivery of the head
- Shoulders delivered after head
- Umbilical blood sent for ABO and Rh testing

During third stage:

- Delivery of placenta

During fourth stage:

- Evaluation of cervix, vagina, vulva, perineum, and urethral area for lacerations and/or traumatic injury

4. Abnormal Labor

Dystocia = Difficult Labor

Dystocia is detected by evaluation three criteria, they are:

1. **Power** – referring to the strength, duration, and frequency of contractions.

- For cervical dilation to occur, there must be more than 3 contraction in 10 minutes

2. **Passenger** – referring to fetal weight + fetal lie + presentation + position

3. **Passage** –

- Pelvic diameter
- Ability of pelvic to accommodate passage of baby
- Distended organs or masses can hinder passage

Labor stage	Definition	Duration	Abnormalities
Stage 1 – Latent Phase (Effacement)	**Begins with onset of regular uterine contractions.** **Ends with acceleration of cervical dilation.**	**<20 hours in primipara** **<14 hours in multipara**	**Prolonged latent phase:** **Cervix dilated <3cm,** **No cervical change in 20hr (primi) or 14hr (multi)** **MCC is analgesia** **Manage with rest and sedation**
Stage 1 – Active phase (Dilation)	**Cervical preparation for dilation.** **Begins with acceleration of cervical dilation.** **Ends at 10cm dilation.** **Rapid cervical dilation**	**>1.2cm/hr (primipara).** **>1.5cm/hr (multipara)**	**Active Phase prolong or arrest:** **Cervix dilated ≥ 3cm** **Prolongation: cervical dilation of <1.2cm/hr (primi) or <1.5cm/hr (multi)** **Cause: passenger abnormality** **Mgmt:** **If hypotonic contraction: IV oxytocin** **If hypertonic contraction: morphine sedation** **If adequate contractions do emergency C-**

			section
Stage 2 – Descent	**Begins when 10cm dilated.** **Ends when baby is delivered**	**<2hr if primi** **<1 hr if multi** **Additional hour if given epidural**	**Second stage arrest:** **Failure to deliver within 2hr(P) or 1hr(M)** **Cause: abnormality with passenger, pelvis, or power** **Mgmt:** **Fetal head not engaged (do emergency C-section)** **Fetal head engaged (forceps or vacuum extraction)**
Stage 3 – Expulsion	**Begins with baby delivery and ends with placental delivery**	**<30 minutes**	**Prolonged stage if placenta takes longer than 30 minutes to deliver.** **Mgmt: IV oxytocin, manual removal.** **Hysterectomy if these fail.**

Prolongation Disorders

Dystocia can be divided into prolonged latent phase and prolonged active phase

1. Prolonged latent phase
- Latent phase >20hr in primigravid or >14hr in multigravid
- May be due to ineffective uterine contractions, disproportion between pelvis and fetus, and even excessive anesthesia

2. Prolonged active phase
- Considered prolonged when it lasts >12hr and/or the rate of cervical dilation is <1.2cm/hr in a primigravid or <1.5cm/hr in multigravid.
- May be due to excessive anesthesia, ineffective contractions, disproportion between pelvis and fetus, fetal malposition, rupture of membranes before onset of active labor
- This can increase the risk of intrauterine infection and increased need for C-section

Arrest Disorders

An arrest disorder occurs when cervical dilation ceases during the active phase for ≥ 2hr. Most commonly due to either cephalopelvic disproportion or ineffective contraction of the uterus.

Management of Abnormal Labor

Labor induction = stimulation of uterine contractions before spontaneous onset of labor

Augmentation of labor = stimulation of uterine contractions that began spontaneously but have since become infrequent, weak, or both

Induction trial → should be attempted only if cervix is prepared or "ripe". Cervical ripening via prostaglandin E2 gel.

Indications for induction:

- Suspected fetal compromise
- Fetal death
- Pregnancy induced hypertension
- Past date pregnancy

- Maternal medical complication
- Premature rupture of membranes (PROM)

Contraindications for induction:

- Placenta previa
- Active genital herpes
- Cord presentation
- Abnormal fetal lie

Risks of prolonged labor:

- Infection
- Exhaustion
- Lacerations
- Uterine atony with hemorrhage

Postpartum Hemorrhage

Uterine Atony:

- The most common cause of excessive postpartum bleeding

Management:

- Uterine massage and uterotonic agents (oxytocin, methylergonovine)

Laceration: Management with surgical repair

Retained Placenta:

- Treatment involves manual removal or uterine curettage
- Placenta accreta/increta/percreta is the diagnosis if the exam shows placental villi infiltration
- Hysterectomy may be needed to control intractable bleeding

DIC: Most commonly associated with placental abruption, severe preeclampsia, amniotic fluid embolism, or prolonged retention of a dead fetus.

Uterine Inversion:

- Suspect if a beefy-appearing bleeding mass in the vagina and failure to palpate the uterus.
- Manage with uterine replacement followed by IV oxytocin

Urinary Retention:

- May occur with hypotonic bladder
- Residual volume >250ml requires bethanecol
- If bethanecol fails manage with urinary catheterization for 2-3 days

<u>Premature Rupture of Membranes (PROM)</u>

- Rupture of chorioamniotic membrane before the onset of labor
- Occurs in 10%-15% of all pregnancies
- Labor usually follows in 90% of patients within 24hrs
- PROM at ≤ 26 wks of gestation is associated with pulmonary hypoplasia

Diagnosing:

- Perform vaginal exam with testing of non-bloody fluid
- Nitrazine test: tests pH
- Fern test: amniotic fluid placed on slide, looking for branching fern leaf pattern when slide dries
- US confirms diagnosis: finds oligohydramnios

Treatment:

- If an intrauterine infection is suspected, give broad-spectrum antibiotics, otherwise treat as a preterm labor.

Postpartum Contraception

Breastfeeding:

- Breastfeeding provides contraception for 3 months because of temporary anovulation

Combined estrogen-progestin formulations:

- OCP, patch, or vaginal ring
- Not started until 3 weeks postpartum to prevent hypercaogulable state and risk DVT
- Not used in breastfeeding women because they diminish lactation

Diaphragm and/or IUD placement:

- Deferred until 6 weeks post-partum

Progestin contraception:

- Mini-pill, Depo-Provera
- Can be safely used while breastfeeding and started immediately after pregnancy

Post-partum Fever

Postpartum Day #	Most likely diagnosis
0	**Atelectasis**
1	**UTI**
2-3	**Endometritis**
4-5	**Wound infection**
5-6	**Septic thrombophlebitis**
7-21	**Infectious mastitis**

Perinatal Infections

1. Group B β-Hemolytic Streptococci (GBS)

- 30% of women have asymptomatic vaginal colonization with GBS
- Vertical transmission can cause pneumonia and sepsis of the neonate within hours of birth
- Mortality rate is as high as 50%

Treatment:

- Intrapartum IV penicillin G
- In patient with PCN allergy, use IV clindamycin, erythromycin, or cefazolin

When are antibiotics given?

1. When GBS (+) urine, cervical, or vaginal culture at any time in pregnancy.
2. Presence of high-risk factors such as:
   - Prolonged PROM
   - Preterm delivery
   - Presence of maternal fever
   - Any previous baby who experienced GBS sepsis

When are antibiotics not given?

1. With planned C-section, when there is no rupture of membranes.
2. Culture (-) in this pregnancy but with (+) culture in previous pregnancy

2. Toxoplasmosis

- Caused by Toxoplasma gondii parasite
- Is most commonly caused by the handling of cat feces and/or litter during pregnancy
- May be caused by drinking raw goat milk or eating raw meat
- Vertical transmission will only occur with primary infection of the mother
- Most serious infection results from infection in the first trimester

Symptoms:

- Most common presentation is a mononucleosis-type syndrome + the presence of a cat in the household
- On US, there may be intrauterine growth retardation

Prevention:

- Avoid handling cat feces, raw goat milk, and undercooked meat
- If infected, mother should take spiramycin to prevent vertical transmission

Treatment:

- After serologic confirmation, give pyrimethamine and sulfadiazine.

Classic congenital triad:

1. **Hydrocephalus**
2. **Intracranial calcifications**
3. **Chorioretinitis**

3. Varicella

- Primary varicella infection in mother causes transplacental infection 25%-40% of the time.
- Greatest risk to fetus is posed if a rash appears in the mother from 5 days antepartum and 2 days postpartum.
- A neonatal infection presents with limb hypoplasia, microcephaly, microphthalmia, chorioretinitis, cataracts, and "zigzag" skin lesions

Preventing Varicella infection:

- Non-pregnant women should receive vaccination
- Post-exposure prophylaxis within 96hrs of exposure with VZV immunoglobulin

Treatment:

- Maternal varicella – anti-varicella antibodies to mother and neonate
- Congenital varicella – anti-varicella antibodies + IV acyclovir to neonate

4. Rubella

- Vertical transmission occurs up to 70%-90% of the time with primary infection
- Neonate may present with congenital deafness, PDA, cataracts, mental retardation, hepatosplenomegaly, thrombocytopenia, and a blueberry muffin type rash.
- Adverse effects occur within first 10 weeks

Prevention:

- Perform a first trimester screening
- Have mother avoid any infected individuals
- Immunize seronegative women after delivery

** There is no post-exposure prophylaxis available

RUBELLA:

"Blueberry Muffin" rash

Deafness

PDA

Cataracts

Retardation

Hepatosplenomegaly

Thrombocytopenia

5. Cytomegalovirus (CMV)

- Is the most common congenital virus in the USA.
- CMV is the most common cause of sensorineural deafness in children
- CMV is spread by body fluid secretions
- Most infections produce a mononucleosis-like syndrome
- Approximately 10% of infants are symptomatic at birth

Symptoms in neonate:

- IUGR
- Prematurity
- Microcephaly
- Jaundice
- Petechiae
- Hepatosplenomegaly
- Periventricular calcifications
- Chorioretinitis
- Pneumonitis

CMV

- IUGR
- Prematurity
- Microcephaly
- Jaundice
- Hepatosplenomegaly
- Petechiae
- Periventricular calcifications
- Chorioretinitis
- Pneumonitis

Diagnosing CMV:

- Viral culture within 2 weeks of birth
- PCR

Prevention:

- Universal precautions with avoidance of body fluids
- Avoid transfusion with CMV-positive blood

Treatment:

- Ganciclovir (prevents hearing loss but does not cure infection)

6. **Herpes Simplex Virus (HSV)**

- Contact with lesions is the most common cause of transmission
- Greatest risk of infection in the 3rd trimester
- 50% risk of transplacental infection with primary infection

A neonatal infection acquired during delivery has a 50% mortality rate.

Symptoms:

- Fever + malaise + diffuse genital lesions
- If infant survives, they may develop meningoencephalitis, mental retardation, pneumonia, hepatosplenomegaly, jaundice, and petechiae.

Diagnosis:

- **(+)** HSV culture from vesicle fluid or ulcer or HSV PCR of maternal blood

Prevention

- A C-section is mandatory if mother has active genital HSV lesions at time of labor.
- It is contraindicated to use fetal scalp electrodes for monitoring; they increase risk of HSV transmission.
- Avoid standard precautions such as sex with outbreak

Treatment:

- Acyclovir for primary infection during pregnancy

7. **HIV**

- The major route of vertical transmission is contact with infected genital secretions at time of vaginal delivery.
- Without treatment vertical transmission rate is 25%-30%
- Elective C-section is most beneficial in women with low CD4+ counts and high RNA viral loads (>1000).
- All neonates of HIV + mothers will test positive due to the transplacental passage of IgG

Prevention and Treatment:

- Triple-drug therapy starting at 14 weeks and continuing throughout the pregnancy.
- IV intrapartum zidovudine (AZT)
- Give infant oral zidovudine for 6 weeks post-delivery

- Prophylax infant for PCP with TMP-SMX after the 6 weeks of zidovudine therapy
- Schedule a C-section at 38 weeks unless the viral load is <1000 viral copies/ml
- Mother cannot breastfeed because HIV passes through breast milk
- Avoid all invasive procedures such as artificial ROM and fetal scalp electrodes

8. Syphilis

- Transplacental infection is more likely in primary and secondary infection, and less likely in tertiary infection

Early-acquired (1st trimester) congenital syphilis includes the following symptoms:

- Hydrops fetalis
- Maculopapular or vesicular rash
- Large and swollen placenta
- Anemia, thrombocytopenia, and hepatosplenomegaly
- There is a 50% perinatal mortality rate

Late-acquired congenital syphilis is diagnosed after 2 years of age, includes the following:

- Hutchinson teeth
- "Saber" shins
- "Saddle" nose
- Deafness (CN8 palsy)
- "Mulberry" molars

A C-section cannot prevent vertical transmission of syphilis. It will be transmitted through the placenta before birth.

Diagnosis:

- VDRL or RPR screen in first trimester
- Confirm a positive test with FTA-ABS or MHA-TP

Treatment:

- Benzathine penicillin IM (1 time for infected mothers)
- If PCN allergy: Desensitization followed by full dose benzathing PCN

After any (+) syphilis screen, FTA-ABS or MHA-TP is done as confirmatory tests.

9. **Hepatitis** B

- Neonatal infection from primary infection in the 3rd trimester or ingestion of infected genital secretions
- 80% of infected neonates will develop chronic hepatitis

Prevention:

- Hep B infection is not an indication for C-section
- Avoid invasive procedures
- After neonate receives immunization and hep B immunoglobulin, can be breastfed

Immunization:

- HBsAg-negative – give active immunization during pregnancy
- Postexposure prophylaxis for the mother – HBIG

Treatment:

- Hepatitis immunization + HBIG in neonate
- Chronic HBV can be treated with interferon or lamivudine

Contraindications to breastfeeding

- HIV
- Active TB
- Hep B (before infant is immunized)
- HSV
- Drug use/medications
- Drugs of abuse (except cigarettes, alcohol)
- Cytotoxic medications
- Conditions of infant that CI breast milk
- HTLV-1

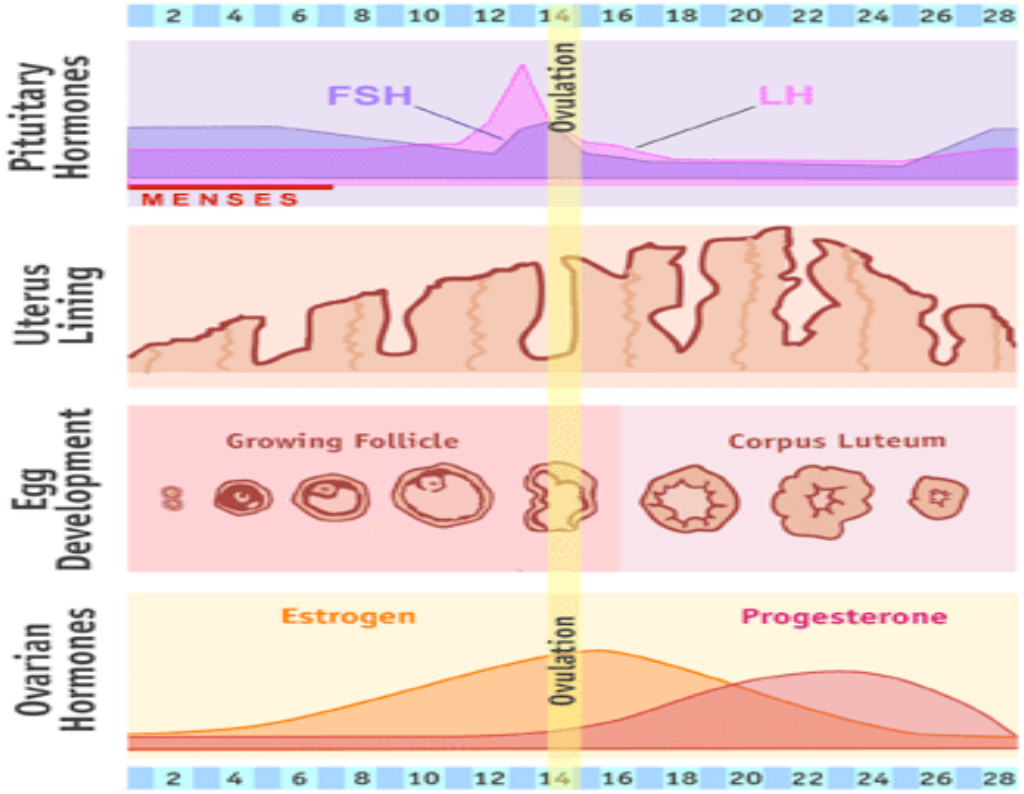
Chapter 3

Gynecology

Benign Gynecology

The Menstrual Cycle:

- At birth, there are approximately 1 million primordial follicles in the ovary, each with an oocyte that is arrested in the prophase stage of meiosis.
- The process of ovulation signals the onset of puberty, which signals follicular maturation.
- Ovulation occurs, and the dominant follicle becomes the corpus luteum, which secretes progesterone to prepare the endometrium for possible implantation.
- If the ovum is not fertilized, the corpus luteum undergoes involution, which causes menstruation.
- The cycle is regulated by hypothalamic gonadotropin-releasing hormone, pituitary release of follicle-stimulating hormones (FSH), luteinizing hormones (LH), and the ovarian sex steroids estradiol and progesterone.
- An ⇑ or ⇓ in any of these hormones can cause dysfunction of the normal menstrual cycle.



This graph demonstrates the process of all three phases of the menstrual cycle, which hormones are in play, the development of the egg, and the days that correspond to all events

Follicular/Proliferative Phase	Ovulatory Phase	Luteal/Secretory Phase
Day 1-13	Day 13-17	Day 15 – Day 1 of menses
Estradiol-induced negative feedback on FSH and positive feedback on LH in anterior pituitary leads to LH surge on days 11-13.	Dominant follicle secretion of estradiol leads to positive feedback to anterior pituitary FSH and LH, ovulation will occur within 30-36hrs after LH surge, small FSH surge also occurs at time of LH surge.	Marked by change from estradiol to progesterone predominance, corpus luteal progesterone acts on hypothalamus, causing negative feedback on FSH and LH, resulting in decreased basal levels prior to next cycle, if fertilization and implantation do not occur there will be a rapid decrease in progesterone.

Contraception:

Oral contraceptives that combine progestin and estrogen

- Estrogen suppresses FSH, which prevents selection and maturation of a dominant follicle.
- Progestin works by suppressing LH, which inhibits ovulation. It also thickens cervical mucosa, which further prevents the ability of semen to pass into the uterus.
- Together, they inhibit pregnancy by thinning the endometrial lining and resulting in light or misses menses.

Phasic vs. Monophasic Pills:

- Phasic pills vary the ratio of estrogen and progestin. This decreases the dose of hormone given but it increases the risk of breakthrough bleeding
- Monophasic pills deliver a constant dose of estrogen and progestin throughout the month

** Following cessation of OCP's, fertilization usually resumes immediately. A small percentage of users will experience a period of infertility, known as "postpill amenorrhea"

Advantages and Disadvantages of OCP's

ADVANTAGES	DISADVANTAGES
• Reliable with failure rate of <1% • Protective against ovarian and endometrial cancer • Decreases incidence of PID and ectopic pregnancies • Makes menses more predictable and less painful/lighter.	• Must be taken daily close to the same time • No STD protection • Up to 30% breakthrough bleeding • May cause depression, weight gain, hypertension, acne, bloating, and weight gain

Alternatives to OCP's

- Progestin-only pills → "mini-pills"
- Depo-Provera → IM injection lasts for 14 weeks
- Norplant → subcutaneous implant lasts up to 5 years
- Intrauterine device → device left in place for several years
- Emergency Contraception → progestin/estrogen taken within 72hr

Pap Smear:

- Begin within 3 years of onset of sexual activity OR age 18 years (whichever comes first).
- When 3 annual pap smears in a row are negative, can do every 2 years until age 70.
- Patients with 1 sexual partner, 3 consecutive normal pap smears, and onset of sexual activity after age 25 can be screened less frequently.

Pap Smear Classifications:

1. **Intermediate smear**: Atypical squamous cells of undetermined significance (ASCUS)

2. **Abnormal smears:**

- Low grade squamous intraepithelial lesions (LSIL): HPV, mild dysplasia, or carcinoma in situ 1 (CIS1)
- High grade squamous intraepithelial lesions (HSIL): moderate dysplasia, severe dysplasia, CIS, CIN 2 or CIN 3
- Cancer: invasive cancers

Workup of an abnormal Pap

Step in the Workup	When is this step warranted	Next Step in management
Repeated PAP	**After first ASCUS find**	**Repeat every 4-6 months until there are 2 consecutive negative Pap smears**
HPV DNA testing	**After first ASCUS is found**	**If HPV 16 or 18 identified perform colposcopy**
Colposcopy and ectocervical biopsy	**Abnormal Pap smear Two ASCUS Pap smears**	**Colposcopy, Abnormal lesions sent for cytology**
Endocervical curettage (ECC)	**All non-pregnant patients showing an abnormal Pap smears**	**Non-pregnant pts undergoing colposcopy for abnormal Pap require ECC to rule out endocervical lesions**
Cone Biopsy	**When Pap smear is worse than biopsy suggested, When ECC is abnormal, With endocervical lesion, When biopsy shows microinvasive carcinoma of cervix**	

Management of abnormal histology

Step in Management	Condition warranting this step	Notes/Details
Observe and follow-up	**CIN1, CIN 2 or 3 after excision or ablation**	**Follow-up tests q4-6 months for 2 years**
Ablation	**CIN2 or CIN3**	**This is: cryotherapy, laser vaporization, electrofulguration**
Excisional procedures	**CIN2 or CIN3**	**LEEP and Cold-knife colonization**
Hysterectomy	**Recurrent CIN2/3**	

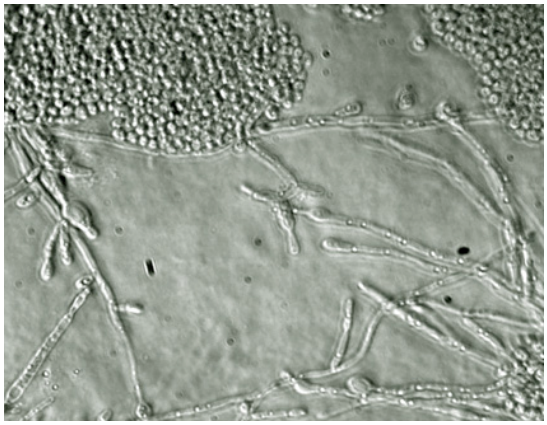
Vaginitis

- The most common presenting symptom is discharge
- Always rule out chemical or allergic causes
- 50% of cases are due to Gardnerella
- 25% due to Trichomonas
- 25% due to Candida

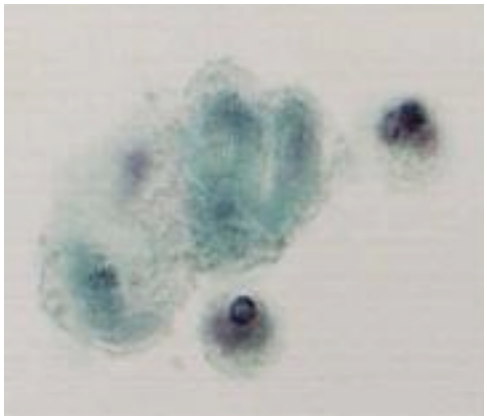
Differentiating between the different causes of vaginitis

	Candida	**Trichomonas**	**Gardnerella**
Vaginal pH	4-5	>6	>5
Odor	NONE	RANCID	"Fishy" on KOH
Discharge	Cottage Cheese-like	Green, frothy	Variable
Si/Sx	Itching, burning, swollen	Severe itching	Variable, none
Microscopy	Pseudohyphae	Motile Organisms	Clue cells
Treatment	Fluconazole	Metronidazole	Metronidazole

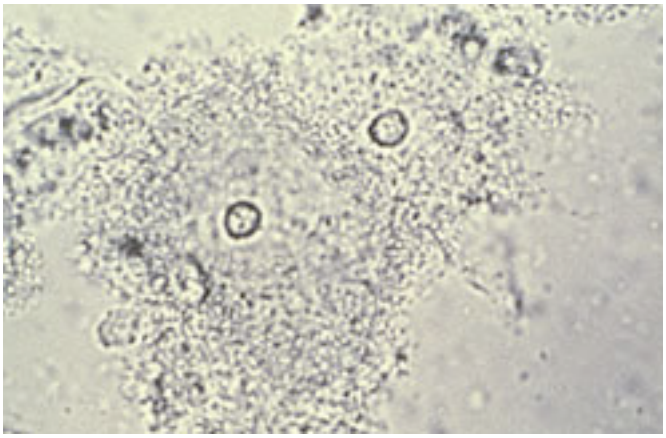
Candida Albicans – note the pseudohyphae



Trichomonas – Motile Organisms



Gardnerella – Clue Cells (large epithelial cells covered with small bacteria)



Endometriosis

- Endometrial tissue outside of the uterus, most commonly found in the ovaries.
- Affects approximately 1%-2% of women
- Approximately 50% of infertile women have endometriosis

Signs & Symptoms:

- The 3 D's → Dyspareunia, Dysmenorrhea, Dyschezia
- May also have pelvic pain, infertility, pain on rectovaginal exam.

Diagnosis:

- Visualization via laparoscopy or laparotomy with histological confirmation.

Treatments:

1. NSAID's first
2. OCP's
3. Testosterone (Danazol – A/E: hirsutism, acne)
4. GnRH analog (gives best result but causes menopausal symptoms within 3-6 months)

Recurrence after cessation of medical treatment is common, definitive treatment is hysterectomy.

Reproductive Endocrinology

Amenorrhea

Primary → woman has never menstruated

Secondary → menstrual aged woman who has not menstruated in 6 months

- Most common cause of amenorrhea is pregnancy
- Every evaluation must by excluding pregnancy (urine β-hCG)
- The most common cause of secondary amenorrhea is Asherman's syndrome (scarring of the uterine cavity after D&C)
- Hypothalamic deficiency is a cause of amenorrhea (excessive weight loss, excessive exercise, psychogenic, drug use)
- Pituitary dysfunction: from ⇓ hypothalamic pulsatile release of GnRH or ⇓ pituitary release of FSH or LH

Ovarian dysfunction:

- Follicles are exhausted of FSH and/or LH OR are resistant to stimulation

Causes:

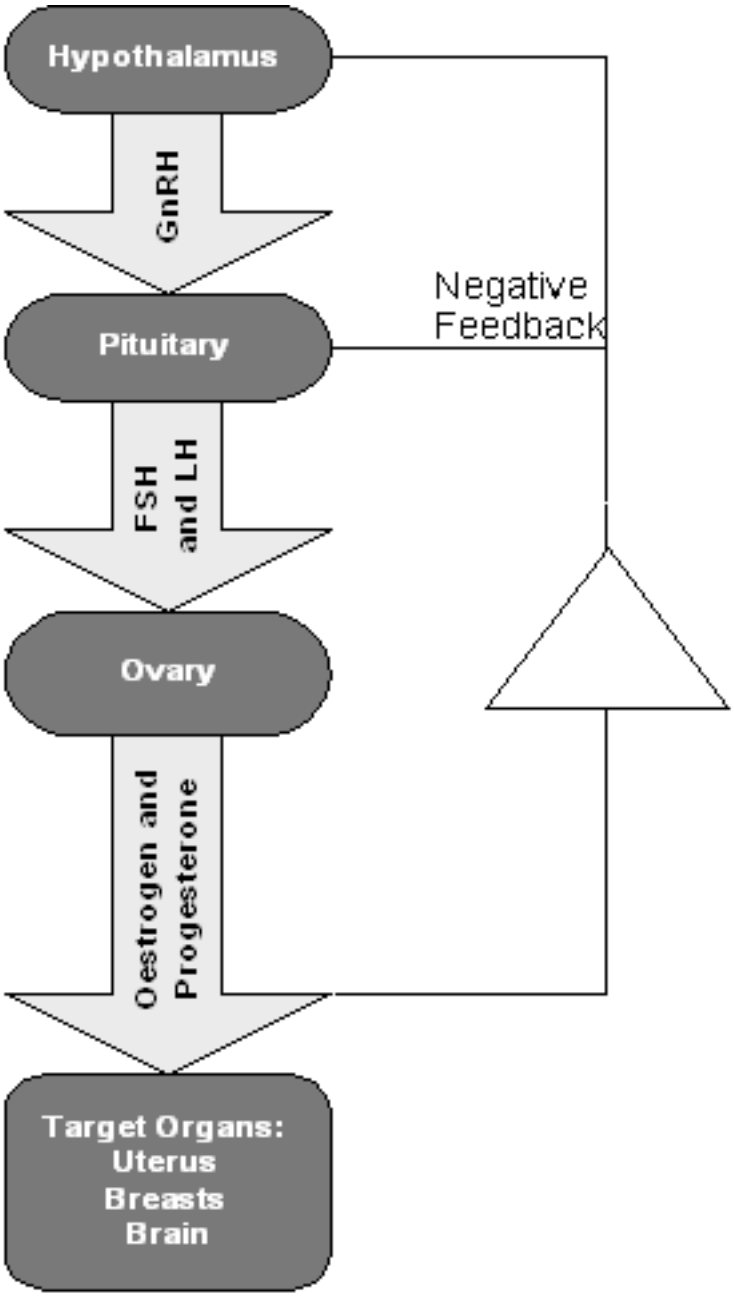
- Inherited disorders such as Turner's syndrome
- Premature menopause
- Autoimmune ovarian failure
- Chemotherapies

Treatment:

- If hypothalamic → treat underlying cause and induce ovulation with gonadotropins
- Tumors → Bromocriptine to shrink tumor and/or excision
- Genital tract obstruction → surgery
- Ovarian dysfunction → Exogenous estrogen replacement

How to differentiate between Menopause and Asherman's syndrome?

→ An ultrasound will help visualize the presence or absence of follicle. Asherman's syndrome will have follicles while menopause will lack the presence of follicles.



Hypothalamic Causes: Stress, Anxiety, Anorexia, And Excessive Exercise

Pituitary Causes: Adenoma

Ovarian Causes: Early menopause, resistant ovary syndrome

Endometrial Causes: Asherman's syndrome

Abnormal bleeding in a reproductive age woman

With the presence of abnormal bleeding, first thing is to rule out "PAD:

P→ pregnancy

A→ anatomical abnormalities

D→ dysfunctional uterine bleeding

Dysfunctional Uterine Bleeding (DUB):

- Anatomic menstruation without anatomic lesions of the uterus
- Is most commonly due to chronic estrogen stimulation
- Abnormal bleeding is defined as bleeding at intervals <21 days or >36 days, lasting >7 days, or blood volume loss >80ml

Diagnosis:

- R/O anatomic causes (fibroids, cervical or vaginal lesions, infections, cervical and/or endometrial cancer)
- Evaluate factors that can affect the hypothalamus-pituitary axis (stress, excessive exercise, weight changes, systemic disease, coagulopathies, pregnancies)

Treatment:

- Give progesterone from day 14-25 of menstrual cycle
- Birth control pills are an alternative

Hirsutism and Virilization

Hirsutism → excessive body hair usually due to polycystic ovaries or adrenal hyperplasia

Virilization → masculinization, associated with marked ⇑ in testosterone, male pattern balding, voice deepening, clitoromegaly, breast involution.

Differentiating causes of Hirsutism and Virilization

Disease	Characteristics	Treatment
Polycystic Ovarian Disease	**The #1 cause of androgen excess and hirsutism.** Related to LH overproduction. Si/Sx: amenorrhea or oligomenorrhea, infertility, hirsutism, acne. Anemia Labs show: ⇑ LH/FSH, and ⇑ testosterone.	OCP's to ⇓ LH production (via feedback changes). Weight loss. Clomiphene may be prescribed to induce ovulation.
Sertoli-Leydig Cell Tumor	Is an ovarian tumor that secretes testosterone (women 20-40). Si/Sx: rapid onset of hirsutism, acne, amenorrhea, virilization. Labs: ⇓LH/FSH, ⇑⇑⇑Testosterone.	Remove affected ovary
Congenital Adrenal Hyperplasia	MC due to 21-α hydroxylase defect. AR pattern. Severe will cause virilization of newborn, while milder forms can present at puberty or later. ⇑ LH/FSH, ⇑ DHEA (DHEA helps determine adrenal source).	Glucocorticoids can suppress adrenal androgen production.

Menopause

- The cessation of menses occurs on average at 51yr of age.
- Cycles no longer associated with pre-menstrual symptoms, no longer regular or predictable.

Signs and symptoms:

- Acute onset of hot-flashes/sweating that cease acutely (within 3-5 minutes)
- Mood disturbances
- Sleep disturbances
- Vaginal dryness (leads to dyspareunia)
- Osteoporosis

Diagnosis:

- Irregular menstrual cycles are most obvious for diagnosis
- Presence of hot flashes suggest menopause
- ⇑ levels of FSH (>30mIU/ml)

Treatment:

- 1st line treatment is estrogen replacement therapy (continuous estrogen with cyclic progestin to allow withdrawal bleeding or daily admin of both estrogen and progestin, which wont cause w/d bleeding).
- Raloxifene: a 2nd generation tamoxifen-like drug (mixed estrogen agonist/antagonist). Shown to prevent osteoporosis, decreases LDL, decreases risk of breast cancer.
- Calcium supplementation is not a substitute for estrogen replacement

Infertility

- Defined as 1yr of frequent, unprotected intercourse without pregnancy.
- 60% female cause, 40% male cause

How to approach infertility:

1st Step: Semen Analysis (volume, motility, # active sperm)

If this is found to be abnormal, attempt Intrauterine Insemination (IUI) or Intracytoplastic Sperm Injection (ICSI)

If semen analysis is normal, consider female factors.

2nd Step: Test cervical mucus to see if it softens (done around time of ovulation – known as "spinnbarkeit".

How to test:

- Patient comes in after intercourse
- With help of speculum get endocervical mucus
- Put mucus on a glass slide and cover with another slide
- Attempt to pull slides apart (should be able to separate at least 6cm before mucus breaks – which indicates soft and favorable mucus)
- If slide breaks too early, mucus is "hostile"
- Also want to check for "mucus ferning" on microscopy

How to treat hostile mucus: **Intra Uterine Insemination (IUI)**

- Perform by inserting a catheter into the cervix and inject sperm past the mucus plug

Another solution: Give estrogen early in the cycle (softens mucus)

Next: If Sperm is OK and Mucus is OK….

3rd Step: Check Ovulation

- Check basal body temperature (⇑ 1deg due to progesterone spike)
- A sign of pregnancy is the lack of body temperature rising
- Do an endometrial biopsy
- Day 22 is the highest day of serum progesterone
- LH surge – there are strips that can measure LH, if + patient will ovulate within 24-48hrs

If you find a problem with ovulation, give fertility drugs.

Treatment:

1st → CLOMID (fools the pituitary into secreting lots of FSH and LH)

OR

2nd → PERGANOL (concentrated amounts of FSH and LH from urine of menopausal women).

** Use CLOMID first, it has a lower rate of multiple gestation and lower risk of "ovarian hyperstimulation syndrome". With PERGANOL, there is a 20% multi-gestation risk.

If semen is normal…

If mucus is normal…

If ovulation is normal…

NEXT STEP: Look for tubal factors (blockage due to PID) – history of chronic pain, TOA, ectopic pregnancy, infertility

Perform: Hystero-Salpingogram to help diagnose a blockage

IF there is a tubal problem, there are 2 choices:

1. Tuboplasty (cuts out the obstruction)
2. In Vitro Fertilization (done in a lab, then put zygote into uterus)

In Vitro Fertilization (IVF):

- Puts 4 zygotes into the uterus
- Putting <4 = low pregnancy rate
- Putting >4 = ⇑ risk of multiple gestation

IVF has a 60% pregnancy rate

Last thing to do in the infertility workup is **Laparoscopy** (checking for endometriosis).

→ 20%-25% of those with unexplained infertility actually have endometriosis.

** If everything is normal, diagnosis is "unexplained infertility"

DATA shows that 50% of people with infertility will get pregnant within 4-5 years.

Androgen Insensitivity

- A diagnosis made when there is an absence of pubic or axillary hair.
- Karyotype reveals a male genotype (XY), and US reveals testicles.
- Testes produce normal levels of both male and female hormones, ie. Estrogen and testosterone.

Management:

Removal of testes before 20yr of age due to increased risk of testicular cancer. After removal patient will require life-long estrogen replacement.

Gonadal Dysgenesis (Turner's syndrome)

- Absence of secondary sexual characteristics
- Karyotype reveals the absence of one of the X chromosomes (45, X).
- Elevated FSH
- Lack of a second X chromosome leads to lack of ovarian follicle development, leading to "streak gonads".

Management:

Estrogen and progesterone replacement to help development of secondary sexual characteristics.

Urogynecology

Pelvic Relaxation*: Most commonly causes the following:*

1. *Uterine Prolapse*
2. *Cystocele (bladder prolapse into vagina)*
3. *Rectocele (rectal prolapse into vagina)*

Uterine Prolapse:

- Occurs when ligaments (suspensary) ligaments can no longer support it
- Most common cause is childbirth

Signs & Symptoms:

- Vaginal pressure sensation
- Vaginal fullness
- Low back pain

** Can cause cystocele and rectocele

Degrees of prolapse:

1st degree → prolapse is above introitus

2nd degree → goes to the introitus

3rd degree → goes past introitus

Cystocele:

- Bladder prolapse into the vagina due to excessive pelvic relaxation
- Severe cases can cause stress incontinence

2 types of incontinence:

1. Stress Incontinence → the most common type (caused by pelvic floor injuries)

2. Neurogenic Incontinence → urge/overflow incontinence (caused by ⇓ innervation and control of bladder function, resulting in involuntary bladder contraction (urges) or bladder atony (overflow).

Testing for stress incontinence:

1. Do a pelvic exam – if you see prolapse this suggests stress incontinence
2. Q-tip test – insert a Q-tip into urethra and have the pt cough. If it rotates >30degrees, pt has stress incontinence.

** If patient describes an "urge" and/or "frequency" for urination, it is suggestive of neurogenic incontinence

Treatments:

Stress Incontinence → 1st – attempt kegel exercises, 2nd – surgery (colporaphy)

Neurogenic → Anti-spasmotics, anti-cholinergics

Other causes of urinary incontinence:

Neuropathic → caused by head injury, spinal injury, or peripheral nerve injury. Treatment: catheter, either indwelling or intermittent

Anatomical → caused by a vesicovaginal fistula. Treatment: repair of defect.

Gynecologic Oncology

1. Endometrial cancer
2. Fibroids/Leiomyoma
3. Cervical cancer
4. Ovarian neoplasm's
5. Vulvar and vaginal cancer
6. Gestational trophoblastic neoplasia

Endometrial Cancer:

- Is the most common reproductive tract cancer with approximately 30,000-40,000 new cases each year.
- Is an "estrogen-dependent" cancer

Risk factors:

- Unopposed postmenopausal estrogen replacement therapy
- Menopause after 52yr of age
- Obesity
- Nulliparity
- PCOS
- Diabetes
- Chronic anovulation

Signs & Symptoms:

- Abnormal uterine bleeding, especially if woman is postmenopausal
- All women >35yr with abnormal bleeding requires an endometrial sampling for histologic examination.

Diagnosis:

- Pap smear not reliable
- Bimanual exam for masses, nodules, induration, and immobility
- Endometrial biopsy by endocervical curettage, D&C, hysteroscopy with direct biopsies.

Treatment:

- ***If simple or complex hyperplasia*** – give progesterone to reverse hyperplastic process promoted by estrogen (Provera for 10 days)

- ***Atypical hyperplasia*** – hysterectomy because of likelihood that invasive carcinoma will ensue
- ***Endometrial carcinoma*** → Total abdominal hysterectomy + bilateral salpingo-oophorectomy + lymph node dissection, adjuvant external-beam radiation, treatment for recurrence is high-dose progestin's.

Prognosis:

- Histologic grade is the most important factor in overall prognosis.
- Depth of myometrial invasion is 2nd most important factor in overall prognosis

G1 – highly differentiated

G2 – moderately differentiated

G3 – solid or completely undifferentiated

G1 tumor that doesn't invade the myometrium has a 95% 5-yr survival

G3 tumor with deep myometrial invasion has ~20% 5-yr survival

<u>Uterine Leiomyomas - Fibroids</u>

- Fibroids are benign tumors whose growth is related to the production of estrogen.
- Growth is often rapid perimenopausally
- 30% of cases warrant a hysterectomy

Signs & Symptoms:

- Menorrhagia
- Pelvic pain and pressure (misdiagnosed as dysmenorrheal)
- Enlarged, firm, asymmetric, non-tender uterus

Diagnosing Fibroids:

- US initially, confirm with tissue sample by either D&C or biopsy (especially in postmenopausal women).

Treatments:

- Mild symptoms only require re-assurance and observation
- Estrogen inhibitors such as GnRH agonists can shrink the uterus, which help to create a simpler surgical procedure.

- Surgery → myomectomy recommended in young patients who want to preserve fertility.
- Hysterectomy is definitive treatment that should be reserved for symptomatic women who have no desire for children and/or any more children.

Adenomyosis

- Abnormal location of endometrial glands and stroma within the myometrium of the uterine wall
- When symptomatic is causes dysmenorrheal and menorrhagia.
- Uterus feels soft, globular, tender, and symmetrical.
- Unlike leiomyomas, there is no change with high or low estrogen states.

Diagnosis	Leiomyoma	Adenomyosis
Symptoms	Secondary dysmenorrhea and menorrhagia. (+) or (–) symptoms of bladder, ureter, and/or rectal compression	Secondary dysmenorrhea and menorrhagia
Pelvic Exam	Asymmetrically enlarged, firm, NONTENDER uterus.	Symmetrically enlarged, soft, TENDER uterus. May be tender immediately before and during menses
Sonogram	Large intramural or subserosal myomas (saline infusion can help visualize this)	Diffusely enlarged uterus with cystic areas within myometrial wall
Hysteroscopy	Direct visualization of tumors	
Histology	Is definitive diagnosis	Is definitive diagnosis

Leiomyosarcoma

- A rare cancer that accounts for approximately 3% of uterine cancers

Signs & Symptoms:

Sarcoma → postmenopausal bleeding, pelvic pain, increasing vaginal discharge

Treatment:

- Hysterectomy with intraopertive LN biopsy
- Surgical staging is an important aspect
- Adjunctive therapies have minimal overall benefit

** Only 50% of patients survive 5 yr

Cervical Cancer

- The most important screening tool is the Pap smear
- The average age of diagnosis is between 45-50yr.
- The most common diagnosis is squamous cell carcinoma (85%), the other 15% are adenocarcinoma

Diagnosis:

Cervical Biopsy→ most commonly is SCC

Next step → metastatic workup (pelvic exam, CXR, IV pyelogram, cystoscopy, and sigmoidoscopy).

** Imaging studies not required (invasive cervical cancer is the only gynecological cancer that does not get staged clinically).

Management:

- Management is simple hysterectomy or modified radical hysterectomy + LN dissection. Survival <40% at 5yr.

- Adjuvant therapy such as radiation and chemo is given when any of the following conditions are present: Mets to LN's, tumor >4cm, + margins, local recurrence

Ovarian Neoplasms

Benign cysts:

- Functional growth resulting from failure of normal rupture of follicle
- Benign tumors are more common than malignant tumors
- Risk of malignancy increases with age

Signs & Symptoms:

- Pelvic pain/pressure
- Acute and severe pain when cyst ruptures

Confirm Diagnosis → With US

Benign Neoplasms

Neoplasm	Characteristics	Treatment
Epithelial Cell	Serous cystadenoma is the most common type. Usually benign (malignancy risk increases when bilateral). Others: mucinous, endometrioid, Brenner tumor (all rarely malignant). Diagnose: Clinical / CT or MRI	Excision
Germ Cell	Most common type is Teratoma (aka dermoid cyst). Almost never malignant. Contains tissues from all 3 germ layers. Unilateral, cystic, mobile, non-tender adnexal mass, often asymptomatic. Confirm diagnosis with US	Excision to prevent torsion or rupture of ovary
Stromal Cell	Are functional tumors secreting hormones. Granulosa tumor makes estrogen (gynecomastia, loss of body hair). Sertoli-Leydig makes androgens (virilization in females)	Excision

Malignant Tumors:

- Most commonly seen in women > 50yr
- OCP use is a protective factor
- Seen in higher frequency in women of low parity, ⇓ fertility, delayed childbearing
- Usually asymptomatic until metastasis has occurred
- Yearly pelvic exams are the most effective screening tools

Signs & Symptoms:

- Vague abdominal pains
- Vague pelvic pains
- Constipation, early satiety, abdominal distention, urinary frequency

Treatment:

- Debulking surgery + chemo/radiation

Malignant Neoplasms

Neoplasms	Characteristics	Treatment
Epithelial Cell	The cause of 90% of all ovarian malignancies. Serous cystadenocarcinoma is the most common type (often develops from a benign precursor).	Excision
Germ Cell	Is the most common ovarian cancer in women <20yr. Produces hCG or AFP, which are useful as tumor markers. Subtypes include: dysgerminomas and immature teratomas.	1st – Radiation 2nd – Chemotherapy >80% 5-yr survival
Stromal Cell	Granulosa cells make estrogen (endometriosis). Sertoli-Leydid cell tumor makes androgens	Total hysterectomy + oophorectomy.

Vulvar and Vaginal Cancers

Vulvar Intraepithelial Neoplasia (VIN):

VIN 1 & 2 → characterized by mild/moderate dysplasia, ⇑ risk of progressing to advanced stages and then carcinoma.

VIN 3 = carcinoma in situ

Signs & Symptoms:

- Pruritis
- Presence of raised lesions
- Irritation

Diagnosis:

- Biopsy required for a definitive diagnosis

Differential diagnosis:

- Malignant melanoma and Paget's disease

Treatment:

- For VIN 1 and 2 → local excision
- For VIN 3 → wide excision

Vulvar Cancer:

- 90% are squamous cell cancers
- Most often this presents in postmenopausal women

Signs & Symptoms:

- Pruritis (may present with or without an ulcerative lesion)

Treatment:

- Excision

** 5yr survival rate ranges from 70%-90% depending on LN involvement.

** With the presence of deep pelvic nodes, survival rate drops to 20%

Vaginal carcinoma in situ and carcinoma:

- 70% of patients with vaginal CIS have previous genital tract neoplasm

Treatment:

- Radiation
- Surgery reserved for extensive disease

Gestational Trophoblastic Disease (GTN)

- An abnormal proliferation of placental tissue that involves both the cytotrophoblast and/or syncytiotrophoblast.
- Can be both benign and malignant

Risk factors:

- Maternal age on low or high spectrum (<20yr and >35yr)
- Folate deficiency is a risk factor

Signs & Symptoms:

- Most common sign is a fundus that is larger than dates should show
- Bilateral cystic enlargement of the ovary
- Bleeding at <16 weeks gestation and passage of tissue from vagina is the most common symptom
- Hypertension
- Hyperthyroidism
- Hyperemesis gravidarum
- No fetal heart tones heard
- Most common site of metastasis is the lung

Diagnosing:

- US reveals homogenous intrauterine echoes without a gestational sac or fetal parts (looks like a "snowstorm")

Management:

- Get a baseline quantitative β-hCG
- Get a CXR to rule out lung METS
- D&C to evacuate the uterine contents
- Place the patient on OCP's so that there will be no confusion between a rising β-hCG titer from recurrent disease and normal pregnancy

BENIGN MOLE

Complete Mole	**Incomplete Mole**
Empty egg	Normal egg
46, XX (dizygotic ploidy)	69, XXY (triploidy)
Fetus is absent	Fetus is nonviable
20% become malignant	10% become malignant
No chemotherapy. Serial β-hCG until completely negative. Follow up for 1yr while on OCP's	No chemotherapy. Serial β-hCG until completely negative. Follow up for 1yr while on OCP's

MALIGNANT MOLES

Non-metastatic	Metastatic: Good Px	Metatstatic: Poor Px
Uterus only	Pelvis or lung	Brain or liver
100% cure	>95% cure	65% cure
Single-agent chemo until after β-hCG is negative for 3 weeks.	Single-agent chemo until after β-hCG is negative for 3 weeks.	Multiple-agent chemo until after β-hCG is negative for 3 weeks.
Follow-up for 1 yr on OCP	Follow-up for 1 yr on OCP	Follow-up for 5 yrs on OCP.

The Breast

Benign Breast Diseases:

- Fibroadenoma
- Fibrocystic disease
- Intraductal Papilloma
- Fat Necrosis
- Mastitis

Malignant Breast Diseases:

- Ductal carcinoma in situ (DCIS)
- Lobular carcinoma in situ (LCIS)
- Ductal carcinoma
- Lobular carcinoma
- Inflammatory breast cancer
- Paget's disease of the breast

Fibroadenoma

- Is the most commonly seen tumor in young women (20's)
- Fibroadenomas grow rapidly but have no increased risk for developing cancer
- Histology shows myxoid stroma and curvilinear/slit ducts

Treatment: Not required and will often resorb within several weeks (re-evaluate at 1 month)

Fibrocystic Disease

- Is the most common tumor in patients between 35-50 years of age.
- Arises in terminal ductal lobular units
- Often arises as multiple bilateral small lumps which are tender during the menstrual cycle

Intraductal Papilloma

- Presents commonly with serous bloody nipple discharge
- Multiple nodules in younger patients and solitary growth in perimenopausal patients

- There is an increased cancer risk with multiple papillomas, but no increased cancer risk with solitary pipillomas

Ductal Carcinoma in Situ (DCIS)

- Usually seen on mammography but not clinically palpable
- Is a premalignancy that will lead to invasive ductal cancer
- Histology shows haphazard cells along papillae, punched-out areas in ducts, with cells infiltrating open spaces.

Treatment:

- Mass excision ensuring clear margins.
- If margins are not clear must excise again with wider margins.
- Give post-op radiation to prevent recurrences.

Lobular Carcinoma in Situ (LCIS)

- LCIS can't be detected clinically but mammography is also a weak tool for diagnosis
- Not precancerous like DCIS but can be a marker for future invasive ductal cancer
- Mucinous cells are almost always present
- There is a "saw-tooth" and clover-leaf configurations in the ducts

Invasive Ductal Carcinoma (IDC)

- Is the most common breast cancer type, seen most commonly in the mid 30's – late 50's, and forms solid tumors
- Most important prognosis factor is the size of the tumor
- LN involvement is also an extremely important factor in prognosis
- There are many subtypes of IDC, such as mucinous and medullary
- Moderately differentiated IDC comes from cribriform or papillary intraductal originators
- Poorly differentiated IDC comes from intraductal comedo originator.

Invasive Lobular Carcinoma (ILC)

- 3%-5% of invasive cancer is lobular
- Most commonly seen from 45-55 yr of age
- Vague appearance on mammogram
- Growth pattern → single file growth pattern within a fibrous stroma

Treating invasive carcinomas:

- If tumor is <5cm perform lumpectomy + radiotherapy +/- adjuvant therapy +/- chemotherapy
- Sentinal node biopsy should be performed over an axillary node biopsy
- Always test for: **1.** Estrogen & Progesterone receptors, and **2**. HER2 protein
- The primary treatment for inflammatory, tumor size >5cm, and METS is systemic therapy

Inflammatory Carcinoma

- Classic symptoms are that of inflammation (warm, red, painful)
- Progresses rapidly and is almost widely metastatic at presentation
- Has a very poor prognosis

Paget's Disease of the Breast

- Very specific presentation of dermatitis + macular rash over the nipple and areola
- There is almost always an underlying ductal carcinoma

More on the breast:

- The most common cause of nipple discharge is intraductal papilloma
- The presence of discharge + palpable mass increases the likelihood of cancer is greater
- If discharge is unilateral, further workup is required
- If discharge is bloody, further workup is required
- If discharge is associated with a mass, further workup is required
- For bilateral, milky nipple discharge → do a workup for prolactinoma

With discharge:

- Next step is a mammogram to look for underlying masses and/or calcifications

- If mammography gives a definitive diagnosis, excision of duct is recommended
- For nipple discharge, never base diagnosis on cytology

Mastalgia:

- Is cyclical or noncyclical breast pain that isn't caused by lumps
- Treat with danazol (induces amenorrhea)

Gynecomastia:

- Enlargement of the male breast, both unilateral and bilateral
- No lobules
- Is caused by an imbalance in estrogens and androgens, most often occurring during puberty
- May be seen in hyperestrogen states (cirrhosis, drugs inhibiting estrogen breakdown → ETOH, marijuana, heroin)

Cancer Risks of Breast Cancers:

- #1 risk factor is gender (females make up 99% of breast cancers)
- In women, age is the #1 factor
- Menarche < 11yr is a risk factor for breast cancer
- Women who are nulliparous at >30yr have an increased risk
- 95% of breast cancer is NOT familial
- Having a first degree relative with a hx of breast cancer increases the risk of breast cancer
- Autosomal dominant conditions that have increased risk, such as BRCA-1, BRCA-2
- Prior cancer in the opposite breast
- Cancer of the breast occurs most commonly in the upper and outer quadrant of the breast.

What to do for certain breast masses?

When to do an ultrasound (US) → this is the first step when finding a palpable mass that feels cystic on physical exam.

When to do fine needle aspiration (FNA) → this is often the first step when finding a palpable mass, and may be done either after an US or instead of an US.

When to do either mammography (if pt >40yr) and biopsy (or biopsy solely if pt is <40yr):

→ If cyst recurs more than twice in 4-6 months

→ If there is bloody fluid on aspiration

→ If mass doesn't appear completely with a FNA

→ There is bloody nipple discharge

→ There is edema of the skin and erythema that suggests inflammatory breast carcinoma (excisional biopsy)

When to order for cytology → any aspirate with gross blood must be send for cytology

When to observe and repeat an exam within 6-8 weeks → whenever a cyst disappears with aspiration, and the fluid is clear, and/or when the needle biopsy and imaging studies are negative.

When should adjuvant therapy be included in the management → Use adjuvant therapy in all hormone receptor (+) tumors regardless of any other factors.

When to test for BRCA1 and BRCA2?

- If there is a history of early-onset breast or ovarian cancer in the family
- If there is breast and/or ovarian cancer in the same patient
- A family history of MALE breast cancer
- If patient is of Ashkenazi Jewish heritage

When is chemotherapy included in tumor management?

- When tumor is >1cm
- When the disease is node (+)

When is trastuzumab included in management?

- Included for metastatic breast cancer that over expresses HER2/neu
- This is a monoclonal antibody directed against the extracellular domain of the HER2/neu receptor

Tamoxifen:

- Competitively binds to estrogen receptors
- Produces a 50% decrease in recurrence and a 25% decrease in mortality
- Excellent for both pre and post-op patients

Aromatase Inhibitors:

- Include drugs such as: Anastrazole, Exemestane, and Letrozole
- These block the peripheral production of estrogen
- These are the standard of care in hormone receptor (+) women who are menopausal (more effective than tamoxifen)
- Will increase the risk of osteoporosis

LHRH analogues:

- A commonly used drug is Goserelin
- An alternative or an addition to tamoxifen in premenopausal women

Benefits of Tamoxifen	**Side effects of Tamoxifen**
⇓ incidence of contralateral breast cancer ⇑ bone density in postmenopausal women ⇓ serum cholesterol ⇓ fractures ⇓ cardiovascular mortality rate	Exacerbates menopausal symptoms ⇑⇑ risk of cancer of the endometrium **All women with a history of tamoxifen use + vaginal bleeding should have an evaluation of the endometrium

Treatment review:

If case describes HR(-), pre or post menopausal → chemo alone

If case describes HR(+), PREmenopausal → chemo + tamoxifen

If case describes HR(+), POSTmenop → chemo + aromatase inhibitor

Chapter 4

Pediatrics

The Newborn

The APGAR score:

This score measures the newborn's need for resuscitation and measures 5 criteria at 1-minute and 5-minutes. At 1-minute we can determine how well the baby did during labor and delivery, while at 5-minutes we can determine the effectiveness of resuscitation (if it was needed)

APGAR SCORING TABLE

	SCORE		
SIGN	**0**	**1**	**2**
Heart rate	Absent	<100	>100
Respiratory rate	Absent	Weak, irregular	Good, crying
Muscle tone	Flaccid	Arms and legs flexed	Well flexed
Reflex irritability	No response	Grimace	Cough or sneeze
Skin color	Blue, pale	Hands and feet blue	Completely pink

Managing the Newborn:

There are some important things that must be done immediately upon delivery of the newborn, upon delivery give the following:

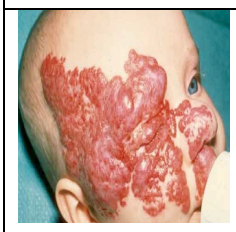
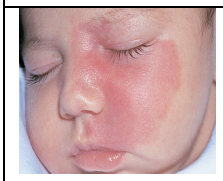
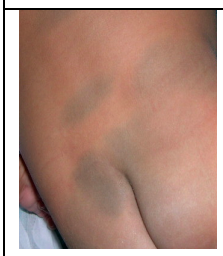
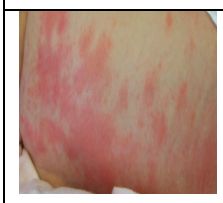
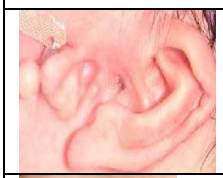
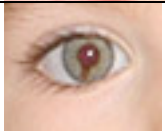
- Give 1% silver nitrate eye drops OR 0.5% erythromycin ophthalmic ointment
- 1mg of intramuscular vitamin K (prevents hemorrhagic disease of the newborn)

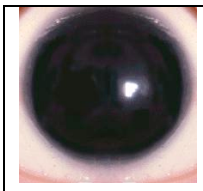
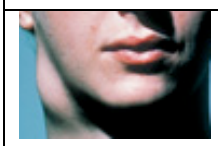
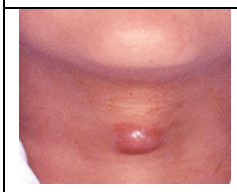
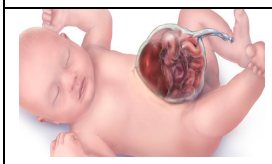
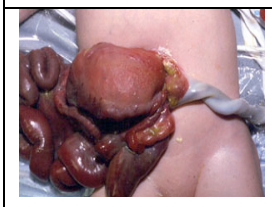
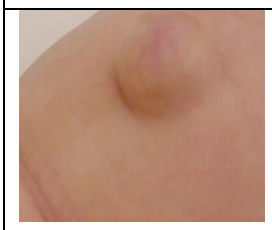
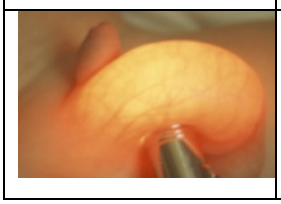
Before discharging the newborn from the hospital, do the following:

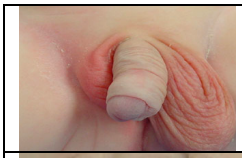
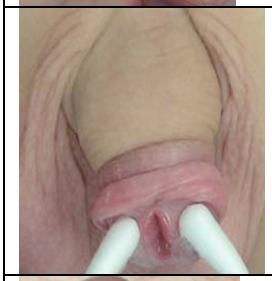
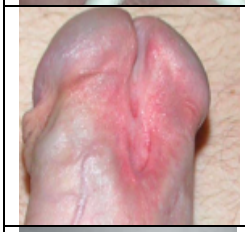
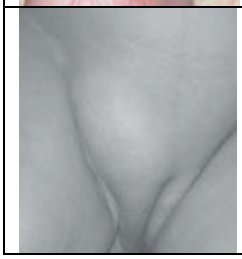
- Hearing test to rule out a sensorineural hearing loss
- Order neonatal screening tests: PKU, galactosemia, hypothyroidism

Abnormalities in the newborn:

There are many abnormalities of the newborn that should be recognizable. It is also important to know which are benign and which require further investigation.

Image	Description	Diagnosis	Co-morbidities	Management
	Red, sharply demarcated raised red lesions	Hemangioma	May be associated with high-output cardiac failure if very large. Consider underlying involvement of organs when large.	Steroids or laser therapy if it involves underlying organs
	Unilateral red formations on head and neck (unilateral and permanent)	Port Wine Stain	May be associated with Sturge-Weber syndrome	Pulsed laser therapy. If Sturge-Weber must evaluate for glaucoma and give anti-convulsives
	Bluish/gray macules on lower back and/or posterior thigh (most commonly)	Mongolian Spots		Rule out child abuse, usually fade within first few years of life
	Firm, yellow-white papules/pustules with erythematous base, peaks on 2nd day of life	Erythema Toxicum	None	None, is self-limited
	Tags or pits in front of the ear	Preauricular Tags	Associated with hearing loss and GU abnormalities	US of kidneys and hearing test
	Defect in the iris	Coloboma of the Iris	Associated with other CHARGE defects	Screen for CHARGE syndrome

	An absence of the iris	Aniridia	Associated with Wilm's tumor	Must screen for Wilm's tumor every 3 months until 8yr of age
	A mass lateral to the midline	Branchial Cleft Cyst	Is a remnant of embryonic development associated with infections	Surgical removal
	A midline mass that rises when swallowing	Thyroglossal Duct Cyst	May have ectopic thyroid, associated with infections	Surgical removal
	A protrusion of GI contents through umbilicus contained within a sac	Omphalocele	Associated with chromosomal disorders and other malformations	Screen for trisomies.
	An abdominal defect that is lateral to the midline with no sac covering contents	Gastroschisis	Associated with intestinal atresia	
	Rectus abdominus weakness that allows bulging of fetal and infant umbilical cord	Hernia (umbilical)	Congenital hypothyroidism	This may close spontaneously. Screen for hypothyroid with TSH screen.
	Scrotal swelling that can be transilluminated	Hydrocele	Associated with an inguinal hernia	Differentiate from inguinal hernia by shining flashlight

	Absence of teste(s) in scrotum	Cryptorchidism	Associated with cancer of the teste(s)	Surgical removal by 1yr
	Opening of the urethra on dorsum of the penis	Epispadias	Urinary incontinence	Evaluate for bladder extrophy
	Opening of the urethra on ventral surface of penis	Hypospadias		No definitive mgmt, but not supposed to circumcise the infant
	Reducible scrotal swelling	Inguinal Hernia		Surgical correction

Developmental Milestones

Developmental milestones show up over and over again on the board exams. By memorizing the important milestones you are going to get 2-3 easy points on the CK exam. Milestones refer to both infant and adolescent (puberty) milestones.

Developmental Milestones

Age	Gross Motor	Fine Motor	Language	Social/Cognition
Newborn	**Moro reflex, grasp reflex**			
2 months	Holds head up	Swipes at objects	Coos	Social smile
4 months	Rolls front to back	**Grasps Objects**	Orients to voice	Laughs
6 months	Rolls from back to front, **sits upright**	Transfers objects	Babbles	**Develops stranger anxiety, sleeps all night**
9 months	Crawls, pulls to a stand	**Pincer grasp,** eats with fingers	**Non-specific words**	Waves goodbye, responds to name
12 months	**Stands on own**	**Mature pincer grasp**	**Specific words "mama"**	Recognizes pictures in a book/magazine
15 months	**Walks**	Uses a cup	Speaks 4-6 words	**Throws temper tantrum**
18 months	Throws a ball, walks up the stairs	Uses spoon for solid foods	Names common objects	**Begins toilet training**
24 months	Starts running, can go up and down stairs	Uses spoon for semi-solids	**Speaks 2-word sentence**	Can follow a 2-step command
36 months	Can ride a tricycle	Can eat neatly with utensils	**Speaks 3-word sentence**	Knows first and last names

Puberty:

The milestones of puberty are as follows and are based on population averages:

MALES	FEMALES
Testicular enlargement – 11.5 yrs	Breast buds – 10.5 yrs
Genitals increase in size	Pubic Hair Growth Begins
Pubic Hair Growth Begins	Linear Growth Spurt – 12 yrs
Peak Growth Spurt – 13.5 yrs	Menarche – 12.5 yrs

Child Abuse

- Suspected child abuse requires your BY LAW to report the suspected abuse.
- You must also explain to the parents why you suspect abuse and that you are legally obligated to report it to child protective services.
- If a parent refuses hospitalization or treatment of their child against the best interest of the child, you must get an emergency court order.

High-risk children:

- Premature infants
- Children with chronic medical problems
- Infants with colic
- Children with behavioral problems
- Poverty stricken children
- Children of teenage parents
- Single parents
- Children of substance abusers

Classic findings:

- Chip fractures (damage to the corner of metaphysis in long bones)
- Spiral fractures
- Rib fractures
- Burns (immersion in hot water, cigarette burns, stocking-glove burns on hands and feet)
- Head injury – MCC of death
- Sexual abuse

When to hospitalize a child under suspect of being abused:

- The hospital is the safest place for the child
- The diagnosis is still unclear
- The child has a medical condition requiring hospitalization

Workup for suspected child abuse:

- PT/PTT/BT
- Full skeletal survey for breaks
- If injuries are severe, get a CT or MRI + a thorough eye exam
- If injury is to the abdomen, get an abdominal CT, check for blood in the stool and urine, and check liver and pancreatic enzymes

Treatment:

1st - Address medical and/or surgical issues before all else

2nd – Report abuse to child protective services (CPS)

Breast Feeding

There are many advantages to breastfeeding:

- Psychological and emotional bonding between mother and infant
- Passive transfer of T-cell immunity decreases risks of allergies and infection

Contraindications to breast feeding:

- HIV
- CMV
- HSV (only if lesions are on breast)
- HBV (unless vaccination is given prior)
- Substance abuse
- Breast cancer
- Acute illness in mother that is absent in infant
- Drugs (list below of contraindicated drugs during breastfeeding)

Absolutely Contraindicated	**Relatively Contraindicated**
Alcohol	Steroids
Nicotine	Neuroleptics
Antineoplastics	Sedatives

Lithium	Tetracycline
Chloramphenicol	Sulfonamides
Iodide and Mercurial Drugs	Metronidazole

High-yield Growth & Development Facts

- The height of a child at 2 years of age normally correlates with the final adult height percentile.
- By 6 months of age the birth weight should double, and by 1 year the birth weight should triple.
- The absolute best indicator for malnutrition is a child who is <5th percentile for height and weight.
- Skeletal and sexual maturity are related more than it is related to chronological age.
- The MCC of failure to thrive (FTT) in all age groups is psychosocial deprivation.
- In patients with genetic short stature or constitutional delay, birth weight is normal.
- Patients with both genetic short stature and constitutional delay have a growth pattern that is below and parallel to the normal growth curve.

Infectious Diseases – The ToRCHS

In general, all will have: jaundice, IUGR, mental retardation, and hepatosplenomegaly. Look for things that stand out with each infection

Disease	Characteristics
Toxoplasmosis	• Acquired by mother through poorly ingested meat • Acquired when mother handles cat feces through litter box • 1/3 of mothers transmit and 1/3 of infants are affected • Causes: **intracerebral calcifications**, IUGR, severe mental retardation, **hydrocephalus**, chorioretinitis, epilepsy, hepatosplenomegaly • If infected → do ultrasound to find any major anomalies
Rubella	• When acquired in 1st trimester there is an 80% chance of transmission • When acquired in 2nd trimester there is a 50% chance of transmission • When acquired in 3rd trimester there is a 5% chance of transmission

	• Signs & Symptoms – **cataracts**, IUGR, blueberry muffin rash, glaucoma, chorioretinitis, PDA, pulmonary stenosis, **ASD, VSD, myocarditis**, hearing loss, mental retardation, **deafness** • Diagnosis – confirm with **IgM** rubella antibody in neonate's serum. • Treat – goal is universal prevention by immunizing all children, there's no therapy for active infection
Cytomegalovirus (CMV)	• Affects 1% of all births and is the most common congenital infection • Infection is often asymptomatic • Approx 1% risk of transplacental transmission, and approx 10% of infected infants manifest defects • Congenital defects – microchephaly, **periventricular intracranial calcifications**, IURG, chorioretinitis, severe mental retardation, **sensorineural hearing loss** • Transmission is through body fluids • **Diagnose with urine CMV culture**
Herpes Simplex Virus	• Vaginal delivery during active infection = approx 50% get infected • C-section is required if active infection • **1st week – pneumonia and shock** • **2nd week – skin vesicles, keratoconjunctivitis** • **Week 3-4 – acute meningoencephalitis** • Treat – acyclovir → significantly decreases mortality
Syphilis	• There is almost a 100% transmission rate, occurs mostly after 1st trimester • 40% death rate (fetal and perinatal) • Manifests early (first 2 years), then late (within next 2 decades) • Signs/Symptoms of early infection – jaundice, increased LFTs, hemolytic anemia, rash that is followed by **desquamation of the hands and feet**, **snuffles** (blood-tinged nasal secretions), **osteochondritis**, sattle nose. • Late symptoms – Hutchinson teeth (upper 2 incisors get notched), mulberry molars, bone thickening (frontal bossing), saber shins (anterior bowing of tibia) • **Best initial test – VDRL screening** • **Most specific test – IgM-FTA-ABS** • Treat – Penicillin G for 10-14 days

Infant Botulism

- An acute and flaccid paralysis caused by C. Botulinum.
- Irreversible blocks release of Ach
- Caused by the ingestion of raw honey

Signs/Symptoms:

- Constipation
- Lethargy
- Weak cry
- Poor feeding
- Hypotonia
- Drooling
- ⇓ suck reflexes
- ⇓ spontaneous movements

Diagnosis: Based on PE and the acute onset of flaccid paralysis

Treatment: Supportive care + intubation

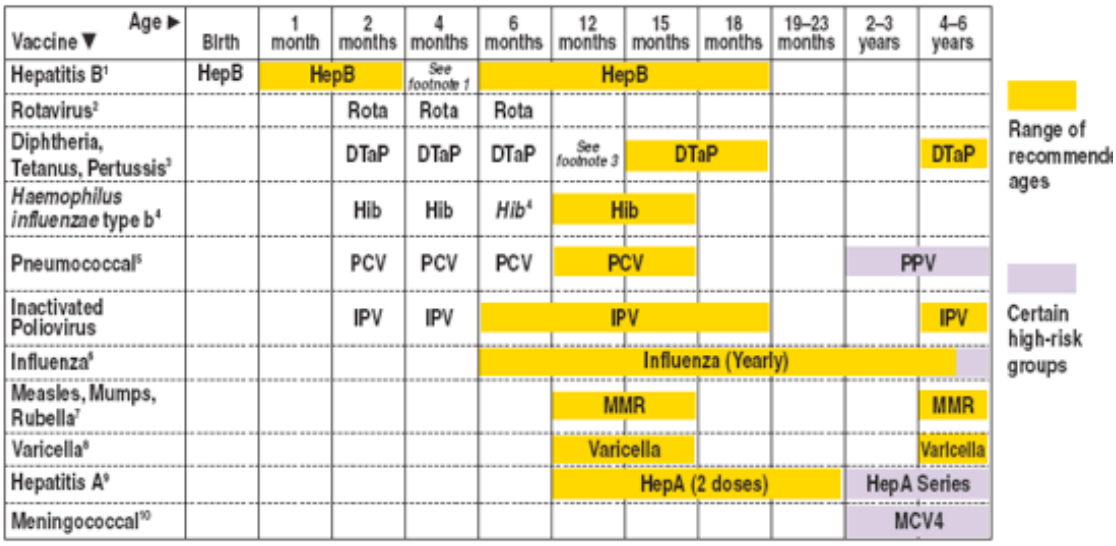
<u>Common Viral Exanthems</u>

Disease	**Signs and Symptoms**
Measles (Rubeola) Paramyxovirus	• Begins at hairline then moves downward, is an erythematous maculopapular rash that erupts 5 days after prodrome. • Pathognomonic "Koplik spots" often disappear before rash starts (white spots on buccal mucosa) • Diagnosis – cough, coryza, conjunctivitis (3C's)
Rubella (German measles) Togavirus	• Suboccipital lymphadenopathy* • A maculopapular rash starts on the face then generalizes • Rash lasts approximately 5 days • Soft palate may show red spots of various sizes
Hand, foot, & mouth disease (Coxsackie A)	• Patient has vesicular rash on the hands and feet + ulcerations in the mouth • Rash lasts approximately 1 week • Is contagious by contact
Roseola Infantum (HHV-6)	• Acute fever lasts 1-5 days, but child shows no physical symptoms and does not feel ill • Once fever drops, a maculopapular rash appears over the whole body (lasts 24hrs)
Erythema Infectiosum (5th disease – Parvo	• "Slapped cheek" syndrome • An erythematous maculopapular rash spreads

B19)	from the arms to the trunk/legs, forms “reticular” pattern • Dangerous if pt has sickle cell disease due to tendency to form aplastic crisis
Varicella (chicken pox)	• Highly contagious, teardrop vesicles that break and crust over. • Starts on face and trunk then spreads • Contagious until crusting over

Vaccinations

This diagram is the typical vaccination recommendation for children from 0-6yr old



RESPIRATORY DISORDERS

Upper Respiratory Infections

Condition	Presentation	Diagnosis	Management	Prognosis
Croup (Parainfluenza 1 or 3, Influenza A or B)	3mnth – 5yr with URI sx + deep barking cough/stridor. Symptoms worsen at night	Nothing needed for diagnosis but a neck-x-ray should be had	1. Humidified 02 2. Nebulized epi + steroids	Spontaneously resolves within 1 wk. Always be wary of the possibility of epiglottis
Epiglottitis (Hib, S. pyogenes, s. pneumo, and mycoplasma)	Acute onset of muffled voice, drooling, high fever, dysphagia, and inspiratory stridor. Patient will lean forward to ease breathing	**Medical Emergency** Mgmt based on clinical dx, stabilize first then do workup: Workup: Neck xray looking for thumbprint sign. Blood cultures Epiglottic swab culture	1. Admit to hospital 2. Anesthesia and ENT consult 3. Intubation 4. Ceftriaxone + steroids 5. Household contacts should get Rifampin if patient is H.Influenza +ve	Without prompt treatment airway obstruction can lead to death
Bacterial Tracheitis (S. Aureus)	Child usually <3yr, after a viral URI gets cough that sounds "Brassy", has high fever, respiratory distress BUT	Dx is clinical but also do a laryngoscopy and CXR CXR looking for sub-glottic narrowing	Antistaph Ab's, if severe intubate	Airway obstruction is a life-threat complication

	no signs or symptoms of severity of epiglottitis	Blood cult + throat cult.		

Otitis Media

- Common in children and often precipitated by an URI
- Conditions that disrupt proper Eustachian tube drainage lead to chronic OM
- MCC are: strep pneumonia, H. Infl, Moraxella, or viral causes

Signs and Symptoms:

- Erythema and ⇓ motility of tympanic membrane
- ⇓ hearing
- Ear pressure
- Bulging tympanic membrane with visualization of fluid behind TM

Treatment:

1st line → Amoxicillin

2nd line → Amoxicillin + Clavulinic Acid (augmentin)

** For recurring OM, ENT consult and tubes may need to be inserted

Bronchiolitis

Classically presents as child <2yr with the following:

- Mild URI
- Fever
- Paroxysmal wheezing cough
- Tachypnea
- Dyspnea
- Wheezing and prolonged expirations

Common causes are:

- RSV (in up to 50%)
- Parainfluenza virus
- Adenovirus

Signs and Symptoms:

- Inflammation
- Air trapping and over inflation (due to ball-valve obstruction)

Diagnosis:

Dx is clinically based.

Best initial test → CXR looking for hyperinflation + patchy atelectasis

Most specific test → Immunofluorescence of nasopharyngeal swab

Treatment:

- Mostly supportive
- If tachypnea is severe hospitalize and give trial of nebulized β-agonists

Pneumonia

There are different causes of pneumonia:

Viral → MCC in children <5yr, MCC is RSV

Bacterial → MCC in children >5yr, MCC are S. Pneumo, Mycoplasm Pneumo

Chlamydial → Common in infants 1-3 month with insidious onset

Viral:

- Tachypnea is the most consistent finding in viral pneumonia
- URI symptoms
- Low grade fever

Bacterial:

- Acute onset with sudden shaking chills
- High grade fever
- Cough
- Chest pain (pleuritis- pain with respiration)

- Diminished breath sounds
- Dullness to percussion of the lung fields

Chlamydial:

- Most common finding are a "staccato cough" and "peripheral eosinophilia"
- No fever or wheezing
- May be conjunctivitis

Diagnosis:

CXR:

- Viral → hyperinflation with bilateral interstitial infiltrates
- Bacterial (pneumo) → lobar consolidation
- Mycoplasma/Chlamydia → unilateral lower-lobe interstitial pneumo that looks worse than the patient's presenting symptoms

CBC:

- Viral → <20000 wbc
- Bact → 15000-40000

Treatment:

- Mild cases can be managed on an outpatient basis, Amoxicillin is the best choice. Augmenting may also be used
- Severe cases require hospitalization and are treated with IV ceftriaxone
- If pneumonia is of viral origin, withold Ab's unless patient deteriorates.
- Chlamydia or Mycoplasma treated with erythromycin

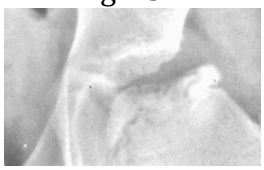
MUSCULOSKELETAL DISORDERS

Limp

- ***Painful limping*** most often occurs acutely, and may be associated with fever, irritability.
- Young infants may refuse to walk
- ***Painless limping*** usually has an insidious onset and is more commonly due to weakness or deformity of the limb secondary to developmental hip dysplasia, cerebral palsy, or leg-length discrepancy

This table shows the different causes of ***PAINFUL LIMP***

Disease	Characteristics	Treatment
Arthritis (Septic)	• The #1 cause of painful limp in 1-3yr old • Is usually monoarticular (hip, ankle, or knee) • MCC S. Aureus ***Si/Sx:*** Acute onset of pain, ⇓ ROM, fever, arthritis, ⇑ wbc, ⇑ ESR ***Xray***: shows joint space widening + soft tissue swelling. ***Diagnose***: joint aspirate shows WBC ≥ 10,000 with PMN predominance	Drainage + antibiotics that are appropriate to the culture obtained from the joint aspirate
Toxic Synovitis	• MC in males 5-10yr and may procede URI ***Si/Sx***: insidious onset of pain, low-grade fever, wbc and ESR are normal Usually no tenderness, warmth, or swelling	Rest + analgesia for 3-5 days

	Xray is normal ***Diagnose***: technetium scan that shows ⇑ epiphyseal uptake	
Aseptic Vascular Necrosis	**Legg-Calve-Perthes dx** • Head of femur • 4-9yr old • Boys 5x more than girls  **Osgood Schlatter** • Tibial tubercle • Active child/adolescent • Rest relieves pain **Kohler's bone** • **Navicular bone** ***Si/Sx***: afebrile with insidious onset of hip pain, pain of inner thigh/knee, ⇑ pain with movement, ⇓ with rest, normal wbc and ESR ***Xray***: femoral head sclerosis and ⇑ width of the femoral neck ***Dx***: technetium scan shows ⇓ uptake in epiphysis	⇓ weight bearing on affected side over long term
Slipped Capital Femoral Epiphysis (SCFE)	• MC in obese males 8-17yr • 20%-30% bilateral • 80% occur slowly and progressively where 20% occur acutely and associated with trauma	Surgical pinning

	Si/Sx: dull, aching pain in hip/knee, pain with activity ***Xray***: "ice cream scoop falling off cone" to describe lateral movement of the femur shaft in relation to the femoral head ***Dx***: strictly clinical	
Osteomyelitis	***Neonates*** – S. Aureus 50% of time ***Children*** – Staph, Strep, Salmonella (sickle cell) ***Si/Sx*** in young infants: only symptom may be fever ***Si/Sx*** in older children: fever, malaise, edema, and ⇓ extremity movement ***Dx***: neutrophilic leukocytosis, ⇑ ESR, blood cultures, bone scan is 90% sensitive. MRI is gold standard	IV antibiotics for 4-6 weeks

Collagen Vascular Disease

Juvenile Rheumatoid Arthritis

- Chronic inflammation of ≥ joints in a patient ≤ 16yr
- Occurs MC in 1-4yr olds, females > males
- There are 3 categories: Systemic, pauciarticular, and polyarticular

Diagnosis: Symptoms that persist for 3 consecutive months with the exclusion of other causes of arthritis or collagen vascular disease.

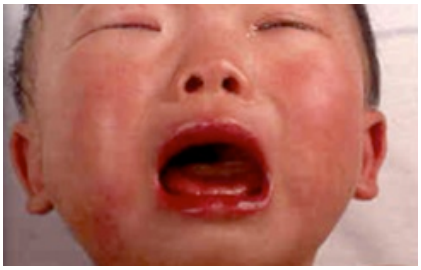
Treatment: NSAIDs, low-dose MTX, and prednisone in acute febrile onset

Types of Juvenile RA

Systemic (Still's disease) – 10%-20%	• Patient has high-spiking fever that returns to normal daily • Small, pale pink macules with central pallor on trunk & proximal extremities with possible palm & sole involvement • Joint involvement may not occur for weeks to months • 1/3 have disabling chronic arthritis
Pauciarticular – 40%-60%	• Involves ≤ 4 joints, primarily affecting large joints (knee, elbow, ankle) • Chronic joint disease is abnormal • Fever/malaise/anemia/lymphadenopathy common 2 Types: Type 1 – MC, females <4yr, 90% ANA (+), incr risk of chronic iridocyclitis Type 2 – MC males >8yr, ANA (-), 75% are HLA-B27 (+), incr risk of Ankylosing spondylitis or Reiter's syndrome later in life
Polyarticular	• ≥ 5 joints are involved, both small & large, insidious onset, fever, lethargy, anemia • There are 2 types that depend on whether rheumatoid factor is (+) or (-) • RF (+) – 80% females, late onset, more severe, rheumatoid nodules present, 75% are ANA (+) • RF (-) – occurs at any time during childhood, mild, rarely ass'd with rheumatoid nodules, 25% are ANA (+)

Kawasaki's Disease

- A mucocutaneous lymph node syndrome
- Affects large and medium vessel vasculitis in children <5yr of age
- More commonly seen in children of Japanese heritage



Diagnosis:

Diagnosis requires the presence of a FEVER > 104F or 40C for more than 5 days that is unresponsive to antibiotics **+** 4/5 of the following criteria:

Using the mnemonic CRASH to remember the criteria

1. **C**onjunctivitis
2. **R**ash (truncal)
3. **A**neurysms of the coronary arteries
4. **S**trawberry tongue
5. **H**and and foot induration (erythema of the palms and soles)

Complications:

- 10%-40% of untreated cases show dilation/aneurysm of the coronary arteries

Treatment:

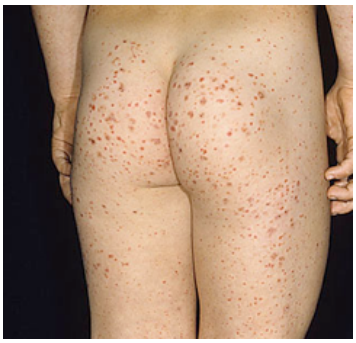
- IVIG to prevent coronary vasculitis + high-dose aspirin
- Do not give steroids as this will exacerbate the condition

Prognosis:

- With response to IVIG + aspirin is rapid and 2/3 become afebrile within 1 day.
- Always re-evaluate in 1 week, repeat ECHO at 3-6wk post illness
- If no further abn on ECHO then no further imaging is necessary

Henoch-Schonlein Purpura

- A small-vessel vasculitis mediated by IgA nephropathy (Berger's disease)



Signs and symptoms:

- A palpable purpura on the legs and buttocks is pathognomonic in children
- May also have abdominal pain due to intussusception

Treatment:

- Self-limited and rarely progresses to glomerulonephritis

Histiocytosis X

- A proliferation of histiocytic cells resembling Langerhan's skin cells

There are 3 common variants:

1. **Letterer-Siwe disease**

- An acute, aggressive, disseminated variant that is often fatal in infants

Signs and Symptoms:

- Hepatosplenomegaly
- Lymphadenopathy
- Pancytopenia
- Lung involvement
- Recurrent infections

2. **Hand-Schuller-Christian**

- A chronic/progressive variant that presents prior to 5 yr

Classic triad: Skull lesions + diabetes insipidus + exophthalmos

3. **Eosinophilic granuloma**

- Extraskeletal involvement usually limited to the lungs
- Has the best prognosis of all variants and often regresses spontaneously

Metabolic Disorders

Congenital Hypothyroidism

- Newborn screening is mandatory by law
- T4 is essential during the first two years of life for normal brain development
- Usually due to secondary thyroid agenesis or enzyme defects
- Birth history is usually normal with a prolonged period of jaundice

Signs and Symptoms:

- At 6-12 weeks the infant develops poor feeding, lethargy, hypotonia, coarse facial features, large protruding tongue, constipation, hoarse cry, and developmental delay

Diagnosis:

- ⇓ T4, ⇑ TSH

Treatment:

- Levothyroxine
- Delay of treatment beyond 6 wks results in mental retardation

Newborn Jaundice

Timeframe	Differential Dx
Within 24hr of birth	• Sepsis • Hemolysis (ABO/Rh isoimmunization, hereditary spherocytosis)
Within 48hr of birth	• Hemolysis • Infection • Physiologic
After 48hr	• Infection • Hemolysis • Breast milk jaundice • Congenital malformation • hepatitis

** 50% of neonates have jaundice during their first week of life

Physiologic jaundice:

- Clinically benign condition that occurs between 24-48hr after birth
- Characterized by unconjugated hyperbilirubinemia
- Cause is increased bilirubin production + a relative deficiency in glucuronyl transferase in the immature liver

Treatment:

- None required

Jaundice present at birth - pathologic

- Is always pathologic and appears within 24hrs of birth
- Bilirubin rises >5mg/dL/day
- Bilirubin >12mg/dL in term infant
- Direct bilirubin >2mg/dL at any time
- Hyperbilirubinemia is present after the 2nd week of life

Workup for pathologic jaundice:

- Total and direct bilirubin
- Direct Coomb's test
- Blood type of infant and mother (ABO or Rh incompatibility)

- CBC, retic #, peripheral smear (assessing for hemolysis)
- U/A and urine culture (if elevated is direct bilirubin – assess for sepsis)

If prolonged >2 weeks, do the following:

If ⇑ conjugated bilirubin

- Initial diagnostic tests → LFT's
- Most specific test → US and liver biopsy

If no elevation of unconjugated bilirubin

- UTI or other type of infection
- Bilirubin conjugation abnormalities (Gilberts, Crigler-Najjar)
- Hemolysis
- Intrinsic red cell membrane defect or enzyme defect (spherocytosis, elliptocytosis, G6PD def, pyruvate kinase deficiency)

Treatment:

- When bilirubin is >10-12 mg/dL → phototherapy
- Exchange transfusion if encephalopathy is suspected or there is failure of improvement with phototherapy

Unconjugated Hyperbilirubinemia

- Caused by ***hemolytic anemia*** or congenital deficiency of glucuronyl transferase (Crigler-Najjar, Gilbert's syndrome)

Hemolytic anemia:

- Congenital or acquired
- **Congenital** → spherocytosis, G6PD, pyruvate kinase deficiency
- **Acquired** → ABO/Rh isoimmunization, infection, drugs, twin-twin transfusion, chronic fetal hypoxia, delayed cord clamping, maternal diabetes

Conjugated Hyperbilirubinemia

- Infectious causes are → sepsis, ToRCH's, hepatitis, syphilis, listeria infection
- Metabolic causes are → galactosemia, α1-antitrypsin def
- Congenital causes are → extrahepatic biliary atresia, Dubin-Johnson syndrome, Rotor syndrome

Treatment:

- UV light to break down bilirubin pigments
- Urgent treatment is imperative in order to prevent kernicterus induced mental retardation

Complications:

- UV light can cause diarrhea, dermatitis, dehydration, and damage to the retina (be cautious of these adverse effects)

Reye Syndrome

- The use of salicylates in children causes an acute encephalopathy + fatty degeneration of the liver
- Most commonly occurs in children aged 4-12yr

Signs and Symptoms:

- Alternates an asymptomatic interval with abrupt onset of vomiting, delirium, stupor, abnormal LFT's
- Rapid progression to seizures, coma, and death

Diagnosis:

- Significantly elevated liver enzymes

Treatment:

- Urgent ICP management with mannitol and fluid restriction
- Glucose administration due to rapid depletion of stores

Prognosis:

- Bad if serum ammonia levels are increased 3fold, and if there is a decreased PT level that WONT respond to vitamin K
- If disease is mild the recovery is usually good and rapid
- A severe disease can result in permanent neuro defects

Seizures

- In the newborn, seizures may present as jitters with repetitive sucking movements, tongue thrusting, and apneic spells.
- Blood counts and chemistries are often WNL
- Neonatal seizures can be diagnosed by the presence of ocular deviation and failure of jitters to subside with stimulus

Diagnosis:

- EEG – often normal
- CBC + chemistry panel → often the cause is hypoglycemia in case of GDM
- Amino acid assay looking for inborn errors of metabolism
- Total cord blood IgM to look for ToRCH infections
- Urine cultures
- LP if suspected meningitis
- US of head if infant is preterm → looking for bleeding

Treatment:

- 1st line DOC is Phenobarbital
- Persistent seizure not responsive to Phenobarbital – give Phenytoin

Febrile Seizures

- Usually between 3mnt – 5yr
- A fever is present with no other signs of infection
- Is the MC convulsive disorder in children and rarely develops into epilepsy
- Seizure occurs during temperature rise (≥ 102F) but not at its peak

Signs and Symptoms:

- MC is a tonic-clonic seizure that rarely last more than 10min + a drowsy postictal period is common
- Seizure lasting >15min is usually due to an infectious process

Diagnosis:

- Clinical diagnosis is usually all that is needed
- Routine labs only required to identify a source of the fever
- LP to rule out meningitis (only if suspected)

Treatment:

- Control fever with antipyretics
- Reassure parents/counsel

- Always do a careful evaluation for the source of the fever

** 30%-50% of children experience recurrent febrile seizures

Genetic and Congenital Disorders

Failure to Thrive

- Is the failure of children to grow and develop at an appropriate rate
- May be due to inadequate calories or inability to absorb the calories
- May be idiopathic or due to other diseases
- Factors such as *poverty*, *family problems*, *neonatal problems*, and *maternal depressio*n should all be included in diagnosis

Diagnosis:

Requires 3 criteria for a FTT diagnosis:

1. Child <2yr with weight <5th percentile for age on >1 occasion
2. Child <2yr whose weight is <80% of ideal for age
3. Child <2yr whose weight crosses 2 major percentiles downward on a standardized growth chart

Exceptions:

- Genetically short stature
- Small for gestational age children
- Preterm infants
- Very lean (be careful here)
- ⇑ height with a ⇓weight gain (causes an overweight child – careful attention for this diagnosis)

Treatment:

Organic causes → treat underlying condition + supplement with sufficient calories

Idiopathic causes → educate parent on nutrition and observe parent while feeding

Older infants/children → offer solids before liquids, ⇓ mealtime distractions, have child eat with others, never force-feed

** Monitor closely for weight gain with adequate calorie consumption

Prognosis:

- In the 1st year of life the px is poor since the brain develops early in life
- 1/3 of children with nonorganic failure to thrive are developmentally delayed

Craniofacial Abnormalities

- The mildest form of craniofacial abnormality is the "bifid uvula", and has no clinical significance

Cleft Lip:

- May occur unilaterally or bilaterally
- Caused by a failure of fusion of maxillary prominences
- MC form is unilateral cleft lip
- No interference with feeding
- Treat with surgical repair

Cleft Palate:

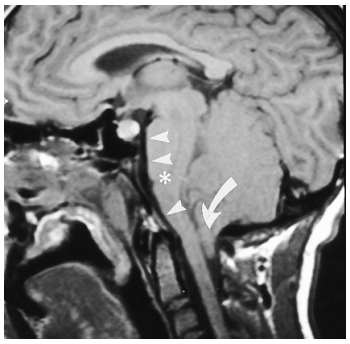
- May be anterior or posterior
- Anterior cleft palate is due to failure of the fusion of the palatine shelves with the primary palate
- Posterior cleft palate is due to failure of the fusion of the palatine shelves with the nasal septum
- These conditions will interfere with feeding and thus require a special nipple for the baby to feed properly
- Treat with surgical repair

Macroglossia:

- Is a congenitally enlarged tongue
- Seen in conditions such as Down's syndrome, hypothyroidism, and gigantism
- Can be acquired later in life via acromegaly and/or amyloidosis
- Is NOT glossitis, which is from a B-vitamin deficiency
- Treatment is aimed at treating the underlying cause

Arnold-Chiari Malformation

A congenital disorder where the cerebellum is caudally displaced, the medulla is elongated and passes into the foramen magnum.



Signs and Symptoms:

- Flattened skull base
- Hydrocephalus
- Aqueduct stenosis

Prognosis:

- Death usually as neonate of toddler

Neural Tube Defects

- Associated with increased α-fetoprotein in the maternal serum
- VERY PREVENTABLE with FOLATE supplementation

Signs and Symptoms:

- Spina bifida (failure of posterior vertebral arches to close)
- Meningocele (lack of vertebrae covering the lumbar spinal cord)

Treatment:

- Prevention is key (folic acid supplementation)
- Neuro deficits remain

Fetal Alcohol Syndrome

- In children born to alcoholic mothers, or mothers who consumed excessive alcohol during pregnancy

Signs and symptoms:

- Characteristic facial abnormalities and developmental delays
- ASD
- Microcephaly
- Smooth filtrum of upper lip

Treatment:

- Cessation of ETOH consumption when pregnant

Congenital Pyloric Stenosis

- Presents with projectile vomiting in first 2wk-2month of life
- Seen more commonly in males and in 1st-born children
- The pathognomonic finding is the palpable "olive mass" in the mid-epigastrium (hypertrophied pyloric stenosis)

Diagnosis:

- Palpation of "olive" mass is often sufficient
- If no mass can be palpated, to an ultrasound (US)

Treatment:

- Longitudinal surgical incision of hypertrophied pylorus

Congenital Heart Diseases

***ASD**:*

- Often asymptomatic and found on routine physicals
- Can predispose to CHF in the 2nd and 3rd decades of life
- May also predispose to strokes (due to an embolus bypass tract)

Signs and Symptoms:

- Midsystolic ejection murmur
- Loud S1
- Wide fixed-split S2

Diagnosis:

- Echocardiography

Treatment:

- Surgical patching
- Treatment is more important for females because they have an increase in cardiovascular stress during pregnancy

Ventricular Septal Defect (VSD)

- Is the MC congenital heart defect
- 30% of these VSD's close spontaneously by 2yr of age

Signs and Symptoms:

- Small defects may be asymptomatic
- Large defects can cause CHF
- Can cause delayed/decreased development and growth
- Holosystolic murmur heard over the entire precordium and maximally at the 4^{th} left intercostal space

Eisenmenger's Complex:

- A right to left shunt secondary to pulmonary hypertension
- RV hypertrophy causes a flow reversal through the shunt, resulting in a R→ L shunt
- Get cyanosis secondary to lack of blood flow to the lungs

Diagnosis:

- Echocardiography

Treatment:

- Simple defects require complete closure

Tetralogy of Fallot

Four defects make up this tetralogy, they are:

1. VSD
2. Pulmonary outflow obstruction
3. RV hypertrophy
4. Overriding aorta

Signs and Symptoms:

- Cyanosis develops within first 6 months of life (not present at birth)
- "Tet Spells" are acute cyanosis accompanied by panic, where child goes into a squatting position because it helps improve blood flow to the lungs

Diagnosis:

- Echo
- CXR shows "boot-shaped" contour of the heart due to RV enlargement

Treatment:

- Surgical repair of VSD and pulmonary outflow tracts

Transposition of the Great Arteries

- Aorta comes off the right ventricle
- Pulmonary artery comes off left ventricle
- Without a persistent AV communication this condition is incompatible with life. Thus requires a PDA or persistent foramen ovale.

Signs and Symptoms:

- Marked cyanosis at birth
- Early clubbing of the digits
- CXR shows an enlarged egg-shaped heart and an increase in pulmonary vasculature

Diagnosis:

- Echo

Treatment:

- Surgical switching of the arterial roots to normal positions with repair of communication defect

Prognosis:

- Without treatment is fatal within several months of birth

Coarctation of the Aorta

- A congenital aortic narrowing that is often asymptomatic in children

Signs and Symptoms:

- Normal BP in arms with decreased BP in legs
- Continuous murmur over collateral vessels in the back
- The classic XRAY shows "rib notching"

Diagnosis:

- Confirm with CT or aortogram

Treatment:

- Surgical resection of coarctation and reanastomosis

Patent Ductus Arteriosus (PDA)

- ⇑ incidence with premature births
- Pt predisposed to endocarditis and PVD's

Signs and Symptoms:

- Continuous machinery murmur that's best heard at 2nd left interspace
- Wide pulse pressure
- Hypoxia

Diagnosis:

- Echo or cardiac catheterization

Treatment:

- Indomethacin induces closure (blocks prostaglandins) for children

- Older children usually require surgical repair

Genetic Anomalies

Condition	Classic Features	Workup/Associations
Down's Syndrome (trisomy 21) Increased risk when maternal age is >35yr	• Epicanthal folds • Slanted palpebral fissures • Speckling of iris • Late fontanel closure • Mental retardation	• Hearing exam • ECHO: VSD, ASD, PDS • GI: TEF, duodenal atresia • TSH for hypothyroidism • ALL, decreased risk with increasing age
Edwards syndrome (trisomy 18)	• Low-set, malformed ears • Microcephaly • Micrognathia • Clenched hand • Rocker-bottom feet • Omphalocele	• ECHO: VSD, ASD, PDS • Renal US: polycystic kidneys, ectopic or double ureter • Most pts don't survive 1st yr
Patau Syndrome (trisomy 13)	• Midface defects • Eye defect • Defective forebrain development • Microcephaly • Microphthalmia • Cleft lip and palate	• ECHO: VSD, PDA, ASD • Renal US: polycystic kidneys • Single umbilical artery
WAGR syndrome	1. Wilm's 2. Aniridia 3. GU anomalies 4. Retardation (mental)	• The presence of aniridia should alert for the workup for WAGR
Klinefelter (XXY) 1/500 males	• Low IQ • Gynecomastia • Behavioral problems • Long/slim limbs	• Testosterone levels: hypogonadism and hypogenitalism • Testosterone replacement at 11-12 yr of age
Turner's syndrome (X0) A sporadic condition with no maternal age association	• Small-statured female • Low IQ • Gonadal dysgenesis • Webbed neck • Broad chest • Wide-spaced nipples	• Renal US: horseshoe kidney, double renal pelvis • Cardiac: bicuspid aortic valve, coarctation of the aorta • Thyroid function: primary

		hypothyroidism • Supplement with estrogen, GH, and anabolic steroids
Fragile X Syndrome	• Microcephaly in early childhood • Large ears • Large testes • Is the MCC of mental retardation in boys	• Ass'd with ADHD
Beckwith-Wiedemann syndrome	• Multi-organ enlargement • Macrosomia • Macroglossia • Pancreatic beta cell hyperplasia • Large kidneys • Neonatal polycythemia	• Increased risk of abd tumors • US and serum AFP q 6mnth up until 6yr of age – looking for Wilm's tumor
Prader-Willi (deletion at 15q11q13 – paternally derived)	• Obesity • Hyperphagia • Small genitalia • Mental retardation	• Morbid obesity decreases life-span
Angelman syndrome (aka "happy puppet" syndrome). (deletion at 15q11q13 maternally derived)	• Mental retardation • Inappropriate laughter • Absence of speech • Ataxia/jerky arm movements • Recurrent seizures	• Epilepsy develops in 80%
Pierre-Robin (ass'd with FAS and Edwards)	• Mandibular hypoplasia • Cleft palate	• Airway obstruction possible over first 4 wks of life, thus monitor airway

Trauma and Intoxication

Poisonings

Signs/Symptoms	Possible Toxins
Lethargy & Coma	ETOH, sedatives, narcotics, antihistamines, neuroleptics, anti-depressants
Seizures	Theophylline, cocaine, amphetamines, anti-depressants, antisphychotics, pesticides
Hypotension	Organophosphate pesticides, β-blockers
Arrhythmia	TCA's, cocaine, digitalis, quinidine
Hyperthermia	Salicylates, anticholinergics

- Approximately 50% of cases occur in children <6yr
- 92% occur at home, 60% with non-pharm agents, 40% with pharm agents
- 75% of cases are due to ingestion, 8% dermal, 6% ophthalmic, and 6% inhalation
- Lavage is often unnecessary in children but may be useful in drugs decreasing gastric motility
- Charcoal is often most effective and safest procedure to prevent absorption (but is ineffective in heavy metal or volatile hydrocarbon poisoning).

Adolescence

Epidemiology

Injuries:

- 50% of adolescent deaths attributed to injuries
- Many due to ETOH & elicit drugs
- Older adolescents have increase deaths due to MVA, while younger adolescents have deaths due to drowning and weapon injuries
- Homicide rates are 5x> for Blacks than White males

Suicide:

- Is the 2nd leading cause of adolescent death
- Females attempt more but males are 5x more likely to succeed
- Suicide attempts are greater in those who abuse ETOH and drugs

Substance abuse:

- A major cause of morbidity in adolescents
- Average age of 1st usage is 12-14yr
- High school seniors on average: 90% tried ETOH, 40% tried marijuana

Sex:

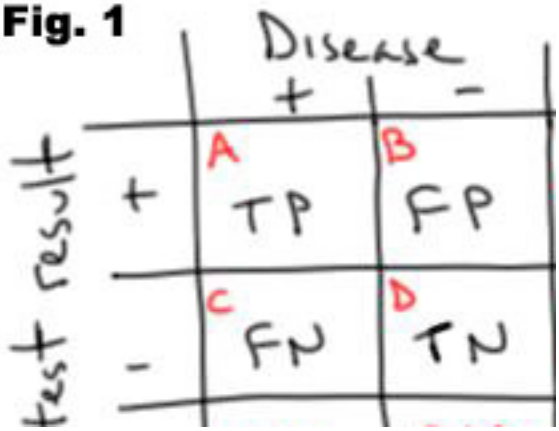
- 61% of all males and 47% of all females in high school have had sex
- Biggest risks: unwanted pregnancy, STD's
- 86% of STD's occur among adolescents and young adults between 15-29yr of age
- >1 million female adolescents become pregnant yearly, 33% are <15yr old

Chapter 5

Biostats

Success in answering the biostatistics questions comes from not only memorizing the following charts, but actually understanding them. If you can grasp what is happening you will not have any issues in this section.

Fig. 1



True Positive: is the # of people who have the disease with +ve results

False Positive: is the # of people who in fact do not have the disease with a +ve test result

True Negative: is the # of people who do not have a disease who tested –ve

False Negative: is the # of people who have the disease who tested -ve

Sensitivity → [A/A+C]

Sensitivity is a tests ability to detect a disease

Specificity → [D/B+D]

Specificity is a tests ability to detect health

Positive Predictive Value → [A/A+B]

The positive predictive value (PPV) detects the likelihood that the patient has a disease when they test positive for a test

Negative Predictive Value → [D/C+D]

The negative predictive value measures how likely a patient is in fact healthy after a test result comes back negative.

Odds Ratio → [(a X d) / (b X c)]

Compares the incidence of disease in people exposed X incidence of non-disease in people not exposed, divided by the incidence of people unexposed and incidence of non-disease in those exposed.

OR >1 = the factor being studied is a risk factor for the outcome

OR < 1 = the factor being studied is a protective factor in respect to the outcome

OR = 1 = no significant difference in outcome in either exposed or unexposed group

Relative Risk → [a/(a+b) / d/(c+d)]

Compares disease risk in people exposed to a certain factor with disease risk in people who have not been exposed

Attributable Risk → [a/(a+b) – d/(c+d)]

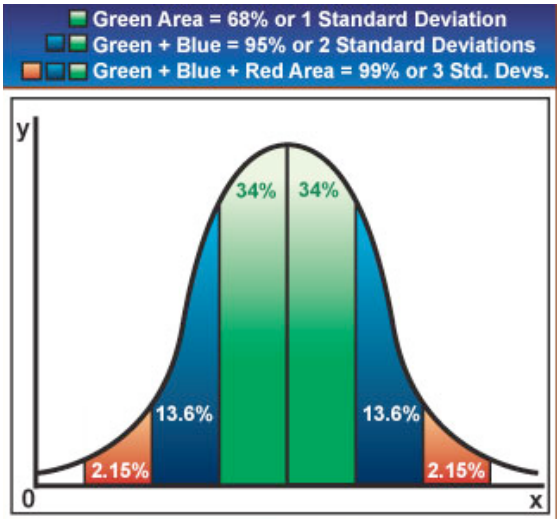
The attributable risk is the # of cases attributable to one risk factor

Standard Deviation

1 standard deviation – 68% fall within 1 SD

2 standard deviations – 95% fall within 2 SD's

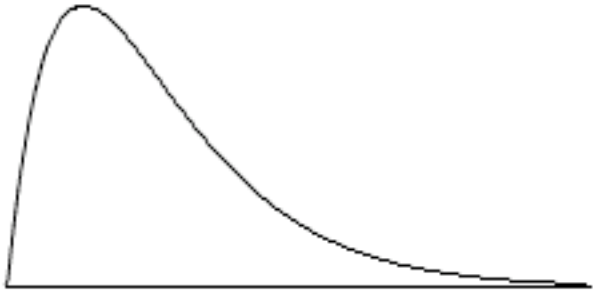
3 standard deviations – 99.7% fall within 3 SD's



MEAN – the average value

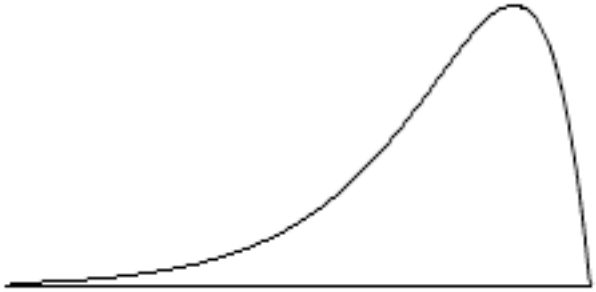
MEDIAN – the middle value

MODE – the most common value



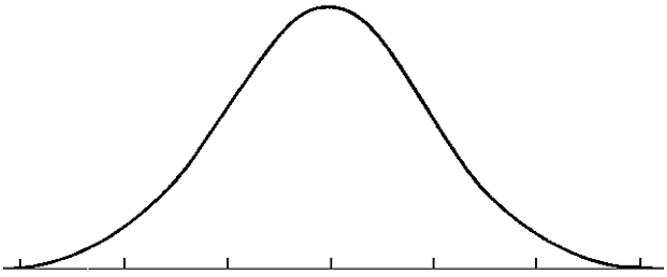
+VE SKEW

A +ve skewed graph means the ***mean>median>mode***



-VE SKEW

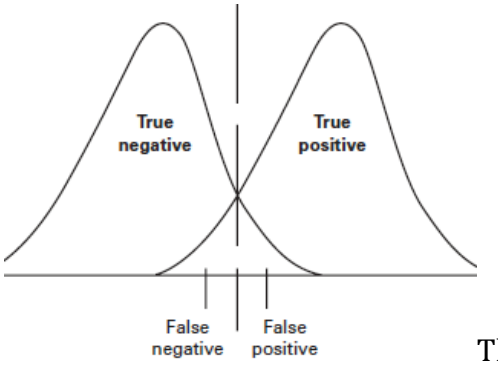
A –ve skewed graph means the ***mean<median<mode***



Normal bell curve

Mean = median = mode

NO DISEASE DISEASE



This chart represents sensitivity & specificity

- If the cutoff point for a disease is moved from false (+) → false (-), there will be an ⇑ in the # of positive results. Thus an ⇑ in sensitivity (TP⇑, FP⇑, FN⇓, PPV⇓]
- If the cutoff point is raised from the false (-) → false (+), this will ⇑ specificity [TN⇑, FP⇑]

Correlation co-efficient

Measures to what degree the variables are related (from -1 to +1)

0 = there is no correlation

+1 = there is a perfect correlation (thus if 1 variable increases so does the other)

-1 = there is a perfect negative correlation (thus if 1 variable increases the other decreases)

Confidence interval and p-value

Two values used to strengthen a finding of a study. For statistical significance, the confidence interval must not contain the null value (RR=1). Further, statistically significant results have a p-value <0.05 (meaning there is <5% chance that the results obtained were due to chance alone).

A p-value <0.05 is generally used as a cutoff for statistical significance in medicine. 0.05 means there is a ≤ 5% chance that results obtained are due to random chance. When the p-value is ≤ 0.05 we reject the null hypothesis (null hypothesis says that a result is due to random error or chance)

The confidence interval is given in 2 digits, and the closer they are, the more confidence there is. * With increased subjects there is a tighter confidence interval

Attributable Risk Percent (ARP)

Measures the impact of a risk factor being studied. The ARP represents the excess risk in a population that can be explained by exposure to a particular risk factor.

Calculate ARP: [(RR -1)/RR]

Incidence vs Prevalance

Incidence → the # of new cases of a disease over a unit of time

Prevalence → is the total # of cases of a disease (new or old) at a certain point in time

If a disease is treated only to prolong life without curing the disease (ie. Terminal cancers), then incidence remains the same but prevalence increases.

** In short term diseases: incidence > prevalence

** In long term diseases: prevalence > incidence

Reliability → gives similar or very close results on repeat measures

Validity/Accuracy → defined as a test's ability to measure what it is supposed to measure (as compared to the gold standard)

Precision → is increased with a tighter confidence interval, and CI is made tighter with a higher # of subjects

Study Types

Case-Control/Experimental → Is the gold standard, compares 2 equal groups where one has a changed variable

Prospective → Also known as: *Cohort, Observational, Incidence*. Takes a sample and divides it into 2 groups based on presence or absence of risk factor, and follows over time to see what develops. ** These are time consuming and expensive.

Retrospective → Chooses a population (after the fact) based on the presence or absence of a risk factor. ** Costs less, less time consuming, better for rare diseases.

Case Series → describes the clinical presentation of people with a certain disease

Cross-Sectional/Prevalence → This study looks at the prevalence of disease and the prevalence of risk factors. Takes sample from a population at one point in time. This compares 2 different cultures.

Epidemic → When the observed incidence greatly exceeds the expected incidence

Pandemic → Is an epidemic seen over a wide geographical area.

Test Methods

Two-sample T-test: is used to compare the means of 2 groups of subjects

ANOVA (analysis of variance): used to compare ≥ 3 variables

Chi-squared: compares the proportions of a categorized outcomes (2x2 table). If the difference between the observed and expected values is large, an associated between the exposure and the outcome is assumed to be present.

Meta-Analysis: is a method of pooling the data from several studies to do an analysis having a big statistical power.

Types of Bias

Selection Bias: Bias type due to manner in which people are selected, or from selective losses from follow-up

Observer & Measurement Bias: Distortion of measurement of association by misclassifying exposed, unexposed, and/or diseases/non-diseased subjects.

Recall Bias: Results from the inaccurate recall of past exposure by people in the study

Hawthorne Effect: Patients change their behavior because they know they are being studied

Confounding: Is bias that results when the expose/disease relationship is mixed with the effect of extraneous factors. (ex. Study of the association of smoking and cirrhosis, and find that there is a strong association. Then the division of drinkers and non-drinkers finds there's no association of smoking to cirrhosis. In this case, alcohol is the confounder). * Matching is an effective way of controlling confounding

Lead Time Bias: Refers to the chronology of the diagnosis and treatment between different cases. (ex. Testing of platelet inhibitors in pilots vs autoworkers, not fair because pilots are undergoing constant health screening)

Admission Rate Bias: Refers to distortion in risk ratio due to different hospitals admission of cases

Unacceptability Bias: Occurs when participants purposely give desirable responses which lead to

Chapter 6

Psychiatry

Mood Disorders

Major Depressive Disorder

Major depressive disorder is characterized by a depressed mood or anhedonia (cannot enjoy things that they once enjoyed), and depressive symptoms lasting at least 2 weeks.

- Look for other possible causes such as hypothyroidism, drug use/substance use.

The classic mnemonic for depression is SIG E CAPS

S: sleep disturbances (too much or too little)

I: interest changes (loss of interst)

G: guilty feelings

E: energy loss

C: concentration disturbances

A: appetite changes (causes weight changes too)

P: psychomotor changes

S: suicidal thoughs/death

- If patient is suicidal or dangerous to others always admit

Treatment:

- 1st line treatment is SSRI such as fluoxetine, paroxetine, sertraline (possible side effect is sexual dysfunction)
- Can include benzodiazepine if patient is agitated
- Therapy is also indicated along with SSRI treatment

Dysthymic Disorder

Same symptoms as major depressive disorder except is more low-level in nature, and is present on most days for at least 2 years.

Treatment:

- SSRI (similar treatment as Major Depression)

It is possible to get a major depressive episode while dysthymic. Treat as MDD in this situation.

Bereavement

- Bereavement is commonly seen after death of a family member (most commonly seen in older people after death of a spouse).

Diagnosis:

- Key to the diagnosis is the time that has elapsed since the onset of the bereavement period.
- Symptoms > 2months makes the diagnosis major depressive disorder instead of normal bereavement.

Treatment:

- Therapy (grief management) is recommended in such conditions

Bipolar Disorder

- A condition with episodes of mania, depression, as well as normal periods.
- Seen in approximately 1% of the population
- Affects males = females
- More common in the younger population
- A mix of mania, depression, or mixed symptoms for at least 1 week

Signs and Symptoms:

- Acute onset of ⇑ energy
- ⇓ need to sleep

Remembering MANIA:

D – distractibility

I - insomnia

G – grandiosity

F- flight of ideas

A – activity increased

S – speech (pressured)

T – takes risks

- Pressured speech
- ⇓ attention span
- hypersexuality
- Reckless behavior (excessive gambling, shopping, spending money)

Differential diagnosis:

- Schizophrenia
- Intoxication (cocaine, amphetamine)
- Certain personality disorders

Diagnosis:

- Episode should last ≥ 1 week and should be abrupt/cause significant disability
- Bipolar 1 → a manic episode with or without depressive episode
- Bipolar 2 → depressive episodes with hypomanic episodes
- Rapid cycling → > 4 episodes in a one-year period

Treatment:

- Hospitalization (is often involuntary because patient is manic)
- Mood stabilizers – Lithium is DOC, can also use valproate or carbamazepine
- Antipsychotics can be used until acute mania is controlled
- If recurrent episodes of depression are present, can give antidepressants only in conjunction with mood stabilizers
- Lithium levels should be checked to prevent toxicity

Cyclothymia

- Is a recurrence of depressive episodes and hypomania for at least 2 years
- Is a milder form of bipolar disorder

Treatment:

- 1st is psychotherapy because many patients can function without medication
- If functioning becomes impaired start patient on valproic acid, which is more effective in cyclothymia than lithium

Drug-Induced Mania

- The most common causes are cocaine and amphetamines

Signs and Symptoms:

- Findings similar to mania
- Mydriasis
- Hypertension
- MI in young people (highly suggestive of cocaine overdose)

Treatment:

- For acute symptoms give CCB's
- Drug treatment programs for long-term management

Post-partum Depression

	Post-partum blues "Baby blues"	**Postpartum Depression**	**Postpartum Psychosis**
Onset	After any birth	MC after 2nd birth	Usually after 1st birth
Mother's emotions toward the baby	Mother still cares about the baby	Thoughts about harming the baby are common	Thoughts about harming baby are common
Symptoms	Mild Depression	Severe Depression	Depressive symptoms + psychotic symptoms
Treatment	No treatment necessary	Antidepressants	If patient not breastfeeding give Mood stabilizers OR antipsychotics + antidepressant If patient is breastfeeding do ECT

PSYCHOSIS

Psychosis is characterized by:

- Hallucinations – false sensory perception that is NOT based on real stimuli
- Delusions – false interpretations of external reality
- Can be of the paranoid nature, grandiosity, religious, or ideas of reference

This table gives a general overview of the causes of psychosis

DISEASE	CHARACTERISTICS
Schizophrenia	• There is a strong genetic predisposition, onset usually late teens through the 20's • +ve symptoms = hallucinations and/or delusions • -ve symptoms = flattened affect • Other symptoms include disorganized behavior and/or speech • Must last ≥ 6 months to be called schizophrenia • If lasting 1-6 months called schizophreniform • If lasting <1 month it is a brief psychotic disorder (these patients often return to normal baseline functioning)
Schizoaffective disorder	• Combination of a mood disorder + schizophrenia
Delusional disorder	• Patient gets non-bizarre delusions
Mood disorders	• Bipolar and/or depression can cause delusions and in extreme cases may cause hallucinations
Delirium	• Often seen in patients who have underlying conditions • No orientation to person, place, or time • Waxing and waning of condition • Treatment involves treating the underlying condition
Drugs	• Cocaine/amphetamines cause paranoid delusions and formication (sensation of bugs crawling on the skin) • LSD/PCP cause hallucinations of vision, taste, touch, and scent
Medical causes	• Endocrine disorders, metabolic disorders, neoplastic disorders, and seizure disorders can

	cause psychosis

Treatment:

- If condition is disabling or potentially dangerous to patient or others, hospitalization is required.
- Pharmacologic therapy is with dopamine antagonists, and the differences amongst the drugs is based on the side effects they produce
- Improve drug compliance by giving depot form of haldol
- Psychotherapy to improve social functioning (behavioral treatment to improve social skills, family-oriented treatment for improved familial functioning)
- Prognosis is dependent of frequency of episodes as well as accompanying symptoms (presence of negative symptoms usually indicates a poor prognosis)
- Patients who were very high-functioning prior to the psychosis onset have a better prognosis

DRUG	**ADVERSE EFFECTS**
Typical Antipsychotics	
Chlorpromazine	Low potency, ⇑anticholinergic effects, ⇓ movement disorders
Haloperidol	High potency, ⇓ anticholinergic effect, ⇑ movement d/o
Atypical Antipsychotics	
Clozapine	For refractory disease, give weekly CBC (agranulocytosis risk)
Risperidone	1st line, minimal averse effects
Olanzapine	1st line, minimal adverse effects

There are many possible movement disorders associated with the use of antipsychotic medications. You will likely encounter one on the CK exam. This table will demonstrate the timeline for certain adverse movement reactions.

DISORDER	TIME FRAME	CHARACTERISTICS
Acute Dystonia	From 4hr – 4 days (4&4)	• Patient experiences sustained spasms, may be anywhere but MC seen in the neck, jaw, or back. • Treatment – IV diphenhydramine (immediately)
Parkinsonism	From 4 days – 4 months	• Patient has cog-wheel rigidity, resting tremor, and shuffling

		gait • Treatment – benztrophine (anticholinergic used in Parkinson's disease)
Tardive Dyskinesia	4 months – 4 years	• Involuntary/irregular movements of the head, tongue, lips, limbs, and trunk • Treatment – change medications immediately (is a permanent condition)
Akithisia	May occur at any time during treatment	• Patient has a sense of discomfort/restlessness • Treat by lower the dose of medication
Neuroleptic Malignant Syndrome	May occur at any time during treatment	• Is a life-threatening muscle rigidity with fever, increased BP and HR, and rhabdomyolysis that appears over 1-3 days • Treatment is supportive, stop all offending drugs immediately, give patient dantrolene (Calcium is inhibited from release into cells), and cool the patient

Anxiety Disorders

Panic Disorder

- A condition seen MC in women in their mid 20's
- Symptoms mimic those of an MI (chest pain, palpitations, diaphoresis, nausea, anxiety, sense of impending doom)
- Symptoms usually escalate for approximately 10 minutes and last at least 30 minutes
- This disorder is very unpredictable, if it occurs in the same type of setting then suspect a specific phobia

Diagnosis:

- Must differentiate from drug use, MI, and other sources of phobias
- Diagnosis of exclusion

Treatment:

- Cognitive-behavioral therapy and/or relaxation training.
- Relaxation is more useful if patient has an agoraphobic tendency
- SSRI's and benzodiazepines can be prescribed

Agoraphobia

- Patient fears being in situations where they cannot escape, bringing about a panic attack
- Patients develop agoraphobia because of recurrent and unexpected panic attacks in certain situations

Diagnosis:

- Is clinical, looking for evidence of social and/or occupational dysfunction

Treatment:

- Exposure desensitization
- β-blockers as prophylaxis from sympathetic activation when in possibly triggering situations

Obsessive-Compulsive Disorder (OCD)

- Patient experiences recurrent thoughts and performs recurrent actions/rituals as a coping mechanism
- Obsessive thoughts provoke anxiety, compulsions are a way of dealing with this anxiety, this anxiety relief is only temporary and thus rituals get performed over and over again.
- Commonly involve cleanliness (fear of contamination) – thus excessive hand-washing is common

Do not confuse OCD with obsessive-compulsive "personality disorder", where the patient sees no wrong in their behaviors.

Diagnosis:

- Patient must be aware of the abnormality of their behavior, and must be disturbed by this.

Treatment:

- 1st line treatment is SSRI
- 2nd line is clomipramine
- Patient must undergo psychotherapy as well, where they are forced to overcome their behavior

Post-Traumatic Stress Disorder

- This is the classic "Vietnam vet" patient, who has undergone a traumatic incident that leaves them emotionally scarred

There are 3 key groups of symptoms:

1. Avoidance of stimuli – associated with their trauma or numbing of responsiveness because it emits emotional pain
2. Re-experiencing the traumatic event – via dreams, thoughts, recollections.
3. Increased arousal – seen as sleep disturbances, emotional lability, impulsiveness, anxiety.

Diagnosis:

- Always differentiate from an acute stress disorder, where symptoms last less than 1 month and occur within 1 month of experiencing the stressor
- Diagnosis requires a traumatic incident and must last longer than 1 month

A patient who functioned very well before the onset of PTSD has a greater prognosis than someone who was less functional.

Treatment:

- When patient is in acute distress, give benzodiazepines to calm them down
- For long-term therapy, give SSRI's + psychotherapy

Generalized Anxiety Disorder

- Patient worries excessively and/or has poorly controlled anxiety on most days for at least 6 months.
- There is no specific event or reason for this anxiety
- Patient has trouble sleeping, the inability to concentrate, excessive fatigue and restlessness
- Be sure to distinguish from specific phobia/anxieties or other causes of anxiety.

Diagnosis:

- Must be evidence of social dysfunction (which rules out normal anxiety)

Treatment:

- Psychotherapy teaching patient to recognize their worrying and finding a way to manage through thought patterns and behavior
- Can give SSRI's, buspirone, and benzodiazepines
- β-blockers to block excessive sympathetic activation

ANXIOLYTICS PRESCRIBED FOR ANXIETY DISORDERS:

PANIC DISORDER: SSRI, Alpralozam, Clonazepam

GAD: SSRI's

OCD: SSRI's and clomipramine

ADJUSTMENT DISORDER: Benzodiazepines

SOCIAL PHOBIA: SSRI + buspirone

Personality Disorders

Some general characteristics of Personality Disorders:

- They cause functional impairments
- Behavior often causes significant disruption to others (co-workers, classmates, family members, etc)
- Patients usually see no problem with their behaviors

CLUSTER A DISORDERS – Paranoid, Schizoid, Schizotypal (Eccentric/Weird)

CLUSTER B DISORDERS – Borderline, Antisocial, Histrionic, Narcissistic (Dramatic/Aggressive)

CLUSTER C DISORDERS – Narcissistic, Avoidant, Dependent, Obsessive-Compulsive (Shy/Nervous)

CLUSTER A DISORDERS

***Paranoid*:**

- These people negatively interpret the intentions of others
- Often use projection as their main ego defense

Schizoid:

- These people are socially withdrawn and introverted
- Avoid forming close emotional connections with others

Schizotypal:

- These people believe in things not normally accepted by society, such as magic
- May have brief psychotic episodes but are not psychotic
- Socially isolated
- Many schizotypal patients have schizophrenic relatives

CLUSTER B DISORDERS

Antisocial:

- Break the law, violate other's rights
- Often seductive in nature
- ***Must be 18yr*** of age for diagnosis + must have been this way since at least 15 yr old (conduct disorder)

Borderline:

- Exhibit self-destructive behavior such as cutting
- Emotionally volatile
- "Splitting" ego defense commonly used (people seen as either great or terrible)
- Have the ability to dissociate from past negative experiences

Histrionic:

- Attention-seekers
- Sexually promiscuous and uses physical appearance for attention
- Very dramatic and exaggerate their behaviors
- Use ego defenses such as dissociation and repression

CLUSTER C DISORDERS

Narcissistic:

- Believe they are superior and are entitled to the best
- Do not handle criticism well

Avoidant:

- Patient feels sensitive and does not handle negative comments well
- Scared to try new things or make new friends for fear of embarrassment

Dependent:

- Scared to be on their own and cannot do much on their own
- Require help with decisions from someone else

Obsessive-Compulsive:

- This person is overly preoccupied with rules, regulations, neatness, etc
- They commonly isolate themselves (ego defense) in order to avoid demonstrating emotions

Ego Defenses

Acting Out: transformation of unacceptable feelings into actions (ex. Tantrums)

Identification: copies the behavior of someone else

Rationalization: a way of making something unacceptable seem acceptable (ex. Boyfriend breaks up with girlfriend and she says she wanted to end it anyway)

Reaction Formation: expressing outwardly the exact opposite of how you feel (ex. Someone addicted to something starts a charity to fight that cause)

Intellectualization: trying to logically explain something in order to make sense of it

Regression: resorting to immature/child-like behavior

Sublimation: funneling unacceptable feelings into positive actions (ex. Funneling sexual feelings into a workout regimen)

Somatoform & Factitious Disorder

Somatoform Disorders

Somatization disorder:

- Most commonly female patients and starts before 30 years of age
- Frequently visits the doctor for many procedures and operations
- Often have a history of abusive and/or failed relationships

Symptoms:

- Somatic complaints involving many different systems, such as:

  GI → nausea, vomiting, diarrhea

  Neurologic → weakness, loss of sensation that is not explained by normal anatomy

  Sexual → irregular menses, etc

- Lab findings do not explain any of the complaints

Somatization and conversion disorder are never intentional. If a question says patient is looking for gain or did something purposely, these two are not the right answer.

Diagnosis:

- Must always rule out medical conditions
- Rule out material gains

Treatment:

- Important to form a strong bond with the patient
- Try to bring to light the fact that there are psychological causes for the condition
- Schedule regular appointments
- Perform a physical exam but do not order lab tests

Conversion disorder:

- Patient experiences neurologic symptoms that cannot be explained by medical or neurological disorder
- Patients are often not overly concerned about the impairment – know as "la belle indifference"
- There are often psychological factors associated with symptoms, such as going limp when someone yells at them

Treatment:

- Formation of a strong relationship with the patient
- Psychotherapy

Hypochondriasis:

- The patient falsely believes they have a specific disease even when they are ruled out with negative workups and/or lab tests

Treatment:

- Regular visits to ONE primary doctor
- Avoid tests/procedures
- Provide psychotherapy
- SSRI's may be useful in some cases

Factitious Disorders

- These patients have intentionally feigned their symptoms
- These patients often see many doctors and visit many different hospitals
- They often have more medical knowledge than the average person (often healthcare workers)

Factitious disorder: purposely faked but not for obvious gain

Malingering disorder: purposely faked for an obvious gain, such as medication, insurance, etc.

- Very demanding of treatment

- A factitious order "by proxy", is made when signs and symptoms are faked by another person (ex. Mother makes up symptoms in her child – known as Munchhausen's by proxy)

Munchhausen's syndrome:

- A factitious disorder mainly with physical symptoms

Munchhausen's by proxy:

- Someone claims non-existent symptoms (MC in their child)
- Motivation is usually to assume the role of caretaker

Diagnosis:

- By exclusion of a real medical condition
- Differentiate between malingering and factitious disorders

Treatment:

- Very difficult, patients often very defensive when it is suggested that they are faking

Childhood and Adolescent Psychiatry

Autism

- Seen in 0.02%-0.05% of children
- Onset before 3yr of age
- Is 3-5x more common in boys
- Develop severe problems in communication
- Have normal hearing
- Significant problems in forming social relationships
- Are comfortable performing repetitive behaviors
- Often perform self-destructive behaviors
- Have subnormal intelligence (<70 IQ) in approximately 2/3 of all patients
- Some have unusual specific abilities
- Prognosis is not good, only 2% are able to work and live independently, but most remain severely impaired in adulthood

Treatment/Management:

- Behavioral therapy to increase social/communicative skills, decrease behavioral problems, and improve their self-care
- It is often more beneficial for the parents, because they have much difficulty raising a child with autism.

Asperger disorder

- This disorder is first seen at 3-5 years of age
- More common in boys
- They have significant problems forming social relationships
- Little or no delay in cognitive or language development
- Prognosis is much better here than it is in Asperger

Childhood Depression

- Presents differently depending on the age group
- Preschoolers may be aggressive and/or hyperactive, while adolescents are irritable or show antisocial behavior
- Important to note that they may also show the same symptoms that adults do when experiencing a major depressive disorder

Treatment:

- Family therapy may be required because this is often a cause of childhood depression
- Use of antidepressants is very controversial in children and teens due to their risk of suicide in this age group

Separation Anxiety

- Child is too attached to parents or other figures in their life
- Child has excessive worry that these figures will be separated from them

Signs and Symptoms:

- Somatic symptoms during times of seperation
- Trouble sleeping

Treatment:

- Desensitization
- Imipramine may be used in some cases

Oppositional Defiant Disorder/Conduct Disorder

Oppositional Defiant:

- Patients are argumentative and temperamental (more so with people close to them)
- Often have no friends and perform poorly in school

Conduct Disorder:

- Patient is a bully to others
- Shows physical cruelty to animals
- Violates and destroys other people's property, steals.

- Has no remorse for their actions
- Family history often shows negligence, and abuse of drugs and/or alcholol
- This may lead to conduct disorder (but not always)

Treatment:

- For both oppositional defiant disorder and conduct disorder, create an atmosphere/setting with strict rules and consequences for not obeying these rules

Attention Deficit Hyperactivity Disorder

- Characterized by overactivity, a limited attention span, poor self-control, impulsiveness, emotional lability, high sensitivity to stimuli, sleep problems

Diagnosis:

- Onset must be before 7yr of age
- ≥ 6 symptoms from both hyperactivity and/or inattention sub-categories

Treatment:

- CNS stimulants are DOC
- Methylphenidate in children >6yr of age
- Other types of CNS stimulants also given
- Note the adverse effects of CNS stimulants can be the inability to gain weight and the inhibition of growth

Tourette's Disorder

- Characterized by involuntary tics, repetitive movements, and vocalizations
- Diagnosis MUST include both a motor tic and a vocal tick that is present for ≥1 yr
- The common stereotype of Tourette's involves uncontrollable swearing, which is known as coprolalia

Treatment:

- Haldol is very effective, but is not used in milder cases
- Psychotherapy is effective in dealing with the social aspects of this disorder, but it does not improve the tics

Anorexia Nervosa

- Often start during adolescence
- There is a profound disturbance in body image and in a person's self-worth

Signs and Symptoms:

- Patients are below the ideal weight for their age and height
- They often have mealtime rituals such as cutting their food into tiny pieces and/or re-arranging it on the plate
- Amenorrhea occurs secondary to the weight loss, and is required for the diagnosis of anorexia

Treatment:

- Hospitalization may be required to restore the patient's weight to a safe level, as well as correct any electrolyte imbalances
- The most severe adverse reaction is cardiac dysfunction
- The mainstay of treatment is psychotherapy

Prognosis is poor if preoccupations with food and weight do not improve

Bulimia Nervosa

- More common than anorexia
- Characterized by binge eating (with a perceived lack of control)
- Often accompanied by purging (laxative use and/or vomiting)
- Often have a normal appearance and normal weight
- Often have cuts on the hands from shoving them down the throat to induce vomiting
- Dental erosions seen due to acidic destruction from constant vomiting

Treatment is same as that for anorexia

Dissociative Disorder (multiple personality disorder)

- A patient possesses different personalities that can each take control at any given time
- Childhood trauma is very common when this condition is present
- Treatment is focused on the gradual integration of these personalities

Two different disorders that should be taken into consideration:

Dissociative Amnesia:

- Person forgets plenty of personal information

Dissociative Fugue:

- A syndrome where someone travels to another place with the inability to remember the past and confusion about their present identity

Adjustment Disorder

- A stressful life event leads to the inability to deal emotionally and/or behaviorally

Diagnosis:

- Symptoms present within 3 months of the stressful event and must disappear within 6 months of the disappearance of the stressor
- Differentiate from a bereavement disorder
- Always aggressively look at whether there is a depressive disorder and/or anxiety disorder, which must be treated

Impulse-Control Disorders

- Patients are unable to resist the drive to perform actions that may be harmful to others and themselves
- There is a feeling of anxiety before performing the impulsive action and a sense of gratification afterwards

Intermittent-Explosive Disorder

- Patient shows aggressive behavior that is way out of proportion to the stressor
- Must not be associated with drug use
- Treat with SSRI's AND a mood stabilizer such as Lithium

Kleptomania

- An individual who repeatedly steals to relieve anxiety
- Person does not steal because they need the object
- Often, the person returns the object after stealing it

Pyromania

- Individual purposely sets fires
- There is no personal gain in pyromania, nor is there any anger in relation to doing this (if there is, this shifts the diagnosis to conduct/antisocial disorder)

Trichotillomania

- Patient impulsively pulls out their hair
- This results in *observable* hair loss

Drugs of Abuse

Alcohol

- Alcohol is a commonly abused drug
- Patients develop different levels of dependence
- ***Alcohol dependence*** is the frequent use of alcohol that results in tolerance, leading to psychological and physical dependence.
- ***Alcohol abuse*** is diagnosed when its use results in failure to perform normally in society (loss of job, social impairment, legal problems)

Diagnosis:

- Lab tests are not required for diagnosis
- The CAGE questionnaire is the most accurate diagnosis

Treatment:

- The most effective management of an alcoholic is always alcoholics anonymous

Management:

- For outpatient management, the first thing is to prevent further intake of alcohol
- If patient is intoxicated prevent them from operating machinery (driving)
- If patient is agitated sedate
- Admit to hospital if patient requires further help

The following table presents the most commonly abused drugs

SUBSTANCE	Si/Sx of intoxication	Treatment of intoxication	Si/Sx of withdrawal	Treatment of withdrawal
Alcohol	Lack of inhibition Talkative	If severe give mechanical ventilation	Tremor Seizures Delirium	Long-acting benzodiazepines.
Amphetamines/ Cocaine	Agitation Mydriasis, Euphoria, Hyperactivity, Stroke/MI	ST antipsychotics	Anxiety Tremor Hyperphagia Depression Suicide risk	Antidepressants
Marijuana	Impaired motor co-ordination, Hyperphagia, Dry mouth, Conjunctival redness	None	None	None
Hallucinogens	Ideas of reference, Hallucination, Dissociative symptoms	Talking down, Antipsychotic, Benzos	None	None
Inhalants	Belligerence, Violent, Impaired judgement, Blurred vision, Stupor, coma.	If delirious or agitated give antipsychotics	None	None
Heroine (opiates)	Dysphoria, Miosis, Drowsiness, Slurred speech	Naloxone	Fever, chills, Abd cramps, Insomnia, Lacrimation	Clonidine, Methadone
Phencyclidine (PCP)	Violent, Panic, Agitation, Nystagmus.	Talk-down, Benzos, Antipsychotics Respiratory support	None	None
Barbiturates	Impaired	Flumazenil	Autonomic	Long-acting

and/or Benzodiazepines	memory or concentration, Lack of inhibition		hyperactivity, Tremor, Insomnia, Seizure, Anxiety.	barbiturates as substitution

Paraphilias

- Involve recurrent, sexually arousing preoccupations that are focused on humiliation and/or suffering and the use of nonliving objects and nonconsenting partners.
- Occurs for >6 months
- Causes social impairment
- Treatment for all is psychotherapy and aversive conditioning
- Severe cases may require anti-androgens or SSRI's to reduce patient's sex drive

Types:

Frotteurism: Touching or rubbing against a non-consenting partner

Exhibitionism: Recurrent urge to expose themselves to strangers

Pedophilia: Urges or arousal toward prepubescent children (is the most common paraphilia)

Voyeurism: Urges to observe an unsuspecting person who is having sex or taking off their clothes

Fetishism: The use of nonliving objects associated with the human body (shoes are common)

Masochism: Recurrent urge or behavior involving being humiliated

Sadism: Causing suffering to a victim is exciting to the patient

Sleep

Normal Sleep

There are two types of sleep:

1. Non-REM (NREM), which has four stages
2. REM – rapid eye movement

The stages of normal sleep

Stages	Characteristics
NON-REM	This stage consists of early, slow-wave sleep
Stage 1	Consists of α-waves and σ-waves
Stage 2	Sleep spindles are present
Stage 3,4	δ-waves are present during these stages
REM	Dreaming occurs here (this stage is affected by elicit drugs and ETOH)

Sleep Disorders

Insomnia

- Patient is unable to fall asleep or stay asleep
- Recurrent over more than a 1-month period
- May be associated with periods of stress, anxiety, or drug use

Treatment:

- A sleep schedule is important to regular internal sleep patterns
- Exercise
- Antihistamines
- 2-week period of benzodiazepines (careful to avoid dependence)

Hypersomnia

***Narcolepsy*:**

- Patient experiences acute attacks of REM sleep
- They suddenly collapse with a complete loss of muscle tone (cataplexy)

Treatment:

- CNS stimulants

Sleep Apnea:

- Apneic periods that occur during sleep
- Most commonly is obstructive (commonly due to excess weight)

Treatment:

- Weight loss
- Continuous positive airway pressure (CPAP)
- If patient doesn't get relief from these then should undergo surgery since sleep apnea is a life-threatening condition

Pickwickian Syndrome (Central Alveolar Hypoventilation)

- A syndrome with somnolence, obesity, and erythrocytosis
- Patient gradually develops hypercapnea, hypoxemia, and erythrocytosis
- This is caused by the weight of excess adipose tissue pressing on the lungs

Treatment:

- Weight loss

Parasomnias

Night Terrors:

- Child arises during NREM sleep, is not aware they are awake, screams in terror, then falls back asleep.
- They do not remember the occurrence when they awaken

Nightmares:

- Occur during REM sleep
- Related to emotional events such as tragedy, scary movie, etc
- Patient remembers the dream

Sleep Walking:

- Occurs during NREM sleep
- Patient gets out of bed and wanders about
- Patient has no recollection of the event

Chapter 7

Cardiovascular

Pleuritic Chest Pain	Positional Chest Pain	Tender Chest Pain
PE	Pericarditis	Costochondritis
Pneumonia		
Pleuritis		
Pericarditis		
Pneumothorax		

Ischemic Heart Disease (CAD)

Major Risk Factors:

- Diabetes
- Smoking
- Hypertension (HTN)
- Hypercholesterolemia
- Family history
- Age

Minor Risk Factors:

- Obesity
- Lack of estrogen (this is why it occurs in men more than women)

The #1 preventable RF is smoking

Stable Angina

- Chest pain that occurs with activity
- Caused by atherosclerosis, whereby the supply of 02 required by the heart is not met

Signs and Symptoms:

- Chest pain that may radiate to the left arm, jaw, and back.
- Relieved by rest and nitroglycerin
- EKG will show ST-segment depression and T-wave inversion

Diagnosis:

- Made by clinical presentation and based on symptoms

Treatment of Angina

Acute	• Sublingual nitroglycerin (acts in 1-2 min) • May take nitro up to 3 times every 3-5 minutes • Lack of relief may indicate infarction in progress
Chronic Prevention	• Long-acting nitrates for prophylaxis • β-blockers ⇓ myocardial 02 consumption when stressed • Aspirin to prevent PLT aggregation in atherosclerotic plaque • Smoking cessation • ⇓LDL ⇑HDL through diet
Endovascular Intervention	• Percutaneous transluminal coronary angioplasty • Indicated when there's a failure in medical management • Stent placement can reduce re-stenosis by 20%-30% • GPIIb-IIIa antagonists further reduce stenosis rate
Surgery	• Coronary artery bypass graft • Indicated when medical treatment fails

<u>Unstable Angina</u>

- Symptoms are similar to angina but occur more frequently and without any relation to exertion/activity, occurring at rest
- Unstable angina = ischemia
- Unstable angina and non-ST-elevation MI are a closely related
- EKG during ischemia usually shows ST-segment depression or T-wave inversion
- LABS are (+) for cardiac enzymes

Treatment:

- Based on the likelihood that it will progress to a potentially fatal outcome (ie. Risk of recurrent unstable angina, infarction, or death 30 days after presentation).

Prinzmetal's Angina

- Is caused by a coronary artery vasospasm
- EKG shows ST-segment elevation
- ST elevation is transient and cardiac enzymes are usually negative, which helps differentiate from an MI
- Treatment is vasodilators (nitroglycerin or CCB's)
- Patient should undergo catheterization because vasospasm often occurs at the site of an atherosclerotic lesion in the coronary arteries.

ST Elevation Myocardial Infarction (STEMI)

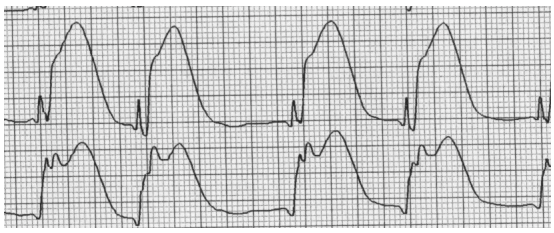
- Infarction usually secondary to acute thrombosis in an atherosclerotic vessel

Signs and Symptoms:

- Crushing substernal pain that is not relieved by rest
- Diaphoresis
- Nausea/vomiting
- Tachycardia or bradycardia
- Dyspnea

Diagnosis:

- EKG will show ST elevation and Q waves



- Cardiac enzymes elevated (CK-MB, troponin I) – CK-MB normalizes within 72hr

Treatment:

- Re-establish vessel patency
- #1 priority → aspirin (proven to ⇓ mortality)
- #2 priority → β-blocker (proven to ⇓ mortality)
- Statins to lower cholesterol (goal is to get LDL <100 post-infarct)
- 02 + morphine (pain control)
- Nitro to reduce preload and afterload
- ACEI's are excellent late and long-term therapy (⇓ afterload and prevent remodeling)
- Consult about smoking cessation

Post-MI Discharge Instructions:

- ASA
- β-blocker
- Statin
- ACEI

EKG findings and Arrhythmias

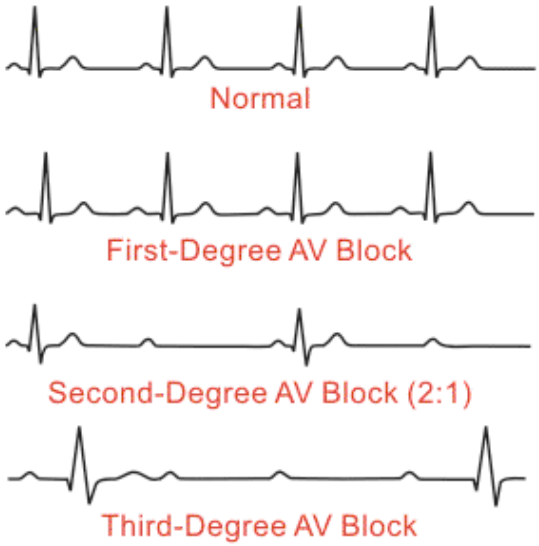
Heart Blocks:

First-degree AV block → normal sinus rhythm with PR interval ≥ 0.2ms

Second-degree, type 1 (Weckenbach) block → PR interval elongates from beat to beat until a PR is dropped

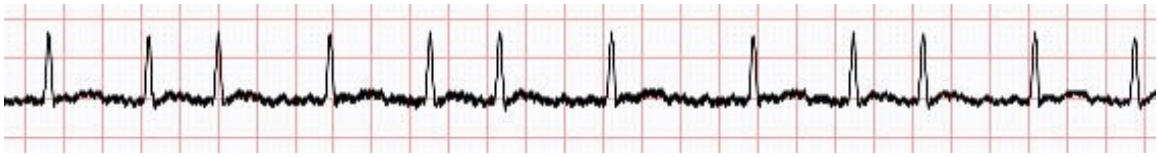
Second-degree, type 2 (Mobitz) block → PR interval fixed but there are regular non-conducted P-waves leading to dropped beats

Third-degree block → no relationship between P waves and QRS complexes. Presents with junctional escape rhythms or ventricular escape rhythm



Atrial Fibrillation

- The most common chronic arrhythmia
- From ischemia, atrial dilatation, surgical history, pulmonary diseases, toxic syndromes
- Classically, the pulse is irregularly irregular



Signs and Symptoms:

- Chest discomfort

- Palpitations
- Tachycardia,
- Hypotension + syncope

Treatment:

- Control rate with β-blockers, CCB's, and digoxin (not acutely)
- If fibrillations last >24hr then should anticoagulate with warfarin for at least 3 weeks before cardioversion (prevents embolisms)
- If you cannot convert to normal sinus rhythm, the patient will require long-term anticoagulation. 1st line is warfarin, 2nd line is aspirin

Cardioversion to convert to normal rhythm:

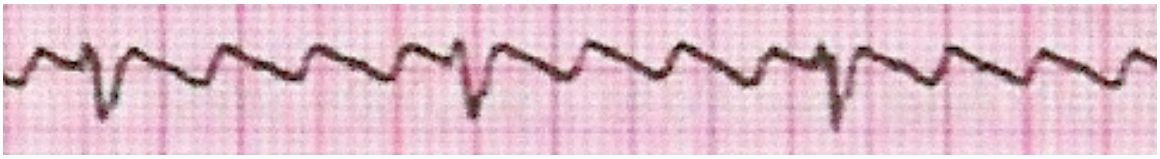
1st line → IV procainamide, sotalol, amiodarone

Electrical → shock of 100-200J followed by 360J

Atrial Flutter

- Less stable than Afib
- The rate is slower than that of atrial fibrillation (approximately 250-350bpm)
- Ventricular rate in atrial flutter is at risk of going too fast, thus atrial flutter is considered to be more dangerous (medically slowing this rate can cause a paradoxical increase in ventricular rates)
- Classic rhythm is an atrial flutter rate of 300bpm with a 2:1 block resulting in a ventricular rate of 150bpm
- Signs and symptoms similar to those of atrial fibrillation
- Complications include syncope, embolization, ischemia, heart failure

Classic EKG finding is a "sawtooth" pattern:



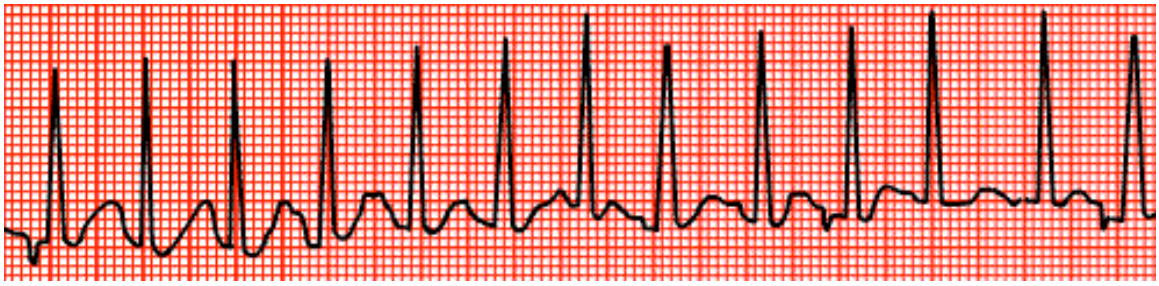
Treatment:

- If patient is stable, slow the ventricular rate with CCB's or β-blockers (avoid procainamide because it can result in increased ventricular rate as the atrial rate slows down)
- If cardioversion is going to take place be sure to anticoagulate for 3 weeks
- If patient is unstable must cardiovert → start at only 50J because is easier to convert to normal sinus rhythm than atrial fibrillation

Multifocal Atrial Tachycardia (MFAT)

- An irregularly irregular rhythm where there are multiple concurrent pacemakers in the atria.
- Commonly found in pts with COPD

EKG shows tachycardia with ≥ 3 distinct P waves



Treatment:

- Verapamil
- Treat any underlying condition

Supraventricular Tachycardia

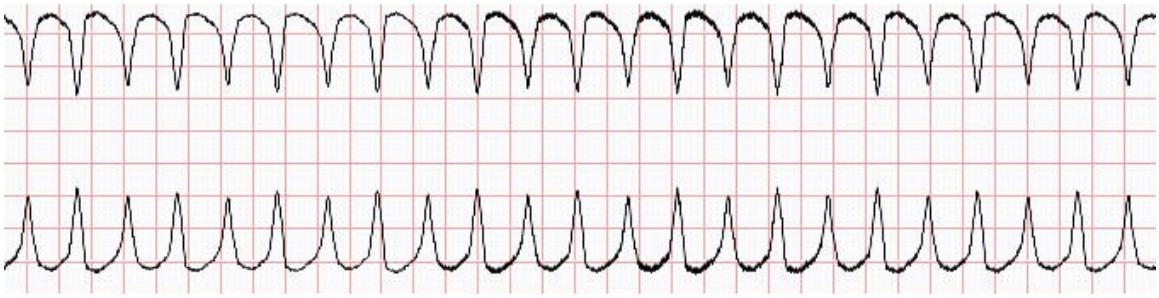
- Many tachyarrhythmias originating above the ventricle
- Pacemaker may be in atrium or AV junction, having multiple pacemakers active at any one time
- Differentiating from ventricular arrhythmia may be difficult if there is also the presence of a bundle branch block

Treatment:

- Very dependent on etiology
- May need to correct electrolyte imbalance
- May need to correct ventricular rate [digoxin, CCB, β-blockers, adenosine (breaks 90% of SVT)]
- If unstable requires cardioversion
- Carotid massage if patient has paroxysmal SVT

Ventricular Tachycardia

- VTach is defined as ≥ 3 consecutive premature ventricular contractions
- If sustained, the tachycardic periods last a minimum of 30s.
- Sustained tachycardia requires immediate cardioversion due to risk of going into ventricular fibrillation

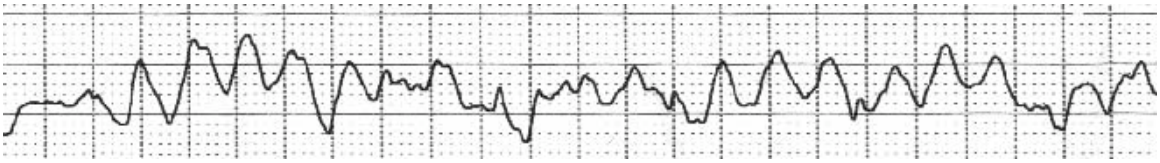


Treatment:

- If hypotensive or no pulse existent do emergency defibrillation (200, then 300, then 360J)
- If patient is asymptomatic and not hypotensive, the first line treatment is amiodarone or lidocaine because it can convert rhythm back to normal

Ventricular Fibrillation

- Erratic ventricular rhythm is a fatal condition.
- Has no rhyme or rhythm



Signs and Symptoms:

- Syncope
- Severe hypotension
- Sudden death

Treatment:

- 1st line - Emergent cardioversion is the primary therapy (200-300-360J), which converts to normal rhythm almost 95% of the time
- Chest compressions rarely work
- 2nd line – Amiodarone or lidocaine
- If treatment isn't given in a timely matter, patient experiences failure of cardiac output and this progresses to death.

Congestive Heart Failure

Definition:

- CHF occurs when the cardiac output is insufficient to met systemic demands
- May be right-sided, left-sided, or both

Causes:

- Valvular diseases
- MI
- Hypertension
- PE
- Anemia
- Cardiomyopathy
- Edocarditis
- Thyrotoxicosis

Signs and Symptoms:

Left-sided: signs and symptoms are due to ⇓ CO and ⇑ cardiac pressures

- Paroxysmal nocturnal dyspnea
- Exertional dyspnea
- Orthopnea
- Cardiomegaly
- S3 gallop
- Renal hypoperfusion (leads to sodium retention and worsened CHF)

Right-sided: signs and symptoms are due to pooling upstream of the right heart

- ⇑ JVP
- Edema
- Hepatic congestion
- Atrial fibrillation (increases risk of embolization)
- Fatigue
- Cyanosis
- Weight loss

Diagnose with echocardiogram

Treatment:

1st line regimen → ACEI, β-blockers, furosemide and spironolactone, and digoxin
If patient cannot tolerate ACEI, try hydralazine + isosorbide dinitrate

ACEIs have been proven to decrease mortality in CHF

β-blockers have been to decrease mortality

- Don't start β-blockers during active failure because they can exacerbate the condition
- Start β-blockers once patient is fully diuresed and is on stable doses of other medications

Spironolactone proven to decrease mortality in class IV CHF

Loops almost always used to maintain dry weight in CHF patients

Digoxin improves symptoms but DOESN'T decrease mortality

Be wary of giving Loop diuretic without spironolactone because this can cause an unsafe hypokalemia that potentiates the effect of digoxin (1st sign of digoxin toxicity is a SVT with AV block and blurry yellow vision)

Cardiomyopathies

	Dilated	**Hypertrophic**	**Restrictive**
Cause	Ischmia, infections, metabolic conditions, drugs	Genetic myosin disorder	Amyloidosis, scleroderma, hemochromotosis, glycogen storage disease, sarcoidosis
Signs & Symptoms	Right and left sided heart failure, S3 gallop, ***systolic dysfunction***	Exertional syncope, angina, LVH, ***diastolic dysfunction***	Pulmonary HTN, S4 gallop, ⇓ QRS dysfunction
Prognosis	30% 5yr survival rate	5% annual mortality rate	30% 5-yr survival
Treatment	Stop offending agents, tx is similar to CHF treatment	β-blockers and diuretics	Diuretics and correction of underlying cause

Diagnosis for each is echocardiography

Valvular Diseases

Presentation:

- Valvular heart diseases all present with shortness of breath as the chief complaint
- Often worsens with exertion/exercise

Clue to Diagnosis	Diagnosis
Young female and/or general population	**Miltral Valve Prolapse (MVP)**
Healthy young athlete	**Idiopathic Hypertrophic Subaortic Stenosis (now called: Hypertrophic obstructive cardiomyopathy HOMC)**
Immigrant, pregnant	**Mitral Stenosis**
Turner's syndrome	**Bicuspid aortic valve**
Palpitations, atypical chest pain not associated with exertion	**Mitral Valve Prolapse**

Physical Findings:

- Murmur and rales (seen in all cases)
- Peripheral edema, gallops, carotid pulse findings (possibly seen)

Murmurs:

Systolic:

- Most commonly seen in aortic stenosis, mitral regurgitation, MVP, and HOCM

Diastolic:

- Most commonly seen with aortic regurgitation and mitral stenosis.

All right-sided murmurs **INCREASE** in intensity with inhalation

All left-sided murmurs **DECREASE** in intensity with exhalation

Location and Radiation of murmurs:

Valvular Lesion	Best heard at
Aortic Stenosis	2nd right intercostal space and radiates to the carotids
Pulmonic valve	2nd left intercostal space
Aortic regurgitation/tricuspid/VSD	Left lower sternal border
Mitral regurgitation	Apex (left 5th intercostal space)

Murmur intensity:

I/VI → only heard with special maneuvers (valsalva)
II/VI and III/VI → majority of murmurs
IV/VI → thrill present
V/VI → can be heard with stethoscope partially off of the chest
VI/VI → can be heard without a stethoscope

Diagnosis:

- Best initial diagnostic test for valvular lesions is echocardiogram
- The most accurate test is left heart catheterization

Treatment:

Regurgitant lesions → best treated with vasodilator therapy (ACEI, ARB)

Stenotic lesions → best treated with anatomic repair (mitral stenosis requires balloon valvuloplasty, severe aortic stenosis requires surgical replacement)

A trick to know what type of therapy you should use:

If the valsalva maneuver improves the murmur, use diuretics.

If amyl nitrate improves the murmur, ACEI is indicated.

Aortic Stenosis

- Most commonly presents with chest pain
- Syncope and CHF are less commonly present with aortic stenosis
- Patient is often older and has a history of hypertension

Prognosis:

- If coronary disease is present then 3-5yr is avg survival
- If syncope is present then 2-3 yr avg survival
- If CHF present then 1.5-2yr avg survival

Diagnosis:

- TTE is the best initial diagnosis
- TEE is more accurate
- Left heart catheterization is the most accurate
- EKG and CXR will show LVH

Treatment:

- Diuretics are the best initial therapy but do not alter the long-term prognosis, and special attention must be paid since over-diuresis is a possibility
- Treatment of choice is valve replacement

Aortic Regurgitation

- HTN
- Rheumatic heart disease
- Endocarditis

Signs and Symptoms:

- Diastolic decrescendo murmur heart best at the left sternal border

Diagnosis:

- TTE is best initial diagnostic test
- TEE is more accurate
- Left heart catheterization is most accurate

Treatment:

- ACEI's
- ARB's
- Nifedipine

If ejection fraction drops below 55% or the LV end-diastolic diameter goes above 55mm, surgery should be done even if the patient is asymptomatic.

Mitral Stenosis:

- MCC of mitral stenosis is rheumatic fever
- Seen in immigrants and pregnant patients (increased plasma vol in pregnancy)

Signs and Symptoms:

- Dysphagia (large left atrium compresses esophagus)
- Hoarseness (pressure on recurrent laryngeal nerve)
- Atrial fibrillation

Physical Exam:

- Diastolic rumble after an opening snap

Diagnosis:

- TTE is best initial diagnostic test
- TEE is more accurate
- Left heart cath is most accurate
- EKG and/or CXR showing left atrial hypertrophy

Treatment:

- Best initial therapy is diuretics, however they do not alter progression of the disease
- Balloon valvuloplasty is the most effective therapy (all pregnant women must have this procedure done)

Mitral Regurgitation

- Caused by HTN, ischemic heart disease, and any condition that may lead to dilation of the heart
- The most common complain is dyspnea on exertion

Physical exam findings:

- Holosystolic murmur that obscures both S1 and S2
- Best heard at the apex, radiates to the axilla

Diagnosis:

- TTE is best initial test
- TEE is more accurate

Treatment:

- ACEI
- ARB's
- Nifedipine
- If LV ejection fraction drops below 60% or LV end systolic diameter is above 45mm, then surgery should be done

Pericardial Disease

Pericarditis

- Pleuritic chest pain
- Relieved by leaning forward
- Pain often described as sharp and brief

Signs and Symptoms:

- Friction rub is commonly found
- No other pertinent physical findings

Diagnosis:

- Best initial test is the EKG
- Diffuse ST-segment elevation
- PR-segment depression is pathognomonic but is not always present

Treatment:

- Best initial therapy is NSAID's
- Patient should return in 1-2 days, if the pain is gone they are cured
- If pain persists after 2 days of NSAID treatment, prednisone orally is treatment

Pericardial Tamponade

- Presents with SOB, hypotension, JVD + clear lungs
- Pulsus paradoxus is present (decreased BP >10mmHg on inspiration)
- Electrical alternans is present (alteration of QRS complex on EKG

Diagnosis:

- Echo is the most accurate diagnostic test
- Earliest finding is usually collapse of the right atrium and ventricle
- EKG shows low voltage and electrical alternans
- Right heart catheterization will show equalization of all pressures in the heart during diastole

Treatment:

- Best initial therapy is a pericardiocentesis
- Most effective long-term therapy is pericardial window placement

Constrictive Pericarditis

- Presents with SOB
- Edema
- JVD
- Ascites
- Hepatosplenomegaly

Unique features of constrictive pericarditis:

- A pericardial knock, which is an extra diastolic sound from the heart hitting the calcified pericardium

Diagnosis:

- CXR showing calcification
- Low voltage EKG
- Thickened pericardium on CT

Treatment:

- Diuretics are the best initial therapy
- Pericardial stripping is the most effective therapy

Chapter 8

Endocrine

Pituitary Disorders

Prolactinoma

- Prolactin-secreting tumor
- Always think of this when there is visual disturbances

Signs and Symptoms:

Men:

- Impotence
- Decreased libido
- Gynecomastia
- Most often men also present with headache and visual disturbacnes

Women:

- Amenorrhea
- Galactorrhea
- Both in the absence of pregnancy

Diagnosis:

- Rule out pregnancy
- Rule out drugs such as: Metoclopromide, Phenothiazines, and/or TCA
- MRI to confirm presence of tumor

Treatment:

- 1st line treatment is a dopamine agonist such as bromocriptine (most prolactinomas respond to DA agonists)
- If medical therapy doesn't work, surgical removal is done

Acromegaly

- Excess production of GH due to a GH-producing adenoma in the pituitary

Signs and Symptoms:

- Enlargement of the head, hands, feet, nose, and jaw
- May be enlargement of the sweat that can cause intense sweating
- Joint abnormalities (excess growth of articular cartilage)
- Amenorrhea
- Cardiomegaly and hypertension
- Colonic polyps
- Diabetes also common because insulin is antagonized by GH

Diagnosis:

- Best initial test is IGF (confirms diagnosis of acromegaly)
- Most accurate test is administration of glucose (normally should suppress GH, if it suppresses GH then this excludes acromegaly)
- MRI done after there is a reason for doing so

Treatment:

- Transphenoidal removal
- DA agonist to inhibit GH release
- Octreotide has some merit in preventing GH release
- Pegvisomant → a GH receptor antagonist

Diabetes

Type 1 DM

- Autoimmune destruction of pancreatic β-cells, leads to insulin deficiency

Signs and Symptoms:

- Polyuria, polyphagia, polydipsia
- Weight loss
- DKA → emergency

Diagnosis:

- Random plasma glucose >200 with symptoms OR
- Two measurement of fasting glucose >125
- 2hr oral glucose tolerant test >200 with or without symptoms

Treatment:

- Insulin replacement

Complications:

- DKA

Signs and Symptoms of DKA:

- Hyperglycemia >250
- Hyperkalemia (due to transcellular shift out of the cell in exchange for H+)
- Low pH
- Elevated levels of acetone, acetoacetate, and β-hydroxybutyric acid
- Increased anion gap

DKA treatment:

1st → IV fluids

2nd → potassium replacement(hyper becomes hypo as DKA is treated), insulin replacement

3rd → addition of glucose to insulin drip when pt becomes normoglycemic (keep giving insulin until ketones are gone)

** insulin is given originally to shut down ketogenesis, not decrease glucose, thus keep giving insulin until ketones are gone despite normal glucose.

Type 2 DM

- A peripheral insulin resistance
- Usually adult onset (changing with the obesity epidemic)
- Family history often plays a strong role
- Ketosis is NOT associated with DM2

Signs and Symptoms:

- Acute → 3P's (polydypsia, polyphagia, polyuria), fatigue, weight loss
- Subactue → infections (yeast infections, Mucor, S. Aureus)

Chronic signs and symptoms:

- Macrovascular → stroke, CAD
- Microvascular → retinitis, nephritis
- Neuropathy → parasthesia, stocking and glove burning sensation, autonomic insufficiency, ⇓ sensation

Diagnosis: same as type 1

Treatment:

- FIRST treatment is always diet and lifestyle modifications
- Oral hypoglycemics for mild/moderate disease
- 1st line → metformin (biguanide), its MOA is blocking gluconeogenesis
- 2nd line → sulfonylurea (glyburide), MOA is ⇑ β-cell insulin secretion
- 3rd line → Thiazolidinediones (pioglitazone), MOA is increasing tissue sensitivity to insulin
- If oral drugs don't work, patient may require insulin
- Lifelong cases most usually will require insulin treatment
- ACEI's important because they slow down the progression of diabetic nephropathy

Monitoring DM with HbA1c:

- HbA1c allows us to get a measure of the average glucose level over the past 3 months
- Tight glucose control is directly responsible for decreasing complications and mortality in both types of insulin
- An HbA1c <7 or 8 is recommended (this # is always decreasing)

Complications of DM2:

Hyperosmolar Hyperglyicemic Nonketotic Coma(HHNK):

- Often precipitated by stress, secondary to hypovolemia
- Glucose can become >1000mg/dL
- There is no acidosis (as in type 1 DM)

Treating HHNK:

- IV fluids are most important, rehydration is often all that is needed.
- May require upwards of 10L of fluids
- Without treatment, mortality rate climbs over 50%

Complications of Diabetes

Hypertension → Control is essential in DM patients because it causes long-term complications of the heart, eye, kidney, and brain. Goal is to keep it <130/90

Lipid Management → Goals are: LDL <100, if patient has CAD + DM, the goal is <70.

Retinopathy → Diabetics require a yearly eye exam to detect proliferative retinopathies. If present, laser coagulation should be performed.

Nephropathy → If any form of protein is present in the urine give the DM pt ACEI's. These prevent nephropathies and ACEI's are 1st line drugs in DM with HTN

Neuropathy → Yearly foot exams are important. If neuropathy is present there is no need to delay treatment with gabapentin or pregabalin.

Erectile Dysfunction → Ask patient about this, sildenafil or tadalafil work well but do not give if they are also on nitrates

Gastroparesis → More common in long-term diabetics, there is impaired stretch-receptors and thus impaired motility. Patient will have bloating, constipation, fullness, and diarrhea. Give metoclopramide or erythromycin (increase gastric motility)

Diabetes	Type 1	Type 2
Onset	Juvenile/childhood	Adult (increasingly common in youth today)

Body Type	Thin	Obese
DKA?	Frequent	Rare
Treatment	Insulin	1st – lifestyle 2nd – oral hypoglyclemic agents

Adrenal Disorders

Cushing's Syndrome

There are 3 sources of Cushing's disease, they are listed in this table along with pertinent information

	Pituitary Tumor	**Ectopic ACTH Production**	**Adrenal Adenoma**
ACTH	High	High	Low
High-dose dexamethasone	Suppression	No suppression	No suppression
Specific test	MRI, petrosal vein sampling	Scan the chest and abdomen	Scan the adrenals
Treatment	Removal	Removal	Removal

There is a common presentation of all patients with hypercortisolism:

- ***Fat redistribution***: Truncal obesity, buffalo hump, thin arms/legs, "moon facies"
- ***Striae and easy bruising:*** Due to a loss of collagen(cortisol thins the skin)
- ***HTN***: Due to fluid and sodium retention
- ***Hirsuitism***: from increased adrenal androgen levels
- ***Muscle wasting***

Diagnosis:

1. 1mg overnight dexamethasone suppression test: normally a person will suppress the 8am level of cortisol if given dexa at 11pm the previous night. A normal test (suppression) will rule out hypercortisolism of all kinds. ** a test may be elevated due to other factors such as depression, alcoholism, or excessive stress

2. 24-hr urine cortisol: this test adds specificity to the overnight dexamethasone test, if the overnight test was abnormal (failing to suppress ACTH), then this test confirms hypercortisolism.

** these tests are to diagnose the presence of Cushing's syndrome, the location is still unknown at this point.

Diagnosing the location:

- Looking at the ACTH can help identify the location
- If ACTH is high → source of problem is the pituitary or ectopic ACTH production
- If ACTH is low → source is the adrenal

Treatment:

- Removal of whatever is causing the problem, identified by MRI or abdominal scan (depending on the location of the problem)

Addison's Disease (adrenal insufficiency)

Can be primary (Addison's) or secondary (⇓ ACTH production from pituitary)

Addison's disease:

- MC is autoimmune disorder
- Waterhouse-Friderichsen may be cause, which is hemorrhagic necrosis of the adrenal medulla during the course of meningococcemia

Signs and Symptoms:

- Fatigue
- Anorexia
- Hyponatremia + hyperkalemia
- Hypotension
- Nausea/vomiting
- Constipation
- Hyperpigmentation (only in primary case)

Diagnosis:

- ⇑ ACTH and ⇓ cortisol (in response to ACTH)
- Hyperpigmentation
- If cause is secondary, then cortisol will ⇑ in response to ACTH

Treatment:

- For acute Addison's give fluids + hydrocortisone
- For stable patient give prednisone
- If patients do not respond to above treatments, give fludrocortisone (highest # of mineralocorticoids)

Adrenal Cortical Hyperfunction

1° Hyperaldosteronism (Conn's Syndrome):

- Most commonly due to an adenoma or hyperplasia of the zona glomerulosa of the adrenal gland

Signs and Symptoms:

- Hypertension
- ⇑Na+
- ⇑Cl-
- ⇓K+
- ⇓ renin

Diagnosis:

- ⇑ aldosterone
- ⇓ renin
- CT showing adrenal lesion

Treatment:

- If adenoma → surgical resection
- If hyperplasia → spironolactone

2° Hyperaldosteronism:

- Increased renin production 2° to decreased renal bloodflow(CHF, shock, renal artery stenosis)

Diagnosis:

- ⇑ renin (this is used to differentiate between 1° and 2° causes

Treatment:

- Treat underlying cause
- Treat HTN

Pheocromocytoma

Patient presents with:

- Episodic HTN
- Headache
- Palpitations
- Tachycardia
- Diaphoresis

Diagnosis:

- Best initial tests → high plasma and urinary catecholamine/plasma-free metanephrine and VMA levels
- Most accurate tests → CT or MRI of adrenal glands

Treatment:

- 1st – phenoxybenzamine to control BP
- 2nd – propranolol (only after α-blockade with phenoxybenzamine)
- 3rd – surgical resection

Male Gonadal Disorders

Disease	Characteristics	Treatment
Klinfelter's Syndrome	• XXY inheritance with variable expressivity • Diagnosis usually at puberty when no virilization • Tall with small testes and gynocomastia • Decreased testosterone • ⇑ LH/FSH (no feedback) • Dx with buccal smear showing barr body	Testosterone Supplements
XXY syndrome	• Mild mental retardation, acne, violent, antisocial behavior • Diagnose with karyotype analysis	None
Testicular feminization syndrome	• Defect in DHT receptor • Female external genitalia with sterile, undescended testes • Patient appears female but is sterile with blind vagina • Testosterone/estrogen/LH are all elevated	No tx Remove testes
5-α-reductase deficiency	• Ambiguous genitalia until puberty • At puberty a burst of testosterone overcomes the lack of DHT (masculinizing external genitalia) • Testosterone and estrogen are normal • Diagnosis is by genetic testing	Testosterone
Congenital adrenal hyperplasia	• A defect in the steroid synthesis pathway causes virilization of females or failure to virilize in males • 21-α-hydroxylase deficiency causes 95% of all CAH cases • severe disease presents in infancy with ambiguous genitalia and excess salt loss • less severe → minimal virilization and salt loss	Hormone replacement
Prader-Willi syndrome	• paternal imprinting • short limbs, floppy baby • hyperphagia (obesity → increases early death likelihood) • mental retardation • classically have almond-shaped eyes with strabismus • diagnosis is genetic analysis	None
Kallmann's syndrome	• AD hypogonadism with anosmia • Decreased production and secretion of GnRH by hypothalamus • Diagnosis made by finding decreased circulating LH and FSH	Pulsatile GnRH

Thyroid Disorders

The clinical differences between hyperthyroidism and hypothyroidism

	HYPOTHYROIDISM	HYPERTHYROIDISM
Weight	Gain	Loss
Intolerance	Cold	Heat
Hair	Course	Fine
Skin	Dry	Moist
Mental	Depressed	Anxious
Heart	Bradycardia	Tachycardia
Muscle	Weak	Weak
Reflexes	Diminished	
Fatigue	Yes	Yes
Menstrual Changes	Yes	Yes

Hypothyroidism

- Most commonly from 'burnout' Hashimoto's thryoiditis.
- Patient is fatigued
- Poverty of movement
- Gaining weight

Diagnosis:

- ⇑ TSH
- ⇓ T4

Treatment:

- Thyroxine
- T4(converted in the tissue to T3 as needed)

Hyperthyroidism

- ⇑ T4 levels
- ⇓ TSH
- 4 forms of hyperthyroidism: Graves(MC), Silent, Subacute, Pituitary adenoma

Grave's Disease:

Has many findings that are unique to this type of hyperthyroidism:

- Exophthalmos and proptosis
- Dermopathy (redness and thickened skin below the knee)
- Onycolysis (seperation of the nail from the nailbed)
- RAIU is elevated

Treatment:

- PTU or methimazole is given acutely to bring the gland under control
- After gland is controlled, use radioactive iodine ablation
- Propranolol used to treat sympathetic symptoms

Silent Thyroiditis:

- An autoimmune process with a non-tender gland and hyperthyroidism
- No eye, nail, or skin finding
- RAIU test is normal
- Gland is not in a state of hyperfunctioning, rather it is "leaking"
- Antibodies to thyroid peroxidase and antithyroglobulin antibodies may be present

Treatment: There is no treatment

***Subacute Thyroiditis*:**

- A condition of viral etiology
- Gland is tender

Diagnosis:

- TSH low
- T4 increased
- RAIU low

Treatment:

- Aspirin to relieve the pain

Pituitary Adenoma:

- Rare condition
- Is the only hyperthyroid disorder with an elevated TSH

Diagnosis:

- MRI of brain

Treatment:

- Removal of adenoma

Exogenous Thyroid Hormones Abuse:

- Will be an elevation in T4 (due to taking thyroid hormone)
- The TSH will be suppressed due to negative feedback
- Thyroid gland will atrophy

Thyroid Storm:

- This is an emergency situation where there is severe release of thyroid hormones from the thyroid gland
- Causes symptoms of extreme sympathetic stimulation (tremor, tachycardia, diaphoresis, etc)

Treatment:

1st → give iodine to block the uptake of iodine into the gland

2nd → give PTU or methimazole to block thyroxine production

3rd → dexamethasone to block the peripheral converion of T4→ T3

4th → block sympathetic effects with propranolol

Myxedema Coma:

- An emergency hypothyroid condition
- May be spontaneous or ppte by cold conditions, infections, sedative drugs, respiratory failure

Signs and Symptoms:

- Hypoventilation
- Hypotension

- Stupor
- Coma
- Seizures

Treatment:

- Levothyroxine
- Cortisone
- Intubate

Thyroid Malignancies

- All solitary dominant nodules should be diagnosed by FNA
- Excision if malignancy is suspected
- Hot nodules (a nodule that takes up more radioactive iodine) are less likely to be malignant
- Cold nodules (nodules that take up less radioactive iodine) are more likely to be malignant

Papillary cancer:

- Most common type of thyroid
- Best prognosis (85% 5-yr survival rate)
- Psammoma bodies & orphan-annie bodies/ground-glass nuclei

Follicular cancer:

- Good prognosis
- Common metastasis to bone and lungs

Medullary cancer:

- Prognosis is intermediate
- Is a cancer of the parafollicular "C" cells that are derived from cells of the 5^{th} branchial pouch
- Secretes calcitonin

Anaplastic cancer:

- Terrible prognosis
- Has a 0% 5-yr survival rate

Multiple Endocrine Neoplasia:

Type 1 (Wermer's)	3 P's: Pituitary, Pancreas, Parathyroid
Type 2 (Sipple's)	Pheocromocytoma, Medullary Thyroid CA, Parathyroid
Type 2b (Type 3)	Pheocromocytoma, Medullary Thyroid CA, Mucocutaneous neuromas(esp in GI)

Chapter 9

Infectious Disease

Answering questions in the infectious disease section are usually pretty straight forward. Being able to correctly answer these questions comes down to understanding the following:

1. Common conditions and their associated organisms
2. Common conditions and the best antibiotics to use
3. Organisms and their gram stain characteristics
4. Common scenarios and their commonly associated organisms

For greatest chances of success with these questions, the following tables should be memorized.

Gram Stain Characteristics

ORGANISM CHARACTERISTICS	STAINING CHARACTERISTICS
Gram (+) organisms	Blue in color
Gram (-) organisms	Red in color
Gram (+) cocci (pairs)	S. Pneumonia
Gram (+) cocci (chains)	Streptococcus
Gram (+) cocci (clusters)	Staphylococcus
Gram (-) diplococci	Neisseria
Gram (-) rods	Hemophilus
Gram (-) rods with mucoid capsule	Klebsiella
Pseudohyphae on stain	Candida
Acid Fast Organisms	Mycobacterium, Nocardia
Silver Staining	Pneumocystis Carinii
Spirochete	Borrelia, Treponema/Leptospira(darkfield)

Common situations and their associated organisms

SITUATION	ASSOCIATED ORGANISMS
Cellulitis from a dog or cat bite	Pasteurella Multocida
Burn wound infection with a blue/green color	Pseudomonas
Baby Paralyzed after eating honey	Clostridium Botulinum
Diarrhea after taking antibiotics	Clostridium Difficile
Pricked by thorn while gardening	Sporothrix Schenckii
Gastroenteritis in young child	Rotavirus
Diarrhea after traveling to Mexico	E. Coli
Aplastic crisis in Sickle Cell patient	Parvovirus B19
Food poisoning after eating reheat rice	B. Cereus
Food poisoning after eating raw seafood	Vibrio Parahemolyticus
Pneumonia in Southwest USA	Coccidioides Immitis
Pneumonia after exploring caves	Histoplasma Capsulatum
Pneumonia after exposure to bird droppings in Ohio	Histoplasma Capsulatum
Pneumonia after exposure to exotic birds	Chlamydia Psittaci
Pneumonia in a patient with silicosis	Tuberculosis
Diarrhea after hiking or drinking from a stream	Giardia Lamblia
B12 deficiency	Diphyllobothrium Latum
Fever and muscle pains after eating raw meat	Trichinella Spiralis
Pneumonia after being near an air conditioner or water tower	Legionella Pneumophilia
Slaughterhouse worker with a fever	Brucellosis
Fungal ball/hemoptysis after TB or cavitary lung disease	Aspergillus

High-yield conditions/illnesses and their most likely organism and treatments

CONDITION/ILLNESS	ORGANISM	BEST TREATMENT
Cellulitis	Staph, Strep	Antistaphylococcus penicillin
UTI	E. Coli	TMP-SMX, Nitrofurantoin (in pregnancy)
Endocarditis	Staph, Strep	Antistaphylococcus, Aminoglycoside
Sepsis	Gram (-) organisms	3rd generation cephalosporin's
Septic Arthritis	Staph Aureus	Antistaphylococcus PCN, Vancomycin (severe)
Meningitis (neonatal)	Group B Strep, E. Coli, or Listeria	Ampicillin + Aminoglycoside
Meningitis (child – adult)	Neisseria Meningitidis	3rd generation cephalosporin
Osteomyelitis	Staph Aureus, Salmonella (Sickle cell patient)	Antistaphylococcus PCN, Vancomycin
Pneumonia	Strep Pneumonia, H. Influenza	3rd generation cephalosporin
Pneumonia (atypical)	Mycoplasma, Chlamydia	Doxycycline, Macrolide
Bronchitis	H. Influenza	Amoxicillin, Erythromycin

Important information regarding HIV

How often should the CD4 count be checked? Q 6 months

At what point is PCP a worry in HIV/Aids patient? When CD4 is <200

What is the most common opportunistic pneumonia in AIDS? PCP

How to prevent PCP infection? Prophylax with TMP-SMX when CD4 ≤ 200

At what CD4 level should prophylaxis against Mycobacterium Avium Complex be started? When CD4 is <50

Which type of cancer are HIV/Aids patients at increased risk for? Kaposi's sarcoma

Should live vaccines be given do these patients? NO

Which is the only live vaccine to be given? MMR

Which type of blood disease is increased in this patient population? NHL

What is the most likely cause of pneumonia in HIV patient? Strep Pneumo

What is the most likely cause of Opportunistic pneumonia in HIV? PCP (be able to recognize if the question is asking most common cause or most common opportunistic cause)

Which organism can cause chronic diarrhea in AIDS patients? Cryptosporidium

Chapter 10

Allergies

Hypersensitivity Reactions

Type 1 – Anaphylactic (Preformed IgE antibodies)

Type 2 – Cytotoxic (preformed IgG and IgM antibodies)

Type 3 – Immune complex-mediated (antigen-antibody complexes deposited in vessels and cause an inflammatory response)

Type 4 – Delayed/Cell-mediated (sensitized T lymphocytes release inflammatory mediators)

Anaphylaxis

- Is a typ1 hypersensitivity reaction
- Due to preformed IgE antibodies that cause the immediate release of vasoactive amines such as histamines and leukotrienes.
- Commonly seen after bee stings and ingestion of medications such as penicillin and sulfa drugs

Presentation:

Symptoms develop acutely and are often very dramatic

- Difficulty breathing
- Hypotension + tachycardia
- Urticaria
- Angioedema

Treatment/Management:

- Secure airway
- Give subcutaneous epinephrine
- If these aren't available give corticosteroids
- Give antihistamines for cutaneous reactions

Angioedema

- Is most commonly caused by a deficiency of C1 esterase inhibitor

Presentation:

- Diffuse swelling of the eyelids, lips, and airway
- Usually occurs after mild facial trauma or ingestion of certain medications (often ace inhibitors)
- There is usually a family history
- C4 levels are low

Treatment/Management:

- Secure airway
- Give subcutaneous epinephrine
- Manage exactly like anaphylaxis

Allergic Rhinitis

- An allergic reaction that is very common
- Recurring nasal stuffiness, itching, rhinorrhea, and sneezing

Treatment/Management:

- The main treatment should be avoidance of the allergen
- Keep air clean
- Close windows and keep air-conditioning running during summer months
- Non-sedating antihistamines such as loratadine are very effective and can be used continually
- Nasal saline sprays/netti pot are effective at washing out the nasal cavity

Primary Immunodeficiencies

IgA Deficiency

- Is the most common primary immunodeficiency, and it is often asymptomatic
- Causes recurrent respiratory and GI infections
- Someone receiving blood products may develop anaphylaxis, which should make you think of an IgA deficiency
- Never give these patients immunoglobulin's

Treatment:

- Manage and treat infections as they arise

Bruton's Agammaglobulinemia

- Is an x-linked disorder affecting males
- Patients present with infections starting around 6 months of age
- Recurrent sinopulmonary infections due to Strep or Hemophilus organisms are classic

Treatment/Management:

- Infusion of IV Ig's

Common Variable Immunodeficiency

- Is a condition that presents in both men and women
- Usually only presents when they are adults
- Patient presents with recurrent sinopulmonary infections
- May get sprue-like abdominal disorders (malabsorbtion, diarrhea, steatorrhea)

Diagnosis:

- IgG levels are low

Treatment:

- Infusions of IVIG is required since IgG levels are low

Severe Combined Immunodeficiency

- An AR or x-linked disorder
- Commonly caused by adenosine deaminase deficiency
- There is a B and T cell defect, thus patient has severe infections early in life
- These are the so called "bubble babies", and require isolation to prevent life-threatening infections

Wiskott-Aldrich Syndrome

- An x-linked recessive disorder affecting only males
- There is a classic triad of eczema, recurring infections, and thrombocytopenia

Chediak-Higashi Syndrome

- Due to a defect in microtubule polymerization
- Giant granules in neutrophils
- Oculocutaneous albinism
- Recurring infections

Chronic Granulomatous Disease

- Usually an x-linked recessive disorder affecting males
- There is a defect in NADPH oxidase, causing recurring infections due to catalase (+) organisms (Staph, Pseudomonas,etc)
- Diagnostic test involves nitroblue tetrazolium dye (normally gets reduced by granulocytes) – measures respiratory burst, which is lacking in these patients

Chapter 11

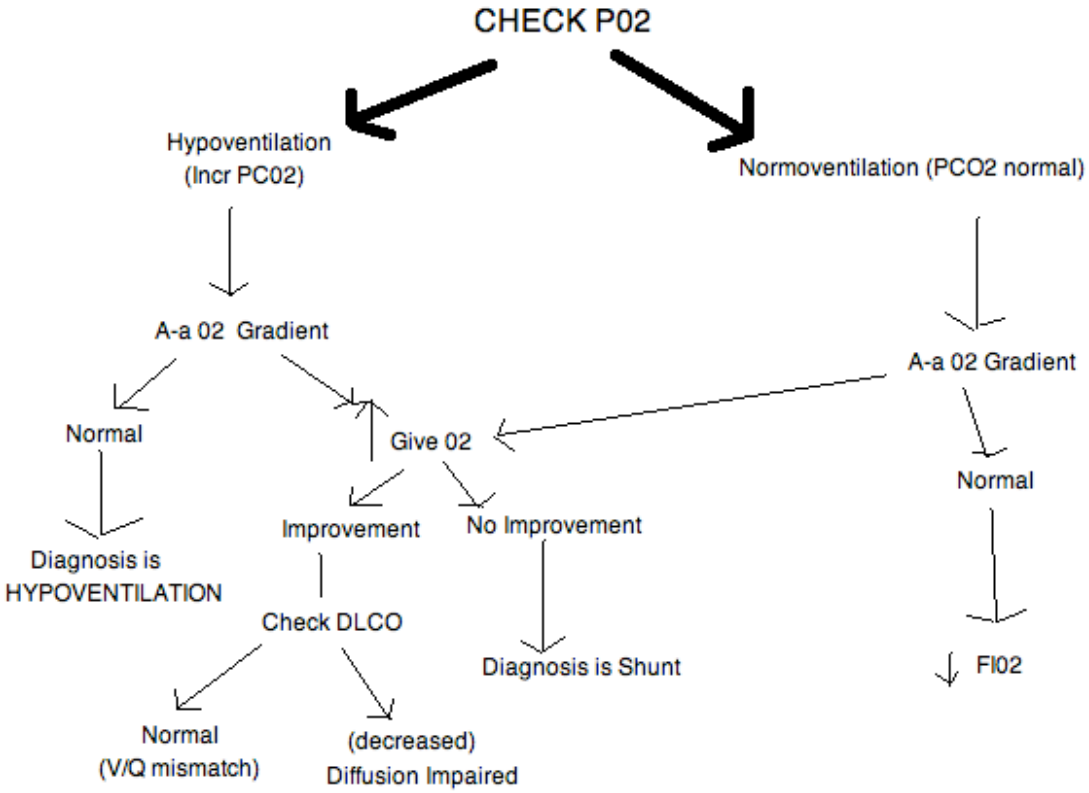
Pulmonary

Hypoxemia

There are 5 causes of hypoxemia:

1. Hypoventilation
2. Diffusion Impairment
3. V/Q mismatch
4. ⇓ Fi02
5. Shunt

Here is an algorithm figuring out the cause of hypoxemia



Signs and Symptoms:

- Tachycardia, dyspnea
- Clubbing and cyanosis
- Crackles and rales

Treatment:

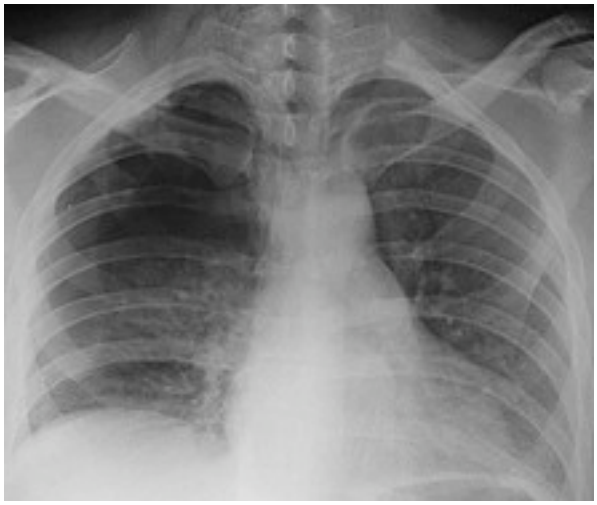
- Treatment requires treating the current hypoxemia and the treatment of underlying disorders
- ⇑ Pa02
- 02 by nasal cannula, or CPAP, or intubation if necessary
- If there is a shunt, the hypoxemia will not improve by increasing the FIO2

COPD's

1. Emphysema
2. Chronic Bronchitis
3. Asthma
4. Bronchiectasis

Emphysema

- Is air space dilation with alveolar wall destruction
- The most common cause of emphysema is smoking
- If a young patient gets this, consider an alpha-1-antitrypsin deficiency



Emphysema – Notice Lung Hyperinflation

Signs and Symptoms:

- Barrel chest
- Hyperventilation
- Pursed lip breathing
- Known as the "pink puffers"

Diagnosis:

- Clinical diagnosis + CXR showing hyperinflation of the lungs

Treatment:

Acute episodes require:

- 02 and an ABG
- CXR
- Albuterol (inhaled)
- Steroids for acute desaturations
- ADVISE PATIENT TO STOP SMOKING

Chronic management of COPD:

- Ipratropium inhaler
- Albuterol inhaler
- Yearly influenza vaccination
- Pneumococcal vaccine
- FUTHER SMOKING CESSATION ADVISING
- Long-term 02 therapy if P02 is <55% or the 02 saturation is <88%

Chronic Bronchitis

- Presents as a productive cough on most days for ≥ 3 months in a row for ≥ 2yrs
- Known as the "blue bloater"

Signs and Symptoms:

- Similar to emphysema however hypoxia is more severe
- RVH + pulmonary HTN
- Neck vein distention
- Hepatomegaly

Diagnosis:

- Diagnosis is largely clinical
- Can confirm with a lung biopsy that shows an increased Reid index, which is a glandular layer that is >50% of the total thickness of the bronchial wall.

Treatment:

- 02
- Bronchodilators

Asthma

- Presents with SOB and expiratory wheezing
- Severe cases may present with the use of accessory muscles
- Caused by bronchial hyperresponsiveness that is reversible

Signs and Symptoms:

- Expiratory wheezing and dyspnea
- Onset is often physical activity
- Condition is reversible with bronchodilators such as albuterol

Diagnosis:

- Highly clinical
- Check for a FEV increase of more than 10%
- Complication includes status asthmaticus, which is refractory attacks that last for days and are fatal

Treatment:

- Treatment with β2-agonists such as albuterol
- Long-term control involves addition of inhaled steroid (if patient isn't experiencing enough control with albuterol)
- If albuterol + steroids are not enough, addition of a long-acting β-agonist such as salmuterol may help
- Last resort in long-term management (refractory to these previous treatments) is oral steroids

Bronchiectasis

- Due to an anatomic defect that causes permanent dilation of the bronchioles
- Patient experiences recurring lung infections that produce massive amounts of sputum
- Patient often has digital clubbing as well

Diagnosis:

- The most accurate diagnostic test is the high-resolution CT scan, which will show thickened bronchial walls and dilated airways
- CXR will show the classic "tram-track markings"

Treatment:

- Must treat infections as they arise because there is no curative therapy
- Antibiotic therapy for recurring infections
- Chest physiotherapy can be helpful in releasing and eliminating sputum
- Long-term "cure" is a lung-transplant

Restrictive Lung Diseases

1. Interstitial Fibrosis
2. Parenchymal disease
3. Extrapulmonary disease
4. Pleural effusion

Interstitial Fibrosis

- Due to chronic insult to the lung tissue by things such as asbestos, chronic infections, organic dusts
- Diagnosis made by a CXR, which shows a "honeycomb" pattern of the lung

Treatment:

- 02, PEEP, steroids if there is collagen vascular disease

Parenchymal Disease

- Parenchymal diseases are caused by things such as infections (TB), inflammation (sarcoidosis), drugs, toxic/chronic inhalation of offending agents (asbestos), and it may be idiopathic
- Patient presents with a dry cough, SOB, and chronic hypoxia

Signs and Symptoms:

- "Velcro" rales
- Clubbing

Diagnosis:

- CXR or high-resolution CT
- Lung biopsy
- PFT (all measurements are decreased proportionately)

Treatment:

- If inflammatory, steroids can help
- There are no definitive cures for other forms of parenchymal disease

Extrapulmonary Disease

- Anything that affects the musculature responsible for aiding in breathing can cause problems
- Multiple sclerosis, ALS, Guillain-Barre, spinal cord trauma
- Anything that presses on the diaphragm can also cause trouble, such as pregnancy and obesity
- Management/treatment is supportive only

Pleural Effusion

- Fluid in the pleural space

Signs and Symptoms:

- Decreased breath sounds
- Dullness to percussion
- Decreased tactile fremitus

Diagnosis:

- The best initial diagnostic test is a CXR (lateral decubidus shows free flowing fluids)
- Most accurate test is thoracentesis (can show which type of fluid it is)

Treatment:

- Small effusions usually resorb spontaneously
- Diuretics can be used if causing respiratory problems
- If effusion is large, insert a chest tube for draining

Pulmonary Embolism

- Patient presents with a sudden onset of shortness of breath
- Lungs are clear

The risk factors for PE are usually telltale in the questions:

1. Immobility (Long airplane ride)
2. Trauma (Broken bone)
3. Surgery (Especially replacement of joints in the leg)
4. Malignancies
5. Thrombophilias

Diagnosis:

- A CXR should be done and is usually normal. May show a wedge-shaped infarct (large PE), atelectasis is a common finding
- EKG often shows non-specific ST-T wave elevations
- Best test to confirm diagnosis of PE is the spiral-CT and should be done if the xray is abnormal
- If the xray is normal but you are still suspicious, a V/Q scan should be performed (the less normal the xray the less accurate the V/Q scan will be)
- Doppler exam is only 70% sensitive, thus many PE's are missed with this test. The benefit of the Doppler is that if it DOES detect a PE, it is 100% accurate
- D-dimer is a highly sensitive test but it has low specificity. This is the best test to use if the patient has a low probability of having a PE and you want a single test to rule out a PE

Pulmonary Hypertension

Pulmonary hypertension is defined as hypertension that is ≥ ¼ that of the systemic pressure. Normally it should be approximately 1/8 that of the systemic pressure

- Active pulmonary hypertension means it is primarily a disease of the lung
- Passive pulmonary hypertension means it is secondary to a condition of the heart

Primary Disease:

- Idiopathic, which occurs commonly in young women
- Interstitial restrictive diseases
- Obstructive pulmonary diseases

Secondary Disease:

- Seen in heart disease
- Commonly seen in patients with HIV/AIDS

Signs and Symptoms:

- Tricuspid regurgitation
- Loud P2
- Right ventricular heave
- Raynaud's phenomenon

Diagnosis:

- Best initial test is the TTE, which will show RVH and an enlarged right atrium
- Most accurate test is right heart catheterization with increased pulmonary artery pressure
- EKG will likely show right-axis deviation

Treatment:

- 02
- Prostaglandins
- Endothelin inhibitors that prevent growth of the vasculature of the pulmonary system

Tuberculosis

- 1° TB affects the lower lobes and is usually asymptomatic
- It occurs in specific groups such as immigrants, HIV+ patients, homeless patients, and alcoholics.

Signs and Symptoms:

- Night sweats
- Fever
- Cough
- Sputum
- Weight loss

Diagnosis:

- CXR is the best initial diagnostic test
- Do an acid-fast stain of the sputum to confirm diagnosis

Treatment:

- Treatment with 4 anti-TB medications should be started with six months of therapy being the standard of care
- Isoniazid (6 months), Rifampin (6 months), Pyrizinamide (2 months), and Ethambutol (2 months)
- Do LFT's because these medications can cause liver toxicity (stop all medications if transaminase levels reach 5x the upper limit of normal)

Specific Toxicities caused by TB drugs:

Isoniazid → peripheral neuropathy, add B6

Rifampin → red/orange colored body secretions

Pyrazinamide → hyperuricemia

Ethambutol → optic neuritis

The PPD test

A screening test for those in risk groups.

Testing criteria is as follows:

- 5mm: close contacts, HIV+, steroid users
- 10mm: for those who are in the "high-risk" groups mentioned above
- 15mm: those with no increased risk

If PPD is positive, do the following:

1. CXR
2. If CXR is abnormal, do a sputum stain
3. If sputum stain is positive, start 4-drug therapy

Cancers of the Lung

- Lung cancers account for the most cancer deaths and are the 2nd most commonly diagnosed cancer
- XRAY is NOT a good screening tool because by the time they are seen metastasis has occurred
- Common signs and symptoms: Cough, hemoptysis, hoarseness, weight loss, fatigue, recurrent pneumonia

The following table demonstrates the common characteristics of different types of lung cancers

CANCER TYPE	CHARACTERISTICS
Adenocarcinoma	Is the most common lung cancer in non-smokers (***periphery and subpleura***) CEA (+), and is used to follow treatment
Bronchoalveolar carcinoma	A subtype of adenocarcinoma that is not related to smoking Presents in the ***periphery*** of the lung
Large Cell carcinoma	In ***periphery*** Is highly anaplastic and has a poor prognosis
Squamous cell carcinoma	Arises from ***bronchus and is a central*** hilar mass Strongly linked to smoking PTHrP release causes hypercalcemia
Small cell carcinoma	***Central hilar*** location Strong link to smoking Secretion of ADH and ACTH causes multiple endocrine problems Treat with radiation + chemotherapy May cause Lambert-Eaton syndrome
Bronchial carcinoid tumor	***Secretes serotonin*** Causes recurrent diarrhea, flushing of the skin, asthmatic wheezing Manage with a 5-HT antagonist
Lymphoangio-leiomyomatosis	Is a ***smooth-muscle*** neoplasm Most commonly seen in menstruating women Presents classically with pneumothorax Treat with either progesterone or a lung transplant

Treat small cell carcinoma with a combination of radiation and chemotherapy

Treat all other types with local resection + radiation (non-metastatic), and radiation + chemo if metastatic

Superior Sulcus Tumor

- Also known as "Pancoast tumor", which causes the following:

1. ***Horner's syndrome*** – Ptosis, Anhydrosis, Myosis because it damages the sympathetic cervical ganglion in the lower neck, AND
2. ***Superior Vena Cava Syndrome*** – obstruction of the SVC causes facial swelling, cyanosis, and dilation of veins of the head and neck

Chapter 12

GI

Esophageal Disorders

The only two esophageal disorders that require an endoscopy are **CANCER** and **Barrett's esophagus**, both which require a biopsy to know the diagnosis.

DYSPHAGIA

Achalasia

- Dysphagia to both solids and liquids in a young non-smoker.
- May be food regurgitation, and aspiration of previously eaten food.
- Involves a failure of the gastroesophageal sphincter to relax, no mucosal abnormalities

Diagnosis:

- Best initial test is the barium swallow
- Most accurate test is an esophageal manometry

Treatment:

- Best Initial treatment is pneumatic dilation, if repeatedly unsuccessful do surgery.
- If patient refuses surgery, we can give them an injection of botulinum toxin.

Esophageal Cancer

Presents w/ the following:

1. Dysphagia: first to solids then to liquids
2. May have heme (+) stool and/or anemia
3. Often pts are >50yr and are smokers/alcohol drinkers.

Diagnosis:

Best initial test is an endoscopy

If endoscopy is not an option, do a barium swallow

Treatment:

- Best initial therapy is a surgical resection (if no local or distant metastasis)
- Follow surgery w/ chemo-based 5FU

Rings and webs

- Also known as peptic strictures.
- Can be caused by repetitive exposure of the esophagus to acids, resulting in scarring and stricture formation.
- Previous use of sclerosing agents for variceal bleeding can also cause strictures(this is why banding is the superior procedure).

Diagnosis:

- Best initial diagnostic test is a barium study

Following are the diff kinds of strictures:

1. ***Plummer-Vinson syndrome***: is a proximal stricture found in association with iron deficiency anemia. Is more common in middle-aged women and is associated with squamous cell esophageal cancer

    - Best initial therapy is iron replacement

2. ***Schatzki's rings:*** is a distal ring of the esophagus that presents w/ intermittent symptoms of dysphagia

    - Best initial therapy is pneumatic dilation

3. ***Peptic stricture:*** results from acid reflux. Treat with pneumatic dilation

Zenker's Diverticulum

Look for pt w/ dysphagia w/ horribly bad breath. There is food rotting in the back of the esophagus from dilation of the posterior pharyngeal constrictor muscles.

Diagnosis:

- Best initial test is a barium study
- Best initial therapy is surgical resection

Spastic Disorders

- Diffuse esophageal spasm and "nutcracker esophagus" are essentially same disease.
- Look for case of severe chest pain, often w/o risk factors for Ischemic heart disease.
- May occur after drinking a cold beverage.
- Pain is always present, but dysphagia isn't always present.
- All cardiac tests are normal

Diagnosis:

- Most accurate diagnostic test is manometry
- Barium studies may show a corkscrew pattern, but only during an episode of spasm

Treatment:

- Calcium channel blockers and nitrates are the best treatment options

Esophagitis

Esophagitis presents with odynophagia as the food rubs against the esophagus.

Diagnosis:

- IF patient is HIV (-), do an endoscopy first
- IF patient is HIV (+), has a CD4 count <100 give fluconazole.. only do endoscopy if the patient doesn't response to fluconazle.

Candida Esophagitis

- Causes 90% of esophagitis in HIV (+) patients
- The other common cause is pill esophagitis, where certain pills can cause esophagitis in the patient.

Treatment:

- Have pt sit upright when taking the pills
- Have patient drink more water and remain upright for 30 minutes after swallowing.

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Mallory-Weiss tear

Is an upper GI bleed with violent retching and vomiting of any cause.
There may be hematemesis or black stool on exam or in the history

Treatment:

- Most cases resolve spontaneously, if bleeding persists, injection of epinephrine can be used to stop the bleeding.

GERD

Patient presents with a history of epigastric pain that is associated with substernal chest pain and possibly a metallic taste in the mouth.

Signs and Symptoms (on top of the classic presenting ones)

- Sore throat
- Metallic or bitter taste
- Hoarseness
- Chronic cough
- Wheezing

NOTE: As many as 20% of people who have a chronic cough are suffering as a result of GERD

Diagnosis:

- PPI admin is both diagnostic and therapeutic.
- Further testing such as 24hr pH monitoring should only be done if there is no response to PPIs and the diagnosis still is not clear

Treatment:Mild disease should be controlled w/ lifestyle modifications such as:

- Weight Loss
- Sleeping in an upright position, or at least somewhat angled in bed
- Smoking cessation
- Limiting alcohol, caffeine, chocolate, and peppermint ingestion
- Avoidance of food and drink within 2-3 hours of going to bed

If those don't work then PPIs are the next best therapy for GERD. They should control 90-95% of cases.

If there is no improvement then a trial of H2-blockers should be tried (many adverse effects with these)

Barrett Esophagus

Is a metaplasia from squamous to columnar cells

Diagnosis:

- Perform endoscopy when there is weight loss, anemia, and/or blood in the stool, and in anyone who has chronic symptoms of reflux disease for more than 5yrs.

FINDING ON ENDOSCOPY	MANAGEMENT
Barrett esophagus	PPI and repeat endoscopy every 2-3 years
Low-grade dysplasia	PPI and repeat endoscopy in 3-6 months
High-grade dysplasia	Distal esophagectomy

Epigastric Pain

Any pt >45yr w/ persistent epigastric pain and/or discomfort should receive an upper endoscopy. This is essential to exclude the possibility of gastric cancer

Non-Ulcer Dyspepsia:

- Is the MCC of epigastric discomfort
- Can only be concluded after endoscopy has excluded an ulcer disease, gastric cancer, and gastritis

Treatment:

- Consists of symptomatic therapy w/ H2 blockers, liquid antacids, or PPIs.

Peptic Ulcer Disease

- Due to hypersecretion of acid
- Can be either duodenal ulcer(DU) or gastric ulcer(GU) diseases
- H. Pylori is the MCC of ulcers, 2nd MCC is NSAIDs, head trauma, burns, intubation, Crohn's disease, and ZES.
- Usually, food improves the pain of a duodenal ulcer and makes the pain of a gastric ulcer worse
- If the pt is above 45 and has epigastric pain, you must scope to exclude gastric cancer.

Gastritis

- Not due to hypersecretion of acid, as in PUD.
- Can be associated with H.Pylori, if this is present treat w/ PPI and 2 antibiotics.
- Gastritis can also be "atrophic" from pernicious anemia and is often associated with a deficiency of vitamin B12

Testing for H. Pylori:

Most accurate test:

- Endoscopy with biopsy (if this is done no further testing is required)
- *Serology* is very sensitive but not specific, if the serology is negative, this excludes H. Pylori.
- A positive test can't distinguish between new and previous infection.

Breath testing and stool antigen testing:

- These are not standard or routinely used. They can however distinguish between new and old disease.

Treating H. Pylori:

- Treat this bacteria with PPI and clarithromycin + amoxicillin.
- ONLY treat if its associated with gastritis or ulcer disease.

There is no need for routine post-treatment testing of H Pylori. there is no benefit in treating H. Pylori that is associated with non-ulcer dyspepsia.

If the treatment of H.Pyolir doesn't succeed, try the following:

1. Repeat treatment with 2 new antibiotics and PPI : Use metronidazole + tetracycline instead
2. If repeat treatment fails, evaluate for ZES (Gastrinoma).

Stress Ulcer Prophylaxis:

Routine prophylactic use of a PPI or H2 blocker or sucralfate should only be used if one of the following is present:

1. Head trauma
2. Intubation and mechanical ventilation

3. Burns
4. Coagulopathy and steroid use in combo

**NSAID or steroid use alone is not an indication for routine stress ulcer prophylaxis

Zollinger-Ellison Syndrome(ZES) or Gastrinoma

ZES is diagnosed by finding an elevated gastrin level and an elevated gastric acid output**.**

****remember that everyone on a H2 blocker or PPI has an elevated gastrin level.**

When to test the gastrin and gastric acid output?

When any of the following are present:

1. large ulcer >1cm
2. multiple ulcers
3. distal location near the ligament of Treitz
4. recurrent or persistent despite H. Pylori treatment

*if the gastrin and acid output level are both elevated, then localization of the gastrinoma is next.

Diagnosis:

- Most accurate is an endoscopic ultrasound
- Nuclear somatostatin scan is also very sensitive because ZES patients have a high number of somatostatin receptors

Treatment:

- Local disease requires surgical resection
- Metastatic disease requires the patient be on lifelong PPIs

Inflammatory Bowel Disease(IBD)

- Both Crohn's and ulcerative colitis can present with fever, abdominal pain, diarrhea, blood in stool, and weight loss.

- UC pres most often with abdominal pain and bloody diarrhea

Extraintestinal manifestations of IBD are:

- Joint pain
- Eye findings
- Skin findings
- Sclerosing cholangitis

Features more common to Crohn's disease are:

- Masses
- Skip lesions
- Involvement of upper GI tract
- Perianal disease
- Transmural granulomas
- Fistulae
- Hypocalcemia from fat malabsorbtion
- Obstruction
- Calcium oxalate kidney stones
- Cholesterol gall stones
- Vitamin B12 malabsorbtion from terminal ileum involvement

Diagnosis:

- Endoscopy is best initial test
- Barium studies are also good diagnostic tests

Crohn's Markers:

- Antisaccharomyces cerevesiae(ASCA) : positive
- Antineutrophil cytoplasmic antibody(ANCA) : negative

UC Markers:

- ASCA: negative
- ANCA: positive

Treatment:

- Best initial treatment for both is mesalamine
- Steroids are useful in acute cases
- Surgery is required if there is no relief from these treatment modalities

DIARRHEA

Infectious Diarrhea

The presence of blood indicates a pathogenic invader, which may include any of the following:

Campylobacter - Is the MCC of food poisoning.

Salmonella - Transmitted by chicken and eggs

Vibrio Parahemolyticus – Associated with infected seafood

E. Coli – There are many different types, which include:

- **E. Coli 0157:H7** → MC associated with haemolytic uremic syndrome(via effects of verotoxin). Look for undercooked beef in the history.
- **Vibrio vulnificus**: Look for shellfish in a person w/ liver disease
- **Shigella**: Secretes Shiga toxin, which is also associated with reactive arthritis
- **Yersinia**: Rodents are natural reservoirs, transmission via veggies, milk-derived products, and meat
- **Amebic**: Perform three ova & stool parasite exams or serologic testing. Treat with metronidazole

Diagnosis:

Best initial test → fecal leukocytes

Most accurate test → stool culture

Treatment:

Mild disease→ Keep the patient hydrated, this usually resolves on its own

Severe disease → Fluoroquinolones are the treatment of choice. Severe disease is defined as presence of any of the following (Blood, fever, abdominal pain, hypotension and tachycardia)

Non-Bloody Diarrhea:

- Non-bloody diarrhea may still be d/t the above pathogens, which can all present with non-bloody diarrhea.
- NO BLOOD in diarrhea will exclude all of the following, which never have blood:

1. **Viruses** → Rotavirus, Norwalk virus (Norovirus)
2. **Giardia** → Camping/hiking. Look for bloating/flatus/steatorrhea, stool ELISA is diagnostic test of choice, treatment involves metronidazole.
3. **Staph Aureus** → Presents with vomiting in addition to diarrhea. This resolves spontaneously
4. **Bacillus Cereus** → Is associated with eating refried rice. This resolves spontaneously
5. **Cryptosporidiosis**→ HIV (+) patient with a CD4 count <100. Diagnosis with acid-fast stain. There is no proven treatment except to raise the CD4 count w/ antiretroviral therapy.
6. **Scombroid**→ Histamine fish poisoning, has fastest onset of poisoning, which is around 10 min after eating infected tuna, mackerel, or mahi-mahi. Patient has vomiting, diarrhea, wheezing, and flushing. Treatment involves giving the patients antihistamines such as diphenhydramine.

Antibiotics Associated Diarrhea (C. Difficile)

Develops several days to weeks after use of antibiotics such as Clindamycin, which is the most common cause. There can be both blood and fecal leukocytes with C.Difficile-related colitis.

Diagnosis:

- Best initial test is a stool toxin assay.

Treatment:

- The best initial therapy is metronidazole

Chronic Diarrhea

The most common cause of chronic diarrhea is lactose intolerance

Diagnosis:

- Removal of milk products will both allow for diagnosis and treatment

MALABSORBTION

- Diarrhea caused by malabsorption is always associated with weight loss
- Fat malabsorption is associated with steatorrhea, which leads to oily/greasy stools that float on the water in the toilet and are foul smelling

The common causes of fat malabsorption are as follows:

1. Celiac Disease
2. Tropical Sprue
3. Chronic Pancreatitis
4. Whipple's Disease

All forms of malabsorption are associated with:

- Hypocalcemia from vitamin D deficiency, which may lead to osteoporosis
- Oxalate overabsorption and oxalate kidney stones
- Easy bruising and elevated PT/INR due to vitamin K malabsorbtion
- Vitamin B12 malabsorption from either destruction of terminal ileum or loss of pancreatic enzymes that are necessary for B12 absorption

Diagnosis:

- The best initial test is the sudan stain
- The most sensitive is a 72-hr fecal fat test

Celiac Disease

Presents with iron malabsorption and microcytic anemia

Diagnosis:

- Best initial diagnostic test is checking for antigliadin, antiendomyseal, and antitissue transglutaminase antibodies
- The most accurate test is a small bowel biopsy

*bowel wall biopsy is always necessary even w/ antibody confirmation, in order to exclude bowel wall lymphomas.

Treatment:

- Elimination of oats, wheat, barley from the diet

Tropical Sprue

Patient will have a history of being in a tropical location, and presents the same way as celiac disease.

Diagnosis:

- Small bowel biopsy is the best test to perform

Treatment:

- Tetracycline or TMP-SMX for 3-6 months

Whipple's Disease

A GI infection presenting with arthralgias, rash, diarrhea, and anemia

Diagnosis:

- The most accurate test is a small bowel biopsy that shows PAS (+) organisms
- Can also do a PCR of the stool looking for T. Whippelii

Treatment:

- Penicillin, Tetracycline, or TMP-SMX for 12 months

Chronic Pancreatitis

- History of alcoholism is usually present
- Lipase and amylase levels are likely normal since these wont drop until the pancrease is calcified and fibrosed.
- Fat soluble vitamins are not absorbed

Diagnosis:

- The best initial test is an abdominal XRAY or a CT without contrast
- The most accurate testis secretin stimulation testing (normal person releases large amount of bicarbonate-rich pancreatic fluid).

Treatment:

- Involves the administration of pancreatic enzymes by mouth.

Irritable Bowel Syndrome

- Syndrome where the patient experiences an alteration in bowel habits (constipation alternating with diarrhea)
- Pain is usually relieved after a bowel movement

Diagnosis:

- Testing may include colonoscopy, xrays, blood tests, but all are negative

Treatment:

- The best initial treatment is fiber supplementation, which helps by bulking the stool and relieving pain
- If fiber doesn't work, can add antispasmotic agents to try and relax the bowel
- If these fail to work, TCA's can be tried

COLON CANCER

- Hamartomas and hyperplastic polyps are benign
- Dysplastic polyps are malignant

** The most important thing to know for colon cancer screening is when and what to do for the patient

***Diagnostic Testing*:**

General Population:

1. Begin screening at 50yr
2. Colonoscopy q10yrs
3. Sigmoidoscopy q 3-5yrs
4. Fecal occult blood testing yearly
5. Barium enema

The best method of screening for colon cancer is performing a colonoscopy every 10yrs

One family member with colon cancer requires colonoscopy starting at 40yr or 10yr before age of family member who had cancer.

Three family member, two generations, one premature(<50) require a colonoscopy every 1-2yr starting at 25yrs. This is a "lynch syndrome" or HNPCC

Familial Adenomatous Polyposis(FAP)

- Start screening sigmoidsocopies at age 12
- Perform colectomy if polyps are found

Gardner's syndrome

- This presents with benign bone tumors known as osteomas, as well as other soft tissue tumors. There is no additional screening indicated for Gardner's syndrome

Peutz-Jeghers Syndrome

- This presents with a patient who has melanotic spots on the
- There are hamartomatous polyps throughout the small bowel and colon
- There is a lifetime risk of colon cancer is 10%.
- No extra screening recommended

Juvenile Polyposis

- There are multiple extra hamartomas in the bowel.
- No increased risk of colon cancer from hamartomas
- No extra screening recommended
- If dysplastic polyps are found, perform repeat colonoscopy in 3-5 years

GENERAL POPULATION	SINGLE FAMILY MEMBER WITH COLON CANCER	THREE FAMILY MEMBERS WITH CANCER	FAP	GARDNERS, PEUTZ-JEGHERS, JP
Start at 50, then q10yr	Start at 40yr or 10yr earlier than when diagnosed	Colonoscopy q1-2yr at 25yr	Sigmoidoscopy q102 yr starting at 12yr	No xtra screening recommended

DIVERTICULAR DISEASE

Includes Diverticulosis and Diverticulitis

Diverticulosis

Incredibly common in older Americans and it most commonly caused by a low-fiber, high-fat diet

Signs and Symptoms:

- LLQ abdominal pain
- Lower GI bleed

Diagnosis:

- The most accurate test is a colonoscopy
- Best diagnostic test is an abdominal CT scan

Treatment:

- High-fiber diet low in saturated fats

Diverticulitis

Is a complication of diverticulosis and presents with:

- LLQ abdominal pain
- Tenderness
- Fever
- Elevated white cell count in blood

Treatment:

- Involves the use of antibiotics. Metronidazole and ciprofloxacin most commonly used.

Gastrointestinal Bleeding

Red blood → lower GI bleed, rarely a very acute upper GI bleed can be red blood

Black stool → upper GI bleed (Proximal to Ligament of Trietz). Black stool usually is 100ml or more blood.

Heme (+) brown stool → can occur from as little as 5-10ml of blood loss

Coffee ground emesis → needs very little gastric, esophageal, or duodenal blood loss, as little as 5-10ml.

**The most important thing to do in acute GI bleeding is to determine if there is hemodynamic instability

Treatment for GI bleeds:

- IV fluids if it is a large bleed
- Correction of anemia or other lab abnormalities

Variceal Bleeding

- Look for alcoholic with hematemesis and/or liver disease
- Other clues are the presentation of splenomegaly, low platelets, and spider angiomas or gynecomastia

Diagnosis:

- Endoscopy

Treatment:

- First thing to do is add octreotide which decreases portal hypertension
- 2nd thing to do is an upper GI to band the variceals
- If banding fails, a shunt between the portal vein and the hepatic vein should be done

Sources of Bleeding

Upper GI:

- Ulcer disease
- Esophagitis
- Gastritis
- Duodenitis
- Varices
- Cancer

Lower GI:

- Angiodysplasia
- Diverticular disease
- Polyps
- Ischemic colitis
- IBD
- Cancer

Diagnosing sources of bleeding:

- Technetium bleeding scan (tagged red cells) detects source of bleed
- Angiography is an excellent preoperative test because it localizes the site of resection
- Capsule endoscopy should be done when the other methods fail. This is a small camera that is swallowed and allows for visualization of the small intestine

Acute Mesenteric Ischemia

Presents with a sudden onset of severe abdominal pain with a normal appearing exam (ie. The pain is out of proportion to the findings on exam)

Diagnosis:

- The most accurate test is an angiography

Treatment:

- Surgical resection of ischemic bowel

Other GI Conditions

Constipation

Initial management of constipation is hydration and fiber supplements

There is usually no clear etiology, for clinical purpose must know possible causes and be able to treat underlying reason.

1. Dehydration: look for signs of dehydration, BUN:Cr of >20:1
2. CCB's
3. Narcotic medication use/sedatives
4. Hypothyroidism: TSH, T4
5. Diabetes
6. Ferrous sulphate iron replacement
7. Anticholinergic medications

Dumping syndrome

- This is a rare disorder related to prior gastric surgery (Often for ulcer disease)
- This is a disorder where stomach contents are quickly passed through to the intestine, and it then draws fluid into the GI, causing an initial ***HYPER***glycemia follow by a reactive ***HYPO***glycemia

Signs and Symptoms:

- Shaking chills
- Diaphoresis
- Weakness
- Hypotension
- Hyperglycemia

Treatment:

- Small and frequent meals

Diabetic Gastroparesis

- Longstanding DM impairs neural supply of bowel, there is impairment of normal motility.
- Patient will present with bloating and constipation as well as diarrhea

Diagnosis:

- Clinical + history of diabetes

Treatment:

- Erythromycin (increases motilin in the gut, thus increasing motility) and metoclopramide.

Acute Pancreatitis

Presents (classically) as severe midepigastric pain and tenderness that is associated with alcoholism and/or gallstones

Diagnosis:

- Lipase (more specific) and amylase

Treatment:

- Keep patient NPO
- Give IV fluids
- Give pain medications

ACUTE HEPATITIS:

All pts present in a very similar way:

1. Jaundice
2. Fatigue
3. Weight loss
4. Dark urine from bilirubin in the urine

*Hepatitis B and C are more likely to pres with serum-sickness phenomenom like joint pain, urticaria, and fever.

- No definitive treatment is available for any form of acute hepatitis.

Diagnosis:

- Conjugated bilirubin levels (will be elevated)
- Viral Hepatitis gives elevated ALT level
- Drug-induced hepatitis is associated with increased levels of AST
- In pregnancy, if a patient gets hepatitis E this can be fatal
- **Most accurate tests** for hep A, C, D, and E, the confirmatory test is serology
- IgM levels acutely rise, and IgG levels rise in the recovery phase.
- Surface antigens, core antibody, e-antigen, or surface antibodies are only associated w/ hepatitis B.

Hepatitis B testing:

- The first test to become abnormal in acute hep B infection is the *surface antigen.*
- Elevation in ALT, e-antigen, and symptoms all occur after the appearance of hepatitis B surface antigen.
- **Chronic hep B** gives the same serologic pattern as acute hep B, but it is based on persistence of the surface antigen beyond six months.

*Tests for active viral replication: hep B DNA polymerase = e-antigen = hep B PCR for DNA, all equal the same thing.

Hepatitis C testing:

- ***Best initial test*** is hep C antibody, this wont tell the level of activity of the virus
- ***Most accurate test*** Hep C PCR for RNA, also is the most accurate way of testing response to therapy
- Liver biopsy is the most accurate for finding out the severity of the disease

<u>Treatment of Chronic Hepatitis</u>

Chronic hep B: The pt w/ surface antigen, e-antigen, and DNA polymerase or PCR for DNA is the pt most likely to benefit from antiviral therapy. Look for >6mnth of positive serology

Treat chronic hep B w/ following single agents:

1. Lamivudine
2. Adefovir
3. Entecavir
4. Telbivudine
5. Interferon (has the most adverse effects)

Chronic hep C: Combine interferon with ribavirin (MC adverse effect is anemia)

Vaccination:

- Hep A and B vaccination is now universally done in children.

- For adults the strongest indication for both types are the following:

1. Chronic liver disease
2. Household contacts
3. MSM (men who have sex with men)
4. Blood product recipients on a chronic basis
5. Injection drug users

Specific indications:

HEP A → travelers
HEP B → health care workers and pts on dialysis.

*there is no vaccine or postexposure prophylaxis for hep C

CIRRHOSIS

No matter what the cause may be, it will have a number of features:

1. Edema due to low oncotic pressure (treat with spironolactone + diuretics)
2. Gynecomastia
3. Palmar erythema
4. Splenomegaly
5. Thrombocytopenia due to splenic sequestration
6. Encephalopathy, which should be treated with lactulose
7. Ascites - Treat with spironolactone
8. Esphageal varices - Propranolol will prevent bleeding, perform banding if bleed continues.

Ascites:

Perform paracentesis for all pts with ascites if a new ascites, pain, fever, or tenderness are present.

Diagnosis:

- Test the fluid albumin level
- SAAG > 1.1 is consistent with portal hypertension from cirrhosis

CHRONIC LIVER DISEASE (Cause of cirrhosis)

Alcoholic cirrhosis

Is a diagnosis of exlusion. Must exclude all other causes of cirrhosis and look for a history of longstanding alcohol abuse. Treat as described above for cirrhosis

Primary Biliary Cirrhosis

Presents most commonly in a middle-aged woman complaining of itching. Xanthalasmas may be found on exam. Also look for a history of other autoimmune disorders

Diagnosis:

- The best initial test is elevated alkaline phosphatase + normal bilirubin level
- The most accurate test is presence of antimitochondrial antibody

Treatment:

- Ursodeoxycholic acid

Primary Sclerosing Cholangitis:

- 80% of those with PSC also have IBD

Signs and Symptoms:

- Urticaria
- Elevated bilirubin levels
- Elevated alkaline phosphatase

Diagnosis:

- Most accurate test is ERCP

- Anti-smooth muscle antibody, and (+) ANCA

Treatment:

- Ursodeoxycholic acid

Wilson's Disease

Involves cirrhosis and liver disease in a person with a choreiform movement disorder and neuropsychiatric abnormalities.

Signs and Symptoms:

- Extrapyrimidal symptoms
- Mania/depression
- Kayser-Fleischer rings around the cornea is pathognomonic for Wilson's disease

Diagnosis:

- Decreased serum ceruloplasmin

Treatment:

- Penicillamine

Hemochromatosis

- Most often from a genetic disorder resulting in overabsorbtion of iron
- Iron deposits throughout the body, most commonly in the liver.

Signs and Symptoms:

- Darkening of the skin (Bronze diabetes)
- Arthralgia

- Cardiomyopathies (Restrictive)
- Infertility
- Hepatoma

Diagnosis:

- Best initial test is serum study showing elevated serum iron and ferritin with a low TIBC. Iron saturation will be grossly elevated
- The most accurate test is a MRI or liver biopsy

Treatment:

- Phlebotomy

Autoimmune Hepatitis

Most often presentation is a young woman who has another autoimmune disease

Diagnosis:

- Best initial test is ANA and anti-smooth muscle antibodies
- The most accurate test is a biopsy of the liver

Treatment:

- Prednisone

Nonalcoholic steatohepatitis (NASH)

Strong association with obesity, diabetes, and hyperlipidemia

Diagnosis:

- Best initial test is liver studies that show ALT>AST
- The most accurate test is a liver biopsy that shows fatty infiltration

Treatment:

- Management of the underlying condition

Chapter 13

Nephrology

Renal Tubular and Interstitial Disorders

Drug Induced Interstitial Nephritis

- Classic drugs causing interstitial nephritis include PCN, NSAIDs, Sulfonamides, and diuretics.

Signs and Symptoms:

- Rash
- Hematuria
- Oliguria
- Fever
- Eosinophilia
- Eosinophiliuria is rare but is pathognomonic for hypersensitivity Allergic Interstitial Nephritis

Diagnosis:

- Diagnosis is mainly clinical, removal of offending agent + improvement helps to confirm diagnosis

Treatment:

- Removal of offending agent
- Steroids can help

Acute Renal Failure

- Presents with ⇑ azotemia
- ⇑ BUN & Creatinine
- Caused by either prerenal, renal, or postrenal azotemia

Prerenal Failure:

- Hypoperfusion will lead to renal failure
- Volume depletion, sepsis, heatstroke, burns, hypotension

***Intrinsic Renal Failure*:**

- ATN is the most common cause
- Renal ischemia also a possibility

***Postrenal Failure*:**

- Is due to obstruction secondary to either BPH, renal calculi, and/or bladder/pelvic tumors

Test	Prerenal	Postrenal	Renal
Urine Osmolality	>500	<350	<350
Urine Sodium	<20	>40	>20
FENa	<1%	>4%	>2%
BUN/Creatinine	>20	>15	<15

Treatment:

- IV fluids to maintain urine output
- Diuretics to prevent fluid overload
- Close monitoring of electrolyte abnormalities
- Dialyze with severe electrolyte abnormalities, unresponsive metabolic acidosis, uremia, and toxic ingestion

<u>Acute Tubular Necrosis</u>

- From either hypoperfusion that leads to tissue death or from insult due to various toxic injuries
- Is the most common cause of acute renal failure

Treatment:

- Remove cause
- Give IV fluids to maintain urinary output
- Closely monitor electrolytes
- Give diuretics as needed to prevent fluid overload

Renal Tubule Functional Disorders

1. Renal Tubular Acidosis
2. Diabetes Insipidus
3. Syndrome of Inappropriate Antidiuretic Hormone

Renal Tubular Acidosis:

Type	Characteristic	Urinary pH
Type I	A defect of the distal tubule (H+ gradient)	>5.5
Type II	Proximal tubule fails to resorb HCO3	>5.5 early then <5.5 as the acidosis worsens
Type IV	⇓ Aldosterone (leading to hyper K+ and hyper Cl- From ⇓ secretion seen in DM, interstitial nephritis, ACEI's, heparin, and NSAID use. May also be due to aldosterone resistance from sickle cell or urinary obstruction	<5.5

Diabetes Insipidus:

There is central and nephrogenic types of DI, both:

- ⇓ secretion of ADH if it is central diabetes insipidus, and an ADH resistance if it is nephrogenic

Signs and Symptoms of both:

- Polyuria
- Polydypsia
- Nocturia
- Urine osmolality ≤ 200 and serum osmolality ≥ 300

Central DI:

- Is either idiopathic (Primary) or caused by insult to brain (Secondary)
- Treat this with DDAVP nasal spray

Nephrogenic DI:

- Is an x-linked disease and may be secondary to sickle cell, pyelonephritis, nephrosis, amyloidosis, multiple myeloma drugs
- Treat by increasing water intake and restricting sodium intake

Diagnosis:

- With DDAVP administration, central DI will have a fast decrease in urine output, while nephrogenic DI will have no change in urine volume
- With DDAVP administration, central DI shows an acute increase in urine osmolality, where nephrogenic DI shows no change in osmolality
- Treat central DI with DDAVP or vasopressin
- Treat nephrogenic DI by correcting the underlying cause (electrolyte imbalances).

Syndrome of Inappropriate Antidiuretic Hormone (SIADH):

There are many possible causes of SIADH:

CNS disease: trauma, tumors, hydrocephalus

Pulmonary diseases: pneumonia, Small cell carcinoma of lung, abscess, COPD

Endocrine disease: hypothyroidism, Conn's syndrome

Drugs: NSAIDs, chemotherapy, diuretics, phenothiazine, oral hypoglycemics

Diagnosis: presence of hyponatremia with a urine osmolality of >300mmol/kg

Treatment: this condition is usually self-limiting, resistant cases may require demeclocycline which induces nephrogenic DI

Chronic Renal Failure:

- Always associated with renal azotemia

Characterized by:

- Azotemia
- Acidosis
- Hyperkalemia
- Hypocalcemia due to lack of vitamin D production
- Anemia (lack of erythropoietin production)
- Hypertension due to RAAS pathway activation

Signs and Symptoms:

- Nausea and vomiting
- Anorexia
- Dementia
- Convulsions
- Coma
- PLT dysfunction (leads to bleeding)

Diagnosis:

- Renal ultrasound showing small kidneys if failure is chronic
- Presence of anemia due to lack of EPO production

Treatment:

- Restrict both water and salt
- Prevent fluid overload with diuretics
- If there are severe electrolyte disturbances or acid-base problems go into dialysis

Glomerular Diseases

Nephritic and Nephrotic Syndromes:

Nephrotic: *Defined by* → hyperproteinuria, hypoproteinemia, hyperlipidemia, edema

- Proteinuria >3.5g/day
- Patient has generalized edema
- Hypercoagulation
- ⇓ albumin
- Hyperlipidemia

Diagnosis:

- ***Best initial test*** is a urinalysis showing significantly increased levels of protein
- Next best step is a spot-urine test for a protein: creatinine >3.5:1
- 24-hr urine protein collection >3.5g
- ***Most accurate test*** → Renal biopsy

TYPE OF NEPHROTIC DISEASE	COMMON CHARACTERISTICS/TX
Minimal Change Disease	This is seen in young children. Treat with prednisone
Focal Segmental Glomerulosclerosis	Similar in presentation to MCD but occurs in adults. Most commonly idiopathic. Commonly presents in young hypertensive males. Treat with prednisone and cyclophosphamide
Membranous Glomerulonephritis	Is the most common 1° cause of nephritic syndrome in adults. A slowly progressive disorder. Many causes: HBV, HCV, syphilis, certain drugs, malignancies, SLE. Treat with prednisone + cyclophosphamide. 50% of cases progress to end-stage renal failure
Membranoproliferative Glomerulonephritis	Type 1 is slowly progressive while Type 2 is aggressive. Autoantibody against C3 convertase (⇓C3 levels) Treat with prednisone, plasmapharesis.

Treatment:

- Protein and salt restriction
- HMG-CoA reductase inhibitor for hyperlipidemia

Nephritic:

- This happens when there is diffuse glomerular inflammation

Signs and Symptoms:

- There is an acute-onset of hematuria
- Oliguria
- Hypertension
- Edema
- ⇓GFR
- ⇑ Bun:Cr

TYPE OF NEPHRITIC DISEASE	COMMON CHARACTERISTICS/TX
Post-streptococcal Glomerulonephritis	An acute condition. Classically occurs after Strep Pyogenes infection. Immunofluorescence shows coarse granular IgG or C3 deposits. Labs show increased red cells and casts, ⇓ serum C3, ⇑ ASO titer.
Rapidly Progressive Glomerulonephritis (Crescentic)	A nephritic condition that progresses rapidly to renal failure. Goodpasture's disease is in this category. Immunofluorescence shows smooth and linear IgG deposits. Treat with prednisone and plasmapharesis.
Berger's Disease (IgA nephropathy)	Most common type of nephropathy. IgA deposits in mesangium. Presents with recurrent hematuria + low-grade proteinuria. Usually harmless, however 1 in 4 may progress to renal failure. Treat with prednisone
Henoch-Schonlein Purpura	Always in children, is an IgA nephropathy. Presents with abdominal pain, GI bleed, vomiting, and hematuria. Classically find palpable purpura on

	buttocks and legs Is a self-limiting disease that requires no steroids.
Multiple Myeloma	There is an increased light-chain production. Find Bence-Jones protein in urine. Hypercalcemia seen. Patient becomes succeptible to encapsulated bacteria because there is a defect in normal antibody production. Treatment must be on the underlying myeloma.

Systemic Glomerulonephropathies:

DISEASE	CHARACTERISTIC NEPHROPATHY
Diabetes Mellitus	Is the MCC of ESRD. Early manifestation is microalbuminuria (give ACEI's, strict glycemic control).
HIV	MC seen when HIV is acquired by IV drug use. Presents as focal segmental glomerulonephritis. Early treatment with antiretroviral
Renal Amyloidosis	Diagnose with birefringence on congo red stain. Treat with a transplant
LUPUS	
Type 1	No renal involvement
Type 2	Is a mesangial disease with focal segmental glomerular pattern. Treatment isn't typically required
Type 3	Is a focal proliferative disease. Treat aggressively with prednisone and cyclophosphamide
Type 4	Diffuse proliferative disease. Combination of both nephritic and nephritic disease. Wire-loop abnormality on LM Treat with prednisone and cyclophosphamide
Type 5	Is a membranous disease that is indistinguishable from other primary membranous diseases. Treat with prednisone

Renal Artery Stenosis

- Presents with sudden onset of hypertension along with hypokalemia
- Abdominal bruit heard with stethoscope
- Can be caused by plaque, fibromuscular dysplasia

Diagnosis:

- ***Best initial test*** is a renal ultrasound with Doppler
- If small kidneys are then seen, do an MRA
- The ***most accurate test*** is a renal angiogram

Treatment:

- Angioplasty and stenting

Obstruction of the Urinary Tract

- BPH and stones are the MCC in adults
- There is an increased risk of stasis thus increasing the risks of UTI's
- Present with urinary colic, intense pain that radiates from the back around to the pelvis and the groin

Nephrolithiasis:

Calcium Pyrophosphate:

- 85% of stones are calcium pyrophosphate
- Are radiopaque and associated with hypercalcemia
- 50% of time it's associated with hypercalciuria
- Treat calcium stones with hydration and loop diuretics (LOOPS eliminate calcium)

Ammonium Mangesium Phosphate (Struvite):

- 2nd MCC of stones
- Are radiopaque
- Usually caused by urease (+) Proteus or Staph Saprophyticus
- May form large staghorn calculi
- Treatment involves taking care of the underlying infection

Uric Acid Stones:

- Often secondary to gout or conditions that cause increased cell turnover, such as myeloproliferative disease
- Treat by alkalinizing the urine and/or treating any underlying conditions

Kidney Tumors

- The most common renal malignancy is **renal cell carcinoma**, which occurs MC in males from 50-70yr of age
- Presents with hematuria, flank pain, fever, palpable mass, and secondary polycythemia
- Treatment involves interleukin and resection
- The most common childhood renal malignancy is **Wilm's tumor**, which occurs MC between 2-4yr old
- Presents with a palpable flank mass
- Part of WAGR complex (Wilms, Aniridia, GU malformation, Retardation – mental and motor)

Treatment:

- Removal of kidney plus chemotherapy and/or radiation

Chapter 14

Hematology

Anemias:

Mild to moderate anemia presents with:

- Fatigue, pallor, pale conjunctiva, flow murmur

Severe anemia presents with:

- SOB, light-headedness, confusion

Diagnosis:

- The most reliable test for iron deficiency anemia is serum ferritin, which will be decreased.
- If a patient has iron-deficiency anemia and does not respond to treatment, do hemoglobin electrophoresis to look for an α or β thalassemia

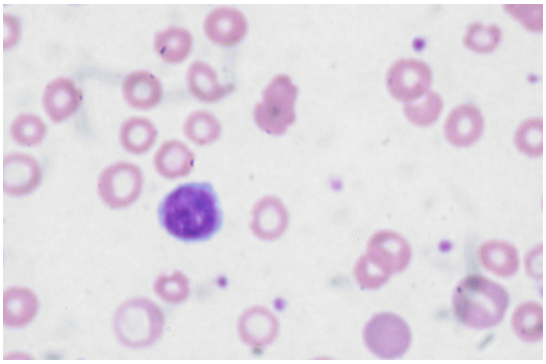
LABS for suspected anemias:

1. Iron studies (most imp)
2. CBC w/ peripheral smear (paying attention to MCV, MCHC)
3. B12/folate(B12 = neuropathies, folate = no neuropathies)
4. RDW (newer smaller RBCs cause change in RDW)
5. Reticulocyte count (determines whether site of problem is bone marrow synthesis of new RBCs)
6. LDH, bilirubin, haptoglobin (all determine whether hemolysis is in play)
7. TSH with T4 (see whether hypothyroidism is cause of fatigue)
8. CXR/blood culture/UA for suspected sickle cell disease

Iron deficiency Anemia

Signs and Symptoms:

- Fatigue
- Pallor
- Pale conjunctiva
- Low ferritin, low iron, increased TIBC



Diagnosis:

- Best diagnostic test for iron-deficiency anemia is iron studies
- The most accurate test is a bone marrow biopsy

Treatment:

- Oral ferrous sulfate supplement

Anemia of Chronic Disease

Signs and Symptoms:

- Same signs and symptoms with the addition of a history of chronic inflammatory disorder or autoimmune disorder

Diagnosis:

- Best diagnostic test is iron studies (will come back normal)

Treatment:

- Correct the underlying disease

Thalassemia

Signs and Symptoms:

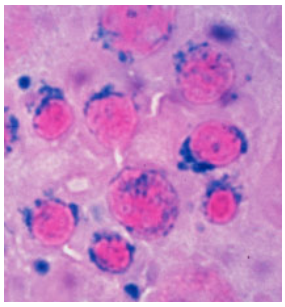
- Small MCV
- Presence of target cells
- Very other symptoms

Diagnosis:

- Best initial test is an iron study
- The most accurate test is hemoglobin electrophoresis
- Beta-thalassemia has elevated HgA2 and HgF
- Alpha-thalassemia has normal levels

There is no treatment for these conditions

Sideroblastic Anemia



Signs and Symptoms:

- Patient has a history of alcohol abuse, exposure to lead, or the use of isoniazid (INH)

Diagnosis:

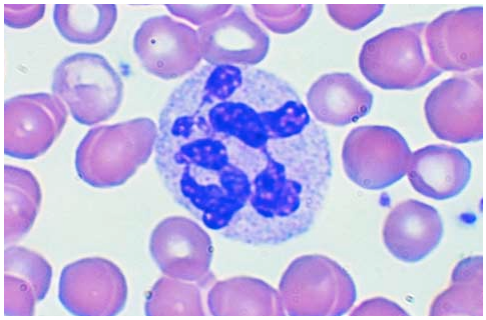
- Best initial test is iron studies
- The most accurate test is the Prussian blue stain

Treatment:

- Minor cases require only pyridoxine replacement
- Severe cases require the removal of exposure to toxin

Macrocytic Anemia

- Presents similary to microcytic anemia with fatigue, pallor, light-headedness, but is caused by a deficiency of either vitamin B12 or folic acid



Vitamin B12 Deficiency:

Signs and Symptoms:

- Parasthesias
- Peripheral neuropathies
- Dementia is the least common occurrence

Diagnosis:

- CBC with peripheral smear, paying special attention to neutrophils
- Neutrophils are large and hypersegmented
- B12 deficiency can also cause glossitis and diarrhea

Treatment:

- Replace either folate or vitamin B12

NOTE:

Approx 30% of B12 deficiencies show normal B12 levels because transcobalamine is an acute phase reactant which elevates with any form of stress. Thus is you suspect B12 deficiency and levels are normal, get a methylmalonic acid level.

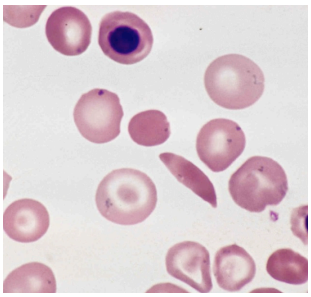
After finding low B12 or elevevated methylmalonic acid, the ***best confirmatory test*** *is* antiparietal cell antibodies or anti-intrinsic factor antibodies (both confirm pernicious anemia as the cause of B12 def).

Treatment:

- Replace B12 or folate

** Folate deficiency is most commonly caused by a poor diet, classically described as a "tea and toast diet". Folate stores deplete within 3 months, thus with poor diets this can be seen quickly. Treat this with diet modifications and immediate folate replacement.

Sickle Cell Anemia



- Patient presents with extreme pain in the chest, back, and thighs
- When a patient presents with a sickle cell crisis, give immediate oxygen, normal saline, and analgesics.
- If patient presents with these symptoms + fever, give IV antibiotics as well

When to do exchange transfusion in sickle cell pt?

1. Presence of visual disturbances due to retinal infarct.
2. Pulmonary infarct leading to pleuritic pain and abnormal xray.
3. Priapism due to infarct of prosthetic plexus of veins.
4. Stroke

What causes sudden drops in hematocrit in sickle cell pt?

- Either due to a folate deficiency or parvo B19 virus, which causes an aplastic crisis
- All Sickle cell pts should be on folate supplements, thus if that's the case it is due to parvo B19
- Do a PCR for DNA of parvo B19

Treatment

- Transfusion and IVIG

What should all sickle cell patients who are being discharged be given?

- Folate replacement
- Pneumococcal vaccine
- Hydroxyurea (This increases hemoglobin F, stops sickling of cells, and prevents further crises)

Hemolytic Anemia

All forms of hemolytic anemia present with a sudden onset of weakness and fatigue that is associated with anemia.

- Are premature destruction of red blood cells
- Bone marrow responds appropriately by increasing erythropoiesis and thus there is an increase in reticulocyte count
- If bone marrow doesn't work properly, anemia will ensue

Hemolysis will show the following:

1. Increased indirect bilirubin (Gets released with RBC destruction)
2. Increased reticulocyte count (Determines bones marrow's ability to make new RBCs)
3. Increased LDH
4. Decreased haptoglobin (ordered to distinguish between anemia and hemolytic anemia)

Intravascular hemolysis will also show the following:

1. Abnormal peripheral smear (Schistocytes, helmet cells, fragmented cells)
2. Hemoglobinuria
3. Hemosiderinuria (Metabolic, oxidized product or hemoglobin in urine).

Causes: Factors external to RBC defects OR intrinsic RBC defects

1. *Factors external to RBC defects (Most cases acquired)*

- Immune hemolysis
- Mechanical hemolysis (caused by prosthetic heart valves)
- Medications, burns, and toxins

2. *Hemolysis due to intrinsic RBC defects (Most cases inherited)*

- Caused by sickle cell disease, hemoglobin disease, thalassemias
- Membrane defects such as hereditary spherocytosis, paroxysmal nocturnal hemoglobinopathy
- Enzyme defects such as G6PD deficiency and pyruvate kinase deficiency

Autoimmune Hemolysis

- Patient often has a history of autoimmune diseases, cancers, or medication use

Diagnosis:

- Most accurate test is Coomb's test
- Look for an increased LDH and increased reticulocyte count
- Look at the peripheral smear for spherocytes

Treatment:

- The best initial therapy is prednisone
- With no response to prednisone, IVIG can stop acute episodes
- With recurring episodes of hemolysis, a splenectomy is most effective

*NOTE: warm antibodies are the cause here, which are always IgG. ***Only IgG responds to steroids and splenectomy.***

Cold-Induced Hemolysis (Cold agglutinins)

- Coomb's test is negative
- There is often a mycoplasma or EBV infection
- There is no response to steroids, splenectomy, or IVIG

Hemolysis due to intrinsic defects

Glucose-6-Phosphate dehydrogenase deficiency

- Presents with a severe and acute onset of hemolysis
- Is an x-linked disorder, thus seen in males
- Most commonly there is a history of sulfa drugs, primaquine, or dapsone use
- Ingestion of fava beans is classically asked on board exams

Diagnosis:

- The best initial test is the peripheral smear that shows Heinz bodies and bite cells
- The most accurate diagnostic test is a glucose-6-phosphate level. The problem with this test is that it will only show up after 2 months, and is not a good test early on in an acute haemolytic episode

Treatment:

- Avoidance of oxidative stress

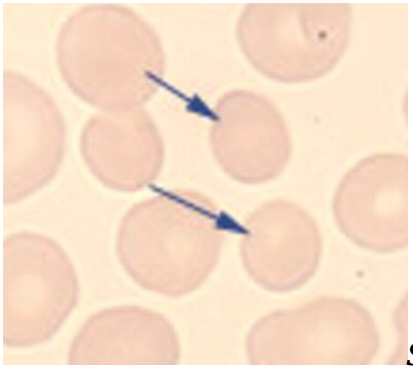
Pyruvate Kinase Deficiency

Presents the same way as G6PD deficiency, but the cause is unknown

Hereditary Spherocytosis

Signs and Symptoms:

- Jaundince in childhood
- Splenomegaly
- Bilirubin gallstones
- Recurrent episodes



Spherocytes

Diagnosis:

- The most accurate and best initial test is the osmotic fragility test
- Peripheral smear showing spherocytes

Treatment:

- Folic acid supplementation
- Splenectomy for severe disease

Hemolytic Uremic Syndrome (HUS)

- **HUS** in kids, there is usually a history of E.Coli 0157:H7

Signs and Symptom:

- Acute renal failure
- Abdominal pain

- Bloody diarrhea
- Seizures

Treatment:

- Dialysis in children, adults this isn't useful and there is a much poorer prognosis

Thrombotic Thrombocytopenic Purpura (TTP)

- Is an idiopathic disease that is often seen in HIV patients

Signs and Symptoms:

There is a pentad of:

- Haemolytic anemia
- Renal failure
- Fever
- Thrombocytopenia
- Neurological diseases

Treatment:

- Plasma exchange until symptoms subside
- Without treatment this is fatal

Paroxysmal Nocturnal hemoglobinuria (PNH)

Presents with recurring episodes of dark urine, mostly seen in the morning.

Signs and Symptoms:

- Pancytopenia
- Recurring episodes of dark urine in the morning
- Portal vein thrombosis is a complication that leads to death
- May progress to cause aplastic anemia and/or AML

Diagnosis:

- The most accurate test is the presence of decay accelerating factor antibody

Treatment:

- Prednisone or other steroids

Methemoglobinemia

- Blood locked in the oxidized state cannot pick up and transport oxygen.
- Patient will present with shortness of breath with no reason

Signs and Symptoms:

- SOB with no known cause (There will be clear lungs on exam with a normal CXR)
- Blood (if seen) will have a brownish appearance, which indicates it is locked in the oxidized state.

Diagnosis:

- Look for exposure to drugs like nitroglycerin, amyl nitrate, or nitroprusside
- CXR and PE show no reason for SOB
- Look for a history of anesthetic use

Treatment:

- Give methylene blue

Transfusion Reactions

1. **ABO incompatibility** → presents with acute symptoms of hemolysis while the transfusion is happening. Ex: during a transfusion, a patient becomes hypotensive and tachycardic. She has back and chest pain, and there is dark urine. LDH and bilirubin are elevated, and haptoglobin are low.
2. **Transfusion related acute lung injury(leukoagglutination rxn)** → presents with acute SOB from antibodies in the donor blood against recipient white cells.
3. **IgA Deficiency** → presents with anaphylaxis. In future use donation from IgA deficient donor or washed red cells.
4. **Febrile nonhemolytic rxn** → results in a small rise in temperature and needs no therapy, the reaction is against the donor's white cell antigens. Prevent by using filtered blood transfusion in future.
5. **Minor blood group incompatibility** → results in delayed jaundice, no therapy needed.

Leukemia

Acute leukemias present with signs of pancytopenia, such as fatigue, bleeding, and infections from non-functional white blood cells.

Acute Myelogenous Leukemia

- AUER RODS
- Mostly occurring in adults (up to 80%)

Diagnosis:

- The best initial test is peripheral smear showing blasts

Treatment:

- Best initial therapy for AML is ***Idarubicin*** (or daunorubicin) and ***cytosine arabinoside***

Acute Lymphoblastic Leukemia

- The most common malignancy in children
- Is the leukemia most responsive to therapy
- Is a neoplasm of early lymphocytic B cell precursors
- Histology reveals predominance of lymphocytes
- Poor prognosis when age < 2 or > 9, WBC's >10^5, or CNS involvement

Diagnosis:

- The best initial test is peripheral smear showing blasts

Treatment:

- **Idarubicin** + **cytosine arabinoside** + **intrathecal methotrexate**.

Acute Promyelocytic Leukemia(M3)

This leukemia is associated with disseminated intravascular coagulopathy

Treatment:

- **Idarubicin + cytosine arabinoside + all trans retinoic acid(ATRA)**

Chronic Myelogenous Leukemia(CML)

Signs and Symptoms:

- Elevated white cells that are predominantly neutrophils
- Splenomegaly is frequent.
- Untreated CML has the highest risk of transformation into acute leukemia of all forms of myeloproliferative disorders.
- **Associated with Philadelphia Chromosome**

Diagnosis:

- Best initial test is **Leukocyte alkaline phosphatase score (LAP score**). An elevated PMN count with low LAP score is CML. Reactive high white counts from infection give an elevated LAP score, LAP is up in normal cells, not CML.
- The most accurate test is finding the Philadelphia Chromosome

Treatment:

- Best initial treatment is Imatinib (Gleevec)
- Bone marrow transplant is the only cure for CML, but is never the best initial therapy, because Imatinib leads to 90% hematologic remission with no major adverse effects.

Chronic Lymphocytic Leukemia(CLL)

Seen in people older then 50yr of age

Signs and Symptoms:

- Often asymptomatic
- Organomegaly
- Haemolytic anemia
- Thrombocytopenia

Diagnosis:

- Best initial test is the peripheral smear showing 'smudge cells'

Treatment:

- At stages 0 and 1, there is no treatment required
- At advanced stages give fludarabine or chlorambucil

Hairy Cell Leukemia

Seen in middle-aged people and presents with massive splenomegaly and pancytopenia

Diagnosis:

- The Most accurate test is the tartrate resistant acid phosphatase(TRAP) smear showing hairy cells

Treatment:

- The best initial therapy for Hairy Cell leukemia is cladribine or 2-CDA

Myelofibrosis

- Presents similarly to hairy cell leukemia except there will be a normal TRAP level
- The key diagnostic feature is the "tear-drop" shaped cells on peripheral smear
- There is no specific therapy for myelofibrosis

Polycythemia Vera (Erythrocytosis)

This presents with headache, blurred vision, dizziness, and fatigue. All due to thickened blood

Signs and Symptoms:

- Everything above
- Pruritis following hot showers or baths due to histamine release
- Splenomegaly

Diagnosis:

- Very high hematocrit with a low MCV
- Get an ABG to rule out or in hypoxia as a cause of erythrocytosis

Treatment:

- The best initial therapy is phlebotomy
- Hydroxyurea can be given to lower the cell count
- Give daily aspirin

PLASMA CELL DISORDERS

Multiple Myeloma

This condition presents most commonly with bone pain due to fractures occurring from normal use

Diagnosis:

- The most specific test is a bone marrow biopsy
- Skeletal survey to detect punched out osteolytic lesions
- Serum protein electrophoresis(SPEP) to look for elevated monoclonal antibody (usually IgG)
- Urine protein electrophoresis(UPEP) to detects Bence-Jones proteins
- Peripheral smear showing "rouleaux" formation of blood cells.
- Elevated calcium levels due to osteolytic lesions
- Beta 2 microglobulin level is a prognostic indicator
- BUN and Creatinine to detect renal insufficiency

Treatment:

- Steroids and Melphalan
- The most effective therapy is bone marrow transplant
- Treat all underlying co-morbidities

Waldenstrom's Macroglobulinemia

This is a hyperviscosity of the blood due to overproduction of IgM

Signs and Symptoms:

- Blurred vision
- Confusion
- Headache
- Enlarged lymph nodes
- Splenomegaly

Diagnosis:

- The best initial test is the serum viscosity (increased significantly) and SPEP for IgM levels
- There will be no specific finding on the CBC

Treatment:

- Plasmapharesis is the best initial therapy
- Can also give fludarabine or chlorambucil

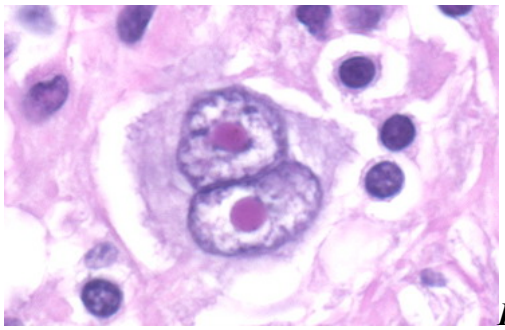
Aplastic Anemia

- Presents as pancytopenia with no identifiable etiology.
- If pt is < 50 and has a match, best therapy is BM transplantation.
- If BM transplant isn't an option (>50), give antithymocyte globulin and cyclosporine.

LYMPHOMAS

Present with enlarged lymph nodes that are most commonly seen in the cervical area

Hodgkin's disease occurs in a bimodal age distribution (young and old), and is characterized by Reed-Sternberg cells



Reed-Sternberg Cell

Non-Hodgkin's disease is commonly seen in HIV patients

Diagnosis:

- The best initial diagnostic test for both types of lymphomas is lymph node biopsy

Once excisional biopsy shows abnormal architecture, further testing to determine stage of the lymphoma needs to be done.

1. Stage 1 → single lymph node group
2. Stage 2 → 2 LN groups on one side of diaphragm
3. Stage 3→ LN involvement on both sides of diaphragm
4. Stage 4→ widespread disease

HD and NHL present with stages as follows:

HD - 80-90% pres w/ stage 1 and 2

NHL - 80-90% pres w/ stages 3 and 4

How to stage: CXR, CT with contrast (Chest/abdomen/pelvis/head), and BM biopsy.

Treatment:

1. ***Localized disease*** (stage 1,2) without "B" symptoms is treated predominantly with radiation.
2. ***More advanced stages*** 3,4 is treated with chemotherapy.

B SYMPTOMS:

Are systemic symptoms such as:

FEVER

Specific treatments:

1. HD: ABVD (adriamycin[doxorubicin], bleomycin, vinblastine, dacarbazine)

2. NHL: CHOP (cyclophosphamide, hydroxyadriamycin, oncovin[vincristine], and prednisone)

* Also test for anti-CD20 antigen and if present, add Rituximab, which adds efficacy to CHOP.

COAGULATION DISORDERS

Von Willebrand's disease (VWD)

Is due to platelet dysfunction, not a lower number of platelets

- Bleeding from PLT dysfunction, superficial bleeds from skin and mucosal surfaces, such as gingival, gums, and vagina.
- Epistaxis common
- Bleeding often worsened with use of aspirin
- PLT count is normal

Diagnosis:

- ***Best initial test*** is platelet function test
- The ***most accurate test*** is a ristocetin cofactor assay and a VWF level (If VWF is normal, ristocetin tells you if it is working properly)

Treatments:

- First line treatment is desmopressin or DDAVP, which causes the release of subendothelial stores of VWF and co-factor VIII
- If desmopressin doesn't work, give factor VIII replacement

Platelet-type bleeding causes: petechiae, epistaxis, purpura, gingival, gums, vaginal

Factor-type bleeding causes: hemarthroses, hematoma

Idiopathic Thrombocytopenic Purpura (ITP)

ITP is a diagnosis of exclusion that presents with platelet-type bleeding and a platelet count of < 50,000

Diagnosis:

- Perform a sonogram to assess the size of the spleen
- Check for anti-platelet antibodies
- Bone marrow biopsy looking for megakaryocytes
- Antibodies to glycoprotein IIb/IIIa receptors

Treatment:

- With mild ITP (platelets of ~ 20,000) give prednisone
- With severe ITP (platelets of < 20,000) give IV immunoglobulins, which is the fastest way to increase the platelet count
- NEVER transfuse platelets because this exacerbates the condition

Uremia-Induced PLT dysfunction

This presents as platelet-type bleeding in a patient with renal failure, where renal failure causes uremia, which prevents the degranulation of platelets and thus stops them from working

Signs and Symptoms:

- Platelet type bleeding

Diagnosis:

- Ristocetin and VWF levels (normal)
- Check platelet levels (They will be normal)
- Look for renal failure (This is the key to diagnosing)

Treatment:

- Desmopressin

Heparin-Induced Thrombocytopenia (HIT)

A condition where platelets drop at least 50% a few days after the use of heparin

Signs and Symptoms:

- The most common clinical manifestation is thrombosis, where venous thrombosis is the most common type

Diagnosis:

- The best initial diagnostic test is platelet factor 4 antibodies
- Other important test is heparin-induced anti-platelet antibodies

Treatment:

- The best initial therapy is to stop heparin and use direct thrombin inhibitor such as “argatroban” or “lepirudin”.

Chapter 15

Rheumatology

Rheumatoid Arthritis

An autoimmune disease presents most commonly in females > 50.

- Joint pain and morning stiffness that is symmetrical and in multiple joints of the hands.
- Lasts longer than 1hr in the morning with symptomatic episodes going on for at least 6 weeks.
- Often a prodrome of fatigue and malaise, but this isn't enough for a clear dx.

Diagnosis is based on having 4 or more of the following:

- Morning stiffness lasting more than 1hr
- Wrist and finger involvement
- Swelling of at least 3 joints
- Symmetric involvement
- Rheumatoid nodules
- Xray abn's showing erosions
- (+) rheumatoid factor

Rheumatoid arthritis is a group of physical findings, joint problems, and lab tests. There is no single diagnostic criteria to confirm the diagnosis, nor is there one single therapy to control and treat the disease.

Diagnosis:

- The single most accurate lab test is the anticitrullinated cyclic peptide (anti-CCP)
- Normocytic, normochromic anemia is very characteristic of rheumatoid arthritis

Joint Findings:

- MCP swelling and pain
- Boutonierre deformity: flexion of PIP w/ hyperextension of the DIP
- Swan neck deformity
- Baker's cyst

- C1/C2 cervical spine subluxation
- Knee: although knee is commonly involved, multiple small joints are involved more commonly over time.

Treatment:

- NSAIDs and Disease modifying anti-rheumatic drugs(DMARDS)
- Steroids can be used acutely to help control the disease while DMARDs take effect, but they are only used as a bridge to DMARD therapy, not used long-term

Seronegative Spondyloarthropathies

1. Ankylosing spondylitis
2. Reactive arthritis (Reiters syndrome)
3. Psoriatic arthritis
4. Juvenile RA

This group of inflammatory conditions all the following characteristics:

- Negative for RF
- Predilection for the spine
- SI joint involvement
- Associated with HLA B27

Ankylosing Spondylitis(AS)

- AS presents in a young male (<40) w/ spine or back stiffness.
- Peripheral joint involvement is less common
- Pain is worse at night and relieved by leaning forward, which can lead to kyphosis and diminished chest expansion.
- Rare findings include: uveitis and aortitis.

Diagnosis:

- The best initial test is an xray
- The most accurate test is an MRI

Treatment:

- *NSAIDS*
- Sulfasalazine
- Biological agents (Infliximab, Adalimumab)

Reactive Arthritis(formerly Reiter's syndrome):

- Presents with asymmetric arthritis and a history of urethritis or GI infection.
- Patient may have constitutional symptoms such as fever, malaise, and weight loss.

Diagnosis:

- Clinical diagnosis based on the presence of the classic triad of urethritis, conjunctivitis, and arthritis

Treatment:

- NSAIDs

Psoriatic Arthritis:

- Presents as joint involvement with a history of psoriasis.
- Rheumatoid factor (-)
- SI joint is involved.

Key features of this disease are:

- Pitting of the nails
- Involvement of the distal interphalangeal joints
- "Sausage-shaped" digits

Treatment:

- Initial treatment should be with NSAIDs
- If this isn't working, add biological agents such as Infliximab
- If these don't work, try methotrexate

Juvenile RA (aka adult onset Still's disease)

There is no specific diagnostic test.

JRA is characterized by the following:

- Ferritin levels are high
- WBC count is elevated
- RF (-) and ANA (-)

Treatment:

- NSAIDs
- If unresponsive to NSAIDs, give methotrexate

Whipple's Disease

- Causes diarrhea and fat malabsorbtion, and is most commonly presented with joint pain
- The most specific test for diagnosis is a biopsy of the bowel

Treatment:

- TMP/SMX

Osteoarthritis

Osteoarthritis is the most common joint abnormality and is associated with excessive joint usage and increased age

Signs and Symptoms:

- Morning stiffness lasting < 30 minutes
- Joint crepitus seen
- Affects the distal interphalangeal joint (whereas RA does not)
- Heberden's nodes are seen as DIP osteophytes
- Bouchard's nodes are seen as PIP osteophytes

Diagnosis:

- The best initial diagnostic test is an xray of the joint

Treatment:

- NSAIDs
- Glucosamine sulphate and chondroitin sulphate are used to slow joint deterioration.

	OA	RA
Morning Stiffness	<30min	>1hr
DIP	YES	NO
PIP	YES	YES
MCP	NO	YES
RF, anti-CCP	NO	YES
Joint fluid	<2000	5000-50000

SLE

Classically, a rash + joint pain + fatigue = Lupus

There are 11 criteria for LUPUS, 4 must be present to make the diagnosis.

SKIN	- Malar Rash - Photosensitivity rash - Oral ulcers rash - Discoid rash
Arthralgias	Present in 90% of pts
Blood	Leukopenia, thrombocytopenia, hemolysis. Any blood involvement counts as 1 criteria.
Renal	Varies from benign proteinuria to ESRD.
Cerebral	Behavior changes, stroke, seizure, meningitis
Serositis	Pericarditis, pleuritic chest pain, pulmonary HTN, pneumonia, myocarditis.
Serology	- ANA (95% sensitive) - DS-DNA (60% sensitive) Each of the serologic abn's count as 1 criteria. If person has joint pain, rash, and both ANA and DS DNA, the pt has 4 criteria.

Diagnosis:

- The best initial diagnostic test is the ANA
- The most specific test is the anti-ds DNA or anti-Smith antibody

How to follow the severity of a lupus flare-up?

1. Complement levels will drop in flare-up
2. anti-ds DNA will rise in flare up

Treatment/Management:

- For acute flare-ups give prednisone and other glucocorticoids
- Give NSAIDs for joint pain
- If there is no response to those medications, antimalarials and hydrochloroquine will work
- For severe disease that recurs give cyclophosphamide and azthioprine

Drug-induced Lupus

The most likely causes of drug-induced lupus are hydralazine, procainamide, and isoniazid

Diagnosis:

- There will be anti-histone antibodies
- There is never renal or CNS involvement
- There are normal levels of complement and anti-ds DNA antibodies

Sjorgen's syndrome

Seen most commonly in women (9x greater in women than men)

Signs and Symptoms:

- Dry eyes, dry mouth
- There is a sensation of sand under the eyes
- Dysgeusia is common
- Commonly a loss of teeth due to the lack of adequate saliva

Diagnosis:

- The most accurate test is a lip biopsy
- The Schirmer test is will show inadequate lacrimation
- Commonly see "anti-Ro or anti-La antibodies

Treatment:

- Pilocarpine to increase Ach and thus increase lacrimation and salivary secretions

Scleroderma

Patient has tight skin, heartburn, and Raynaud's phenomenon

Signs and Symptoms:

- ***Skin findings*** include a tight, fibrous thickening that causes tight and immobile fingers as well as a tight face
- ***Joint Pain*** that is mild and symmetrical
- ***Raynaud's Phenomenon***, may result in ulcerations

Scleroderma can present diffusely with the following:

Lungs: fibrosis and pulmonary HTN(the leading causes of death in this disease)

GI: Wide mouth colonic diverticula and esophageal dysmotility, leading to reflux and Barrett's esophagus*.* 15% of pts get primary biliary cirrhosis

Heart: Restrictive cardiomyopathy

Renal: may lead to malignant HTN.

Diagnosis:

- There is not a single best diagnostic test
- Though not specific, ANA is seen in 95% of cases

Treatment:

- There is no treatment that slows the progression or treats this condition
- If there is renal involvement give ACEIs
- If there is pulmonary hypertension, give Bosentan and prostacyclin analogs
- For Raynaud's give calcium channel blockers

Eosinophilic Fasciitis

Is similar in presentation to scleroderma but there is no Raynaud's, nor is there any of the other systemic findings

Signs and Symptoms:

- Thickened skin similar to scleroderma
- Marked eosinophilia gives the appearance of peau d'orange

Treatment:

- Manage this condition with corticosteroids

Polymyositis(PM) and Dermatomyositis(DM)

In both conditions, the patient cannot get up from a seated position without using the arms. There can also be muscle pain and tenderness.

For polymyositis→ Weakness + increased CPK + increased aldolase

For dermatomyositis → Weakness + increased CPK + increased aldolase + skin rash.

Diagnosis:

- Clinical
- Should order LFT's and ANA

Treatment:

- Corticosteroids

Fibromyalgia

A pain syndrome seen mostly in females that presents with aching muscles, stiffness, trigger points for pain, and sleep that isn't refreshing

Signs and Symptoms:

- Muscle aches
- Muscle stiffness
- Depression and/or anxiety
- Extreme sensitivity and pain to trigger points on the body

Diagnosis:

- Normal blood tests and no objective evidence of disease

Treatment/Management:

- Exercise can help relieve some pain and stiffness
- TCA's can be given

Polymyalgia Rheumatica (PMR)

Seen most commonly in those > 50yr

Signs and Symptoms:

- Patient older than 50yr with significant pain and stiffness of the proximal muscles (shoulders, pelvic girdle)
- Stiffness is usually worse in the morning and is usually more localized to the muscles
- Patient may have fever, malaise, and weight loss

Diagnosis:

- CBC will likely show normocytic anemia
- Muscle biopsy and labs are all normal

- Lack of muscle atrophy

Treatment: Steroids

Following explains difference between chronic fatigue, fibromyalgia, and PMR

	Chronic fatigue syndrome	**Fibromyalgia**	**Polymyalgia rheumatica**
Fatigue/malaise	+++++ >6mnth	++	++
Nonrefreshing sleep	+++++	++	No
Trigger points	NO	YES	NO
Blood tests	All normal	All normal	Incr ESR
Treatment	None	Pain relief	Prednisone

Vaculitis

Common features among different types of vasculitis include:

- Fatigue, malaise, wt loss
- Fever
- Skin lesions such as palpable purpura and rash
- Joint pain
- Neuropathy

Common lab findings:

- Normocytic anemia
- Elevated ESR
- Thrombocytosis

Diagnosis:

- The most accurate test is a biopsy

Treatment:

- The best initial therapy is steroids

→ If steroids aren't effective, alternate and/or additional therapies are the following

- Cyclophosphamie
- Azathioprine/6-mercaptopurine
- Methotrexate

Polyarteritis Nodosa (PAN)

PAN has all the features of vasculitis described above, what differentiates it is the unique features that accompany PAN.

- Abdominal pain (65%)
- Renal involvement (65%)
- Testicular involve(35%)
- Pericarditis(35%)
- HTN(50%)

Diagnosis:

- The best initial test is angiography of the abdominal vessels
- The most accurate test is a biopsy of the muscles, sural nerve, and skin

Treatment:

- Prednisone and Cyclophosphamide

Wegener's Granulomatosis

A disorder that like PAN can affect the majority of the body. Look for the addition of upper and lower respiratory findings and the presence of c-ANCA

Diagnosis:

- The most accurate diagnostic test is a biopsy
- Upper and lower respiratory findings
- Presence of c-ANCA

Treatment:

- Treatment involves Prednisone and Cyclophosphamide

Churg-Strauss

- Can affect any organ in the body, but the key to making its diagnosis is presence of ***vasculitis, eosinophilia, and asthma.***
- P-ANCA and anti-myeloperoxidase can be positive too, but these findings aren't as unique as the presence of eosinophilia.
- Most accurate test is biopsy

Treatment:

- Steroids

Temporal Arteritis

A type of giant cell arteritis, related to PMR.

Signs and Symptoms:

- Fever
- Malaise
- Fatigue
- Weight loss
- Headache
- Visual disturbances
- Jaw claudication

Diagnosis:

- Elevation of ESR
- Clinical findings

Treatment:

- Steroids

Takayasu's Arteritis

Seen in young asian females

Signs and Symptoms:

- Dimishes pulses
- Vasculitis commonly seen before loss of pulse
- Patient often gets TIA and/or stroke caused by the vascular occlusion

Diagnosis:

- MRA or arteriography

Treatment:

- Steroids

Cryoglobulinemia

Is similar to to vasculitis syndromes, the difference is the association with hepatitis C and kidney involvement

Treat with interferon and ribavirin

Behcet Disease

This condition presents in pts of Middle Eastern or Asian ancestry.

Signs and Symptoms:

- Oral and genital ulcers
- Ocular involvement that can lead to blindness
- Skin lesions: "pathergy" which is hyperreactivity to needle sticks, resulting in sterile skin abscesses.

Diagnosis:

- There is no specific test for diagnosis, we must use the features seen above

Treatment:

- Prednisone and colchisine

INFLAMED JOINTS

The key to diagnosing inflamed joints is to look at the fluid within the joint. It is the most accurate diagnostic test for gout, pseudogout, and septic arthritis

Synovial fluid cell count

Normal	Inflammatory(gout/pseudogout)	Infectious
<2000 WBC's	2000-50000 WBC's	>50000 WBC's

Gout

Look for a man with a sudden onset of severe pain in the toe at night. The toe will be red, swollen, and tender.

Things that precipitate a gouty reaction are:

- Alcohol ingestion
- Thiazide diuretics
- Nicotinic acid
- Foods high in protein

Diagnosis:

- The best initial test is aspiration of the joint fluid
- The most accurate test is light exam of the fluid showing negatively birefringent needle-shaped crystals

Treatment:

- The best initial therapy for an acute gouty attack is NSAIDs.
- Give colchisine within 24hr of the attack

- Allopurinol is best as prophylaxis and prevention of future attacks, which works by reducing the levels of uric acid

Pseudogout (Calcium Pyrophosphate Deposition Disease)

- Involves most commonly the wrist and knees, but doesn't involve the toes
- Onset is much slower than gout, and doesn't acutely attack the patient

Diagnosis:

- Aspiration of joint fluid shows positively birefringent rhomboid-shaped crystals

Treatment:

- NSAIDs are the best initial therapy
- Can give colchisine, but is not as effective here as it is in gout

Septic Arthritis

- Any arthritic or prosthetic joint put patient at risk of septic arthritis
- With increased joint abnormality comes increased risk of sepsis

More risk factors for septic arthritis:

- Osteoarthritis is more of a risk than having normal joints
- RA is more of a risk than osteoarthritis
- Prosthetic joints are more of a risk than anything else.

Signs and Symptoms:

- Red, swollen, tender, immobile joint

Most common causes of septic arthritis:

- Staph aureus (40%)
- Strep (30%)
- Gram-ve bacilli (20%)

**Septic arthritis requires consult w/ orthopaedic surgeon.

Diagnosis:

- The best initial test is arthrocentesis showing > 50,000 WBC
- The most accurate test is a culture of the fluid

Treatment:

- Empiric IV antibiotics such as Ceftriaxone and Vancomycin once suspected

Paget's Disease of Bone

A condition with pain, stiffness, aching, and fractures associated with softening of the bones.

Signs and Symptoms:

- Stiffness
- Pain
- Fractures
- Bowing of the tibias

Diagnosis:

- The best initial test is for the elevation of alkaline phosphatase
- The most accurate test is xray of the bones

Treatment:

- Best treated with bisphosphonates and calcitonin

* In cases of Paget's, osteolytic lesions will be found initially. These may be replaced with osteoblastic lesions

** If you see osteolytic think Paget's or osteoporosis

*** If you see osteoblastic, think about metastatic prostate cancer in differential.

Baker's Cyst

A posterior herniation of the synovium of the knee

Signs and Symptoms:

- Patient often has osteoarthritis
- Palpation of the sac in the posterium of the knee
- Swelling of the calf is common (exclude a DVT when you see this)

Treatment:

- NSAIDS are mainstay of treatment
- Severe cases may require steroid injections

Plantar Fasciitis and Tarsal Tunnel Syndrome

Following table compares the two conditions:

Plantar Fasciitis	Tarsal tunnel syndrome
Pain on bottom of foot	Pain on bottom of foot
Very severe in morning, better w/ walking a few steps	More painful w/ use; may have sole numbness
Stretch the foot and calf	Avoid boots and high heels; may need steroid injection
Resolves spontaneously over time	May need surgical release

Chapter 16

Neurology

Strokes and TIA

Strokes:

- Strokes occur for greater ≥ 24hr and have permanent residual neurological deficits.
- Caused by ischemia most commonly, and hemorrhagic.
- Ischemic strokes occur from emboli or thrombosis, which occur more acutely

TIA:

- Presents the same as a stroke except it lasts <24hr.
- Commonly presents with a loss of vision in only one eye (Amaurosis fugax), due to emboli in the first branch off of the carotid (Ophthalmic artery)
- TIA's are always due to emboli or thrombosis, never hemorrhage

Diagnosing:

- The best initial test is always a CT of the head without contrast

Treatment:

- If possible, give thrombolytics within 3hrs of the onset of symptoms and the CT has ruled out hemorrhage
- Aspirin is the best initial therapy for those who come in too late for thrombosis
- If patient is already on aspirin, can give dipyridamole (Clopidogrel is an alternative)

**** There is no clear evidence that heparin benefits a stroke***

Arteries and their associated symptoms in stroke:

CEREBRAL ARTERY	SYMPTOMS
Anterior cerebral artery	-profound lower extremity weakness - mild upper extreme weakness - personality changes - urinary incontinence
Middle cerebral artery	- profound upper extremity weakness - aphasia - apraxia/neglect - eyes deviate towards lesion side*
Posterior cerebral artery	- contralateral homonymous hemianopia w/ macular sparing - prosopagnosia(cant recognize faces)
Vertebrobasilar artery	- vertigo - N/V - Vertical nystagmus - Dysarthria and dystonia - Sensory changes in face/scalp - Ataxia - Labile blood pressure
Lacunar infarct	- must be absence of cortical deficits** - Parkinson signs - Hemiparesis(most notable in face) - Sensory deficits - Possible bulbar signs
Ophthalmic artery	- amaurosis fugax

Further management of a Stroke or TIA:

After all initial managements are implemented, do the following:

- Echocardiogram
- Carotid Doppler
- EKG and Holter monitor if the EKG comes back normal

Young pts <50yr with no significant past medical history (DM, HTN) should have following done:

1. Sedimentaion rate
2. VDRL or RPR
3. ANA, dsDNA
4. Protein C, protein S, factor V leiden mutation, antiphospholipid syndromes

* The younger the patient, the more likely the cause of stroke is a vasculitis or hypercoagulable state.

Control HTN, DM, and hyperlipidemia in this pt:

Goals:

- HTN: <130/80
- DM: same tight glycemic control as gen pop'n
- Hyperlipidemia: LDL<100

TIA:

Same management as per stroke, except thrombolytics aren't indicated because the administration of thrombolytics are to resolve symptoms, so if symptoms are resolved they aren't necessary.

SEIZURES

For seizure disorders, the only clear diagnostic criteria is for a patient undergoing status epilepticus.

Therapy for status is as follows:

1. Benzodiazepine such as Ativan (Lorazepam)
2. If seizure persists, add fosphenytoin
3. If seizure persists, add phenobarbitol
4. If seizure persists, give general anesthetic such as pentobarbital.

Diagnosis in a patient having a seizure:

- Urgent CT of the head
- Urine toxicology secreen
- Chemistry panel
- Calcium levels
- Magnesium levels

Further management:

- If initial tests don't give clear cause, do an EEG (shouldn't be done first).

- Neurology consult required in any patient having or who had a seizure.

Long-term treatment of first-time seizures:

- If patient only had 1 seizure, chronic therapy is not required unless there is a strong family history, an abnormal EEG, or status epilepticus

Chronic antiepileptic therapy:

- There is no single agent considered to be the best therapy.

1st line therapies include:

- Valproate, carbamazepine, phenytoin, and levetiracetam are all equal in efficacy.
- Lamotrigine has same efficacy but can cause steven-johnson syndrome.

2nd line therapies:

- Gabapentin and phenobarbitol

For absence/petit mal seizures:

- Ethosuxamide is best.

Parkinson's Disease

Is a gait disorder with the following findings:

- Cogwheel rigidity
- Resting tremor
- Mask-like facies
- Orthostasis
- Intact cognition and memory

Diagnosis:

- There is no specific test to make the diagnosis of PD, rather the clinical findings are what is used to make the diagnosis

Treatment:

***Mild symptoms*:**

- In a patient < 60yr of age → Anticholinergics such as Benztropine or Hydroxyzine
- In a patient > 60yr of age → Amantadine (Older patients commonly develop bad reaction from anticholinergic medications)

***Severe symptoms*:**

- Levedopa/carbidopa have the greatest efficacy but has an "on-off" phenomenon with uneven long-term effects and more side effects
- Dopamine agonists such as Pramiprexole, Ropinerole, and Cabergoline are less efficacious but have fewer side effects

** If these medication don't work, use COMT inhibitors (Tolcapone, Entacapone) which inhibit metabolism of dopamine, which extends the effect of dopamine-based medication. Can also try MAOI's.

TREMOR

Type of tremor	Resting tremor	Intention tremor	Tremor at rest and with intention
Diagnosis	Parkinsons	Cerebellar disorder	Essential tremor
Treatment	Amantadine	Treat etiology	Propranolol

MULTIPLE SCLEROSIS

Multiple Sclerosis presents with abnormalities of any part of the CNS. The symptoms improve and resolve, only to have another defect develop months or years later.

Signs and Symptoms:

- The most common abnormality is optic neuritis
- There is commonly fatigue, hyperreflexia, spasticity, and depression
- There is also commonly optic neuritis

Multiple Sclerosis:

"SIN"

1. Scanning speech
2. Intention

Diagnosis:

- The best initial diagnostic test is an MRI
- The most accurate test is also the MRI
- If the MRI doesn't give you the diagnosis, do a lumbar tap

Treatment:

- Steroids are the best initial therapy to resolve an acute exacerbation
- Glatiramer and beta-interferon can both decrease the progression of MS

- Can give Amantadine to combat fatigue and Baclofen to combat spasticity

DEMENTIA

Alzheimers Disease

Is a slow progressive loss of memory exclusively in pts > 65yrs of age

Diagnosis:

- Anybody with memory lost requires a CT of the head, a TSH level, and an RPR/VDRL
- With Alzheimer's disease you will only see diffuse and symmetrical atrophy

Treatment

- Anticholinesterase inhibitors are the standard therapy of choice

Frontotemporal Dementia (Pick Disease)

- Personality and behaviour become abnormal first, then memory loss afterwards.
- Head CT or MRI shows focal atrophy of frontal and temporal lobes
- Treat this like Alzheimer's, but do not expect the same type of response

Creutzfeld-Jakob Disease (CJD)

- Caused by prions, which are transmissible protein particles.
- Manifests as rapidly progressive dementia and presence of myoclonus.
- This presents in those younger than the Alzheimer's patient
- EEG will be abnormal
- Most accurate test is brain biopsy
- CSF: shows 14-3-3 protein, the presence of this will spare the patient a brain biopsy.

Lewy Body Dementia:

Is Parkinson's disease + dementia.

Normal Pressure Hydrocephalus:

- This condition generally presents in older males, but it can affect women as well.
- It presents as a person with incontinence, with gait abnormalities, and with cognitive impairment

Diagnosis:

- Diagnosis should include a head CT and a lumbar puncture

Treatment:

- Shunt placement

Huntington's Disease/Chorea

- Presents in young patients (usually in 30's)
- There is usually a family history

Symptoms:

- Dementia
- Personality changes and psychological disturbance
- Choreic movements

Diagnosis:

- Specific genetic testing will show that inheritance is autosomal dominant

Treatment:

- There is no treatment

HEADACHE

Migraine: 60% are unilateral, often triggered by: cheese, caffeine, menses, OCP's

Do a CT or MRI if HA has any of the following:

- Sudden and/or severe
- Onset after 40yr
- Associated with any neurological findings
- May be preceeded by an aura and/or scotomata (Dark spots in visual field), and abnormal smells

Treatment:

- The best initial abortive therapy is sumatriptan or ergotamine
- Prophylactic therapy takes 4-6wks to work, if patient gets four or more HA per month, prophylax with propranolol.
- Alternate prophylactics with CCBs, TCA's, or SSRI.

Cluster: 10x more frequent in men than women. Are exclusively unilateral w/ redness and tearing of the eye and rhinorrhea.

Treatment:

- The best abortive therapy is 100% 02.
- This treatment is unique to cluster HA's.
- Sumatriptan can also be used in same way as is in migraines.

Prophylactic therapy: there is none because these HA's are numerous but short and intense, and the "cluster" would be over by the time the prophylaxis kicked in.

HA Type	MIGRAINE	CLUSTER
Gender		Men 10x more than women
Presentation	Unilateral or bilateral, aura	Only unilateral, tearing/redness of eye
Abortive	Sumatriptan	Sumatriptan, Special: 100%02
Prophylactic	Propranolol	none

* Sumatriptan is similar to 5HT, and works by causing vasoconstriction in cerebral arteries.

Temporal Arteritis

Patient will present with tenderness over the temporal area and may also complain of jaw claudication

Diagnosis:

- First check the ESR
- The most accurate test is a biopsy of the temporal artery

Treatment:

- Give steroids immediately, do not delay if this is suspected

Pseudotumor Cerebri

- This presents most commonly in a younger woman with a headache and double vision, papilledema
- CT/MRI show up normal
- Vitamin A use is often the cause

Diagnosis:

- LP is the most accurate test because it shows an elevated opening pressure

Treatment:

- Involves weight loss
- Acetazolamide can also be given

Dizziness/Vertigo

- All pts with vertigo will have a subjective sensation of the room spinning around them.
- Often associated with nausea and vomiting
- All pts with vertigo will have nystagmus
- Generally all patients wtih vertigo should have MRI of the internal auditory canal.

Following table summarizes pres of a number of vertigo-causing conditions:

Disease	Characteristics	Hearing Loss/ Tinnitus
BPV	Changes w/ position	NO
Vestibular neuritis	Vertigo occurs w/o position change	NO
Labyrinthitis	Acute	YES
Meniere's disease	Chronic	YES
Acoustic neuroma	Ataxia	YES
Perilymph fistula	Hx of traum	YES

BPV

- Vertigo alone w/ no loss of hearing,no tinnitus, no ataxia.
- Positive dix-hallpike maneuver
- Treat with meclizine(antivert)

Vestibular Neuronitis

- An idiopathic inflammation of the vestibular portion of CN8.
- No hearing loss or tinnitus because only vestibibular portion is affected.
- Most likely to be a viral cause
- Not related to change in position
- Treat with meclizine

Labyrinthitis

- Is inflammation of the cochlear portion of the inner ear.
- There is hearing loss as well as tinnitus.
- Is acute and self-limited
- Treat with meclizine

Meniere's Disease

- Same pres as labyrinthitis (vertigo, tinnitus, hearing loss) but meniere's is chronic with remitting and relapsing episodes.
- Treat with salt restriction and diuretics.

Acoustic Neuroma

- A tumor of CN8 that can be related to neurofibromatosis (von Recklinghaus's).
- Presents with ataxia in addition to hearing loss, tinnitus, and vertigo.

Diagnosis:

- MRI of internal auditory canal

Treatment:

- Surgical resection.

Perilymphatic Fistula

- Head trauma or any form of barotraumas to the ear may rupture the tympanic membrane and lead to a perilymph fistula.

Wernicke-Korsakoff Syndrome

Presents with the following:

1. History of chronic heavy ETOH use
2. Confusion with confabulation
3. Ataxia
4. Memory loss
5. Gaze palsy and/or ophthalmoplegia
6. Nystagmus

Diagnosis:

- Do a head CT
- Vitamin B12 level
- TSH/T4 level
- RPR/VDRL
- Presence of memory loss

Treatment:

- 1st give thiamine then give glucose.

CNS INFECTIONS

Often when a CNS infection is suspected, a head CT should be performed before the LP.

This is the case in the following circumstances:

- A history of CSN disease
- Focal neurological deficits
- Presence of papilledema
- Seizures
- Altered consciousness
- Significant delay in ability to perform LP

** If these are present, get blood cultures and start empiric antibiotics before ordering CT.

CSF:

- Getting CSF is the most accurate test for bacteria meningitis, but cannot wait for culture to start therapy.
- Gram stain: only 50% sensitive, thus if (-) cannot exclude anything. Is highly specific though, so if (+) it is likely to be bacteria meningitis. Ie. Good spec bad sensitivity
- Gram (+) diplococci: pneumococcus
- Gram (+) bacilli: listeria
- Gram (-) diplococci: neisseria
- Gram (-) pleomorphic, coccobacillary organisms: hemophilus

Protein: An elevated protein level in CSF is of marginal diagnostic benefit. Elevated protein is non-specific because any form of CSN infection can elevate the CSF protein. However, a normal CSF protein excludes bacterial meningitis.

Glucose: Levels below 60% of serum levels is consistent w/ bacterial meningitis

Cell count: This is best initial test for the diagnosis of meningitis. If thousands of PMN's are present start IV ceftriaxone, vancomycin, and steroids. Thousand of PMN's is bacterial meningitis until proven otherwise.

Cryptococcus (fungal meningitis)

- Look for HIV(+) patient with <100CD4 cells.
- This infection is slower than bacterial meningitis and may not give severe meningeal signs (neck stiffness, photophobia, and high fever, all at same time).

Diagnosis:

- The best initial test is the India ink stain
- The most accurate test is the cryptococcal antigen

Treatment:

- The best initial therapy is Amphotericin

*follow Amphotericin with oral Fluconazole (continued indefinitely until the CD4 count raises), once CD4 count >100, fluconazole can be discontinued.

Lyme Disease

Patient usually lives in Connecticut area and has a history of camping, hiking, or being in tall grass. The tick exposure is rarely remembered by the patient.

Diagnosis:

- A central clearing target rash is pathognomonic
- There is a history of joint pain
- Bell's palsy

Treatment:

- Oral doxycycline
- If there is cardiac involvement treat with IV ceftriaxone

Rocky Mountain Spotted Fever (RMSF)

Look for camper/hiker with a rash that started on wrists and ankles and moved centrally.

Signs and Symptoms:

- Fever, headache, and malaise always precede the rash

Diagnosis:

- Oral Doxycylcine is the most effective therapy.

TB Meningitis

- This is an extremely difficult diagnosis to pinpoint
- Look for an immigrant with a history of lung TB
- Presents slowly over weeks to months (If acute then not TB meningitis)

Diagnosis:

- Has a very high CSF protein levels
- Acid fast stain of CSF is not accurate, need three high-volume centrifuged samples if going to do acid-fast stain.

Treatment:

- RIPE treatment as with TB, only diff is should add steroids and extend the length of therapy for meningitis when compared to the pulmonary disease.

Viral Meningitis

- Viral meningitis is in general a diagnosis of exclusion.
- There is lymphocytic pleocytosis in the CSF.
- There is no specific therapy for viral meningitis.

Listeria Meningitis

- Look for elderly, neonatal, and HIV (+) patients and those who have no spleen, are on steroids, or are immunocomprimised with leukemia or lymphoma.

* Add Ampicillin to regimen of vancomycin + ceftriaxone when Listeria is suspected. Such as if there is lymphocytosis in the CSF.

Neisseria Meningitides

- Look for patients who are adolescents, in the military, are asplenic, or who have terminal complement deficiency.

Treatment is as follows:

Patient → Respiratory isolation

Close contacts→ Start prophylaxis with rifampin or ciprofloxacin.

Close contacts are: household members, people who share utensils, cups, kisses.

Routine contacts→ Routine school and work contacts do not need to receive prophylaxis.

ENCEPHALITIS

- Look for a patient with a fever and altered mental status over a few hours.
- Almost all encephalitis in the US is from herpes, the patient does not have to recall a past history of herpes in order to make this diagnosis

FEVER + CONFUSION = Encephalitis

Diagnosis:

- The best diagnostic test is a CT scan of the head
- The most accurate test is a PCR of the CSF

Treatment:

- The best initial therapy is acyclovir
- For acyclovir-resistant patients give foscarnet

BRAIN ABSCESS

- Pres with a fever, headache, and focal neurological deficits
- CT finds a "ring" aka contrast enhanced lesion
- Finding a ring means either cancer or infection.

Consider HIV status in context of brain abscess as follows:

- HIV negative patient → brain biopsy is the next step
- HIV positive patient → Treatment for toxoplasmosis with pyrimethamine and sulfadiazine for 2wks and repeat the head CT

Progressive Multifocal Leukoencephalopathy (PML)

- These brain lesions in HIV(+) patients are not associated with ring enhancement or mass effect.
- There is no specific therapy.
- Treat the HIV and raise the CD4, when the HIV is improved, the lesions will disappear.

Neurocysticercosis

- Look for a patient from Mexico with a seizure
- Head CT shows multiple 1cm cystic lesions, over time lesions will calcify.

Diagnosis:

- Confirm with serology

Treatment:

- When still active and uncalcified, the lesions are treated with Albendazole, use steroids to prevent a reaction to dying parasites.

Head Trauma and Intracranial Hemorrhage

Any head trauma resulting in a loss of consciousness or altered mental status should lead to CT of head without contrast

	Concussion	**Contusion**	**Subdural hem.**	**Epidural hem.**
Focal deficit	Never	Rarely	Yes or no	Yes or no
Head CT	Normal	Ecchymosis	Crescent shape	Lens shaped

Treatment for various head traumas are as follows:

1. ***Concussion***: None
2. ***Contusion***: Admit patient, vast majority get no treatment
3. ***Subdural and Epidural Hematomas***: Large ones drained, small ones left alone to reabsorb on their own.
4. ***Large intracranial hemorrhage with mass effect***: 1. Intubate/hyperventilate to decrease ICP, 2. Decrease PC02 to 25-30, which constricts cerebral blood vessels, 3. administer Mannitol as an osmotic diuretic to decrease ICP, 4. Perform surgical evacuation.

Subarachnoid Hemorrhage(SAH)

Look for the following symptoms:

- Sudden, severe headache
- Stiff neck
- Photophobia
- LOC in 50% of patients
- Focal neurological deficits in 30% of patients
- SAH is like sudden onset of meningitis with a LOC but without fever.

Diagnosis:

- The best initial test is a head CT without contrast. It is 95% sensitive, if conclusive, no need to do an LP
- The most accurate diagnostic test is an LP, but not necessary if CT shows blood.

Treatment:

- Perform angiography to determine site of bleed
- Surgically clip or embolize the site of bleeding (If patient re-bleeds there is a 50% change of death).
- Insert a vetriculoperotoneal shunt if hydrocephalus develops
- Prescribe nimodipine orally; which is a CCB that prevents stroke.

** When SAH occurs, an intense vasospasm can lead to non-hemorrhagic stroke(thus CCB used).

SPINE DISORDERS

Lumbosacral strain	Cord compression	Epidural abscess	Spinal stenosis
Nontender	Tender	Tender and fever	Pain on walking downhill.

Syringomyelia

- Is a defective fluid cavity in the center of the cord from trauma, tumors, or congenital problem.
- Presents with a loss of sensation of pain and tenderness in a cape-like distribution over the neck, shoulders, and down both arms.

Diagnosis:

- MRI

Treatment:

- Surgical correction

Cord Compression

- Metastatic cancer presses on the cord, resulting in pain and tenderness of the spine.
- Lumbosacral strain doesn't give tenderness of the spine itself.

Diagnosis:

- The best initial test is an MRI
- The most accurate diagnostic test is a biopsy, only done if diagnosis is not clear from the history

Spinal Epidural Abscess

- Presents with back pain wth tenderness and fever
- Scan spine with an MRI
- Give antibiotics against staphylococcus such as oxacillin or nafcillin
- Large abscesss require surgical drainage.

Spinal Stenosis

- Presents with leg pain on walking and can look like peripheral arterial disease
- Pulses will be intact in spinal stenosis
- Pain worsens when patient leans backwards and/or walks in a downward direction, while it improves when walking in an upward direction
- Diagnose with an MRI and treat with surgical decompression

Anterior Spinal Artery Infarction

- All sensation is lost except position and vibratory sense, which travel down the posterior column of the spinal cord
- No specific therapy can correct this problem

Brown-Sequard Syndrome

- This results from traumatic injury to the spine, such as that from a knife wound.
- Patient loses ipsilateral position, vibratory sense, contralateral pain and temperature

** The most urgent management in cord compression is the administration of steroids as soon as possible and to relieve pressure on the cord. Imaging studies are done after administration of steroids.

AMYOTROPHIC LATERAL SCLEROSIS

- Is an idiopathic disorder of both upper and lower motor neurons.
- Treated with riluzole, a unique agent that blocks the accumulation of glutamate

UMN signs	LMN signs
Hyperreflexia Upgoing toes on plantar reflex Spasticity Weakness	Wasting Fasciculations weakness

PERIPHERAL NEUROPATHIES

Diabetes

- Diabetes is the most common cause of peripheral neuropathies
- Specific testing is not necessary in most cases.

Treatment:

- Gabapentin or Pregabalin are useful in treating neuropathies.
- TCA's are less effective and have more side effects

Carpal Tunnel Syndrome

- Look for pain and weakness of first three digits of hand
- Symptoms may worsen with repetitive use.
- Initial mgmt is a splint.
- Steroid injections may be used if splints provide no relief
- If these don't work, can perform surgery to relieve the pain

Radial Nerve Palsy

- Aka "*Saturday night palsy*", results from falling asleep or passing out with pressure on arms underneath the body or outstretched, perhaps draped over back of a chair (classic presentation on exam question)
- Results in wrist drop, resolves on its own.

Peroneal Nerve Palsy

- Results from high boots pressing at the back of the knee.
- Results in foot drop and inability to evert the foot.
- This palsy will resolve on its own

CN7 palsy (Bell's palsy)

- Results in hemi-facial paralysis of both upper and lower halves of face.
- Thought to be due to a virus
- There may also be a loss of taste in anterior 2/3 of tongue, hyperaccusis, and the inability to close the eye at night.

Treatment:

- Steroids and acyclovir or valcyclovir should be given

Reflex sympathetic dystrophy (chronic regional pain syndrome)

- Occurs in a patient with a previous injury to the extremity
- Light tough such as from a sheet touching the foot, results in extreme pain that is "burning" in quality

Treatment:

- NSAIDs
- Gabapentin
- Occasionally a nerve block may be done if the previous methods don't work
- Surgical sympathectomy may be necessary when refractory

Restless Leg Syndrome

- Uncomfortable feeling in the legs which patient tries to "shake off", which brings only temporary relief

Treatment:

- Pramipexole or ropinerole.

Guillain-Barre syndrome

- Ascending paralysis, thought to be caused by a viral infection.
- Usually presents weeks after a respiratory infection

Management/Treatment:

- First step is to take a peak inspiratory pressure, which can tell if the pt will undergo respiratory failure.
- Second step is to give IVIG's and/or perform plasmapharesis.

MYASTHENIA GRAVIS

- Classically presents with weakness of the muscles of mastication, making it hard to finish meals
- Blurry vision from diplopia results from inability to focus the eyes on a single target.
- Classically the patient reports drooping of the eyelids as the day progresses.

Diagnosis:

- The best initial test is testing for anti-acetylcholine receptor antibodies(ACHR)
- The most accurate test is *clinical presentation and ACHR*, which is more sensitive and specific that tensilon test.

Treatment:

- The best initial therapy is **Pyridostigmine** or **Neostigmine**
- Thymectomy can be performed if pyridostigmine or neostigmine don't work patients < 60 yr of age should undergo thymectomy.

- Give *Prednisone* if thymectomy doesn't work, or if there are no responses to pyridostigmine or neostigmine, then prednisone should be started.
- ***Azathioprine*** *and* ***cyclosporine*** are used to try to keep the patient off of long-term steroids.

Chapter 17

Oncology

Breast Cancer Screening

Screening mammography as follows:

- Start at 40
- Between 40-50 and every 1-2yrs
- At 50, start doing them every year

When a mammogram shows an abnormality?

- 1st thing is to do a biopsy(shows CA and presence of esetrogen and/or progesterone receptors)

***What is the sentinel node*?**

- It is the first node detected in the operative field, detected by dye.
- If this node is free of CA, then axillary node dissection is not necessary, if the node is cancerous, axillary lymph node biopsy is required

***Best initial therapy for breast cancer*?**

- Lumpectomy with radiation treatment(equal to modified radical mastectomy)
- If there is presence of estrogen or progesterone receptor (+), use tamoxifen or raloxifene
- Adjuvant chemotherapy used whenever the axillary nodes are (+) OR the cancer is >1cm in size.

***Hormonal Inhibition Therapy*:**

- **Tamoxifen** and **raloxifene** used if either progesterone or esterogen receptors are positive. They SERMs (selective esterogen receptor modulators). **Adverse Reactions**: DVT, hot flashes,endometrial CA.
- These are different from **aromatase inhibitors**, which do not lead to DVT, but cause osteoporosis due to antagonistic activity in the bone.

Adjuvant Chemotherapy: **Is appropriate when,**

1. Cancer is in the axilla
2. Cancer larger than 1cm
3. More efficacious when pt still menstruation, because BR-ca wont be controlled with estrogen antagonists such as tamoxifen.

TRASTUZUMAB: is a monoclonal antibody against breast CA antigen "HER-2/NEU", useful in metastatic disease, has modest activity with few adverse reactions

Primary Preventative Therapy: Use Tamoxifen in any patient with multiple 1st degree relatives (mom, sister) with breast CA

COLON CANCER

The most imp thing is screening schedule and indicators..

- Colon cancer is treated with surgical resection of the colon and chemotherapy centered around a 5-FU regimen.

***Routine for screening*:**

1. **Colonoscopy** starting at 50, the every 10yrs. If single family member has it, get colonoscopy 10yrs earlier than the age at which that person was diagnosed. Three family members get colonoscopy at 25yr, then every 10yr. FAP do screening sigmoidoscopy at 12yr, then every 1-2yr.
2. **Occult blood testing** starting at 50yr, then every year thereafter.
3. **Sigmoidoscopy** and **double contrast barium** enema at 50, then q 3-5yr.

LUNG CANCER

Remember that small cell carcinoma releases ectopic ACTH, Squamous cell carcinoma releases PTH-like hormone.

- There is no screening test for lung cancer
- Excisional biopsy should be done on solitary lung nodules in pts who are smokers with nodule >1cm.
- Calcifications usually go against malignancy, but if there is history of smoking, a patient >50yr, and nodule >1cm, excision is warranted.

Lung cancer therapy: The most imp issue in treatment is whether the disease is localized enough to be surgically resected. Surgery cannot be done is any of the following are present:

1. Bilateral disease
2. Mets
3. Malignant pleural effusion
4. Involvement of aorta, vena cava, or heart
5. Lesions within 1-2cm of carina.

*** ***Small-cell lung cancer*** is non-resectable because >95% of time is has one of these features.

CERVICAL CANCER

- Start pap smears at 21, or 3yrs after onset of sexual activity
- Do pap smears every 3 yrs until age 65. Stop at 65 unless there has been no previous screening.
- Administer HPV quadrivalent vaccine to all women 13-26yr

Following up an abnormal pap smear:

1. An abnormal pap smear with low-grade or high-grade dysplasia should be followed by colposcopy and biopsy.
2. A pap smear showing atypical squamous cells of undetermined significance (ASCUS), do HPV testing. If HPV (+), proceed to colposcopy, if HPV (-), do repeat pap in 6mnth
3. Once pap smear normal, return to routine testing

* Pap smears lower mortality in screen populations, but still not as much as mammography because incidence of cervical CA is less than that of breast cancer.

PROSTATE CANCER

- No proven screening method that lowers mortality rate. PSA and DRE not proven to lower mortality.
- Do not routinely offer these tests, however if pt asks for them, you should perform.

Treating prostate cancer:

1. ***Localized***: Surgery and either external radiation or implanted radioactive pellets (nearly equal in efficacy)

2. ***Metastatic***: Androgen blockade is standard of care, use flutamide (testosterone receptor blocker) AND leuprolide or goserelin (GNRH agonists).

* There is no good chemotherapy for metastatic prostate cancer, treatment is hormonal in nature.

*Remember the 5-alpha-reductase inhibitor finasteride is used for BPH, not cancer

A man with prostate cancer presents with severe, sudden back pain. MRI shows cord compression, and he's started on steroids. ***What's the next best step in mgmt?***

FLUTAMIDE (to block temporary flare up in androgen levels that accompanies GNRH agonist treatment

OVARIAN CANCER

Key feats are women >50 with increasing abdominal girth at same time as weight loss.

Diagnostic testing:

- There's no routine screening test
- CA125 is a marker of progression and response to therapy, not a diagnostic test

Treatment:

- Treatment is surgical debulking followed by chemotherapy, even in cases of local metastatic disease.
- Ovarian ca is unique in that surgical resection is beneficial even when there's a large volume of tumor spread through the pelvis and abdomen. If possible, removing all visible tumors helps.

TESTICULAR CANCER

- Presents with painless scrotal lump in a man <35yr.
- Is extremely curable with a 90-95% 5yr survival rate.
- NEVER do a biopsy of the testicle.

Diagnostic Testing:

- An inguinal orchiectomy of the affected testicle, never do biopsy.
- Measure AFP, LDH, and bHCG
- Stage with CT of abdomen and pelvis.

Treatment:

1. ***Local disease***: Radiation
2. ***Widespread disease***: Chemotherapy, which is curative of even metastasis in testicular cancer.

***Of all testicular cancers, 95% are germ cell tumors (seminoma and non-seminoma).**

*AFP secreted only by nonseminomas.

*Measure AFP, LDH, and bHCG.

SOME EXTRA PREVENTATIVE MEDICINE

Smoking cessation:

- Screen all and advise against smoking
- Most effective methods are use of oral meds such as Buproprion and Varenicline.
- Less effective are nicotine patch and gum (which should be tried first).

*With buproprion, patient should slowly decrease cigarettes 2wks after starting therapy. Use buproprion in conjunction with counselling and nicotine replacement.

Osteoporosis→ Screen all women with DEXA scan at 65yr of age.

AAA→ All men about 65yr who were ever smokers should be screened once w/ an ultrasound.

DM→ No recommendation for routine diabetes screening

HTN→ All pts should be screened at every visit.

Hyperlipidemia→ Men >35, Women >45

Chapter 18

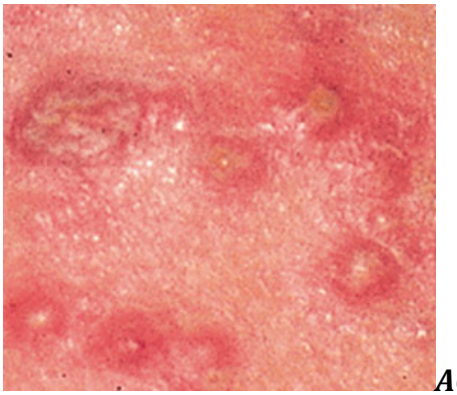
Dermatology

Using Topical Steroids

Potency	Medication	Use
Low	1% hydrocortisone	Face, genitalia, and skin
Moderate	0.1% triamcinolone	Body/Extremities, face, genitalia, and skin folds
High	Fluocinonide	For thick skin. Never on face
Very High	Diflorasone	Thick skin or severe body needs

Acne

- An infection of the pilosebaceous gland caused by the bacteria Propionibacterium acnes



ACNE

Signs and Symptoms:

- Blackheads (open comedones) and whiteheads (closed comedones)

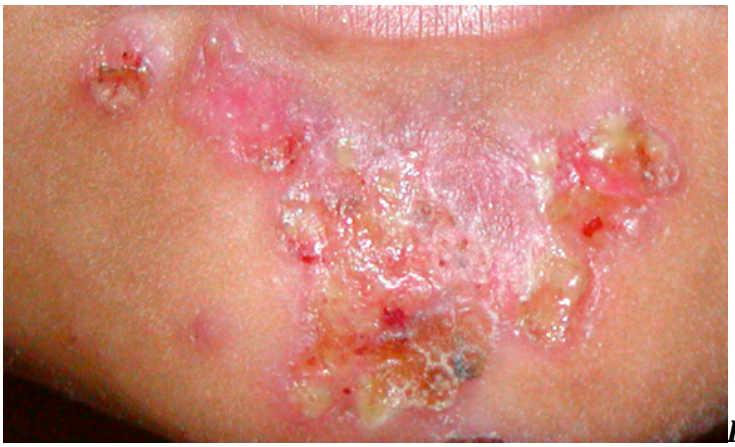
Treatment:

- Mild disease should be managed with topical antibiotics such as clindamycin, erythromycin, in addition to benzoyl peroxide
- Moderate disease should combine benzoyl peroxide with retinoids
- Severe disease should be managed with oral antibiotics and oral retinoic acid derivatives

Bacterial Infections of the Skin

Impetigo

A superficial skin infection limited to the epidermis



Impetigo

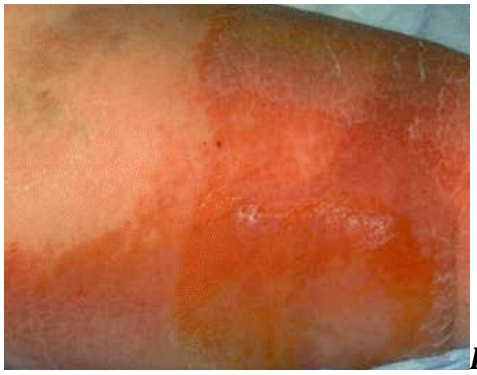
- Is often described as being "honey-colored", "wheeping", or "oozing"
- Usually caused by ***Staphylococcus***, but may also be due to ***Strep Pyogenes***

Treatment:

- Mupirocin (A topical antibiotic) and/or antistaphylococcal oral antibiotics

Erysipelas

A skin infection of the epidermis and the dermis, which is usually caused by Strep Pyogenes. This condition is often described as being bright red, angry, and swollen.

 ***Erysipelas***

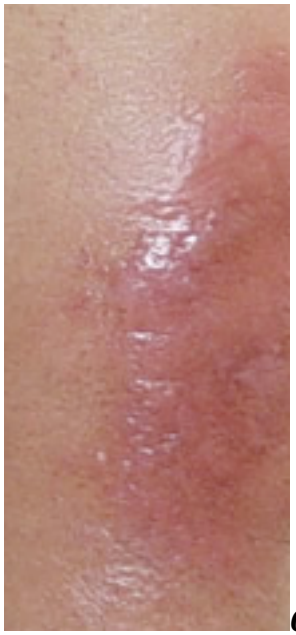
- With erysipelas may come fever and chills

Treatment:

- Penicillin G or ampicillin if diagnosis is Streptococcus

Cellulitis

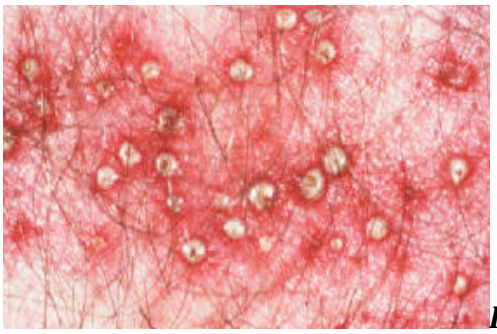
An infection caused by Staphylococcus and Streptococcus that infects the dermis and the subcutaneous tissues. Managed with antistaph drugs such as oxacillin and nafcillin.

 ***Cellulitis***

Folliculitis

An infection of the hair follicle, may progress to a worse infection called a furuncle. Usually caused by staphylococcus, if acquired from a hot-tub it can be due to Pseudomonas, fungi, or virus

Look for this around the beard area where there are accumulations of pus-like material.

 ***Folliculitis***

Treatment:

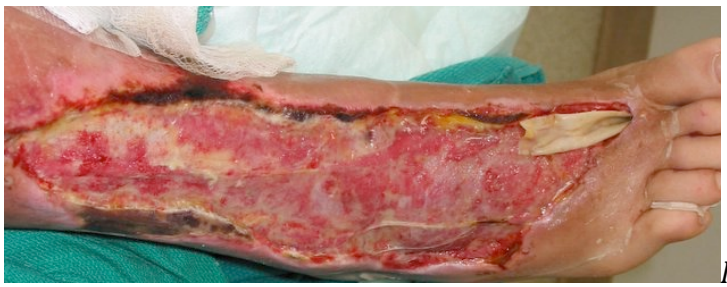
- Local care and topical mupirocin
- Severe cases that progress require systemic antistaphylococcal antibiotics

Necrotizing Fasciitis

Is a severe, life-threatening skin infection. It begins as a cellulitis that dissects into the fascial planes of the skin. MCC by strep and Clostridium

Presentation:

- High fever
- Extreme pain that is worse than it looks
- Bullae
- Crepitus


Necrotizing Fasciitis

Diagnosis:

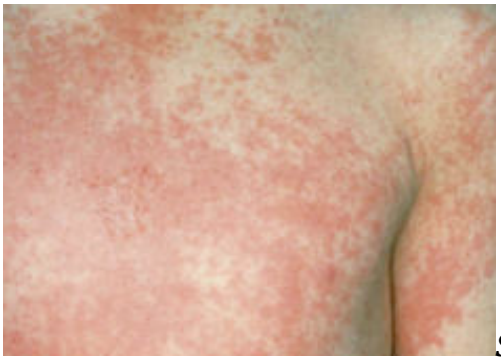
- CT or MRI looking for air in the tissue and/or necrosis
- Elevated creatine phosphokinase

Management/Treatment:

- Surgical debridement
- Combination beta lactam/beta lactamase medications
- If it is caused by Strep Pyogenes, give clindamycin + PCN

Scarlet Fever

Is caused by Strep Pyogenes, and looks like a combination of a rash and goosebumps

 ***Scarlet Fever***

Signs and Symptoms:

- Rough skin
- Strawberry tongue
- Beefy-red pharynx
- Rash is most intense in the axilla and groin (In the creased areas of skin)
- Desquamation of hands and feet occurs as rash resolves
- May have fever, chills, sore throat, cervical adenopathy
- Glomerulonephritis is a complication

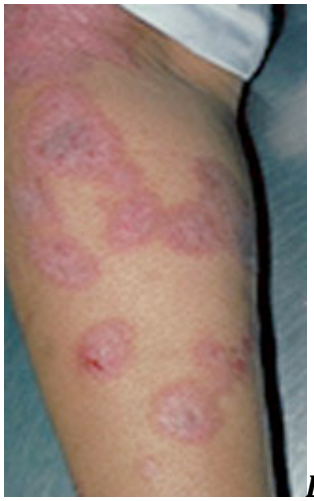
Treatment:

- Penicillin

Common Dermatologic Disorders

Eczema

- Is a superficial, itchy, erythematous lesion
- The rash develops after itching
- Commonly seen on the flexor surfaces
- Diagosis is clinical


Eczema

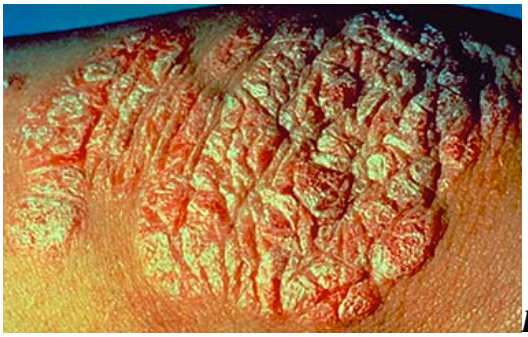
Treatment: Avoidance of irritants and triggers, keep skins moisturized, antihistamines/steroids are good for relief from inflammation and itching

There are many variations of Eczema, including:

1. ***Atopic Dermatitis*** – this is the classic where itching causes a rash
2. ***Contact dermatitis*** – an itchy rash at the site of contact, classically caused by contact with nickel, chemicals, or poison plants
3. ***Seborrheic dermatitis*** – scaling and flaking in areas of sebaceous glands

Psoriasis

- Presents as pink plaques with silver scaling
- Occurs on the extensor surfaces such as the elbows and knees
- There is often pitting of the fingernails
- The classic finding is known as "Auspitz Sign", which is pinpoint bleeding when the scale is removed
- This is diagnosed clinically

 ***Psoriatic Plaque***

Treatment:

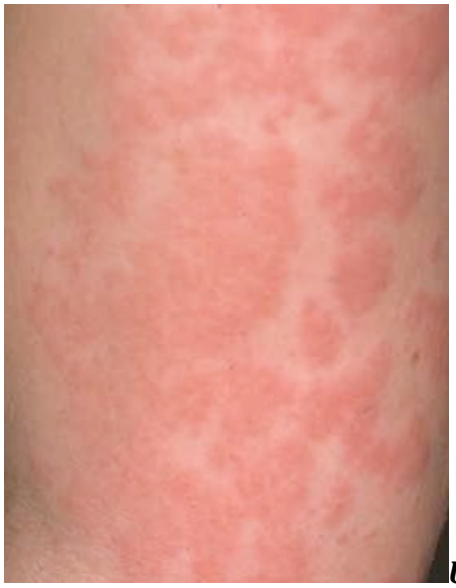
1st line is Topical steroids

2nd line is UVA light and may be used as an adjunct/prophylactic

3rd line is methotrexate and clyclosporin

Urticaria

- Is a condition caused by mast cell degranulation and histamine release
- Presents with the classic "wheals" that are intensely itchy
- "Dermographism" is seen where you can write a word with your finger on the skin and it will remain
- Most lesions are IgE-mediated (thus a type 1 sensitivity)
- Diagnose this condition by scrapings

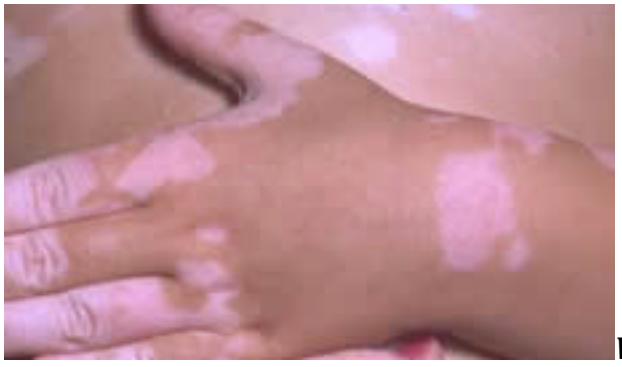
 ***Urticaria***

Treatment:

- Avoid triggers
- Give antihistamines/steroids
- This can possibly affect the respiratory tract which would then involve securing an airway

Vitiligo

- There is a loss of melanocytes in discrete areas of the skin
- Borders are sharply demarcated
- Usually seen in darker patients
- Possibly autoimmune in nature

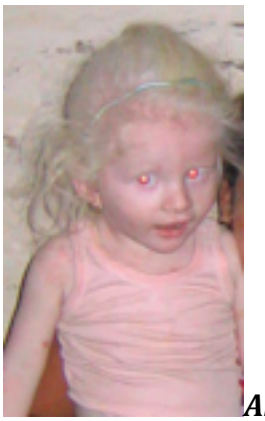
 ***Vitiligo***

Treatment:

- Mini-grafting can restore pigment to areas where it is lacking

Albinism

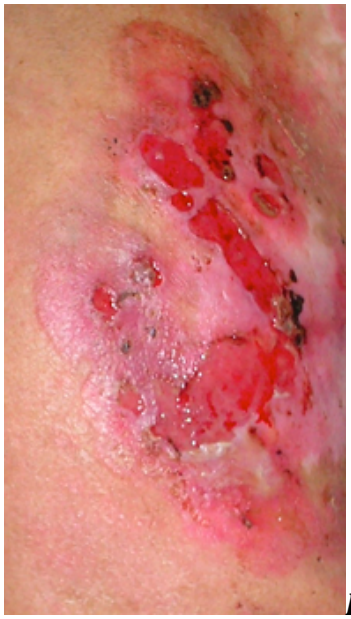
- Failure of melanocytes to produce pigment, due to a tyrosine deficiency
- Patient has white skin, iris translucency, decreased retinal pigment, nystagmus, and strabismus
- Avoid sun exposure and use sunscreen all the time
- There is an increased risk of skin cancer due to lack of protection from the sun

 ***Albinism***

Blistering Disorders

Pemphigus Vulgaris

- A rare autoimmune disorder affecting people between 20-40yr of age
- Bullae slough off easily and leave large denuded areas of skin, this is know as "Nikolsky's sign", this predisposes to an increased risk of infection
- Fatal condition if not managed properly



Pemphigus Vulgaris

Diagnosis:

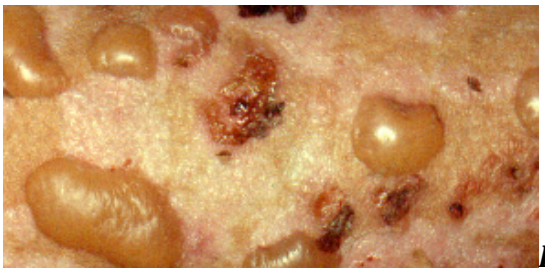
- Immunofluorescence of the surrounding epidermal cells shows a "tombstone" fluorescent pattern

Treatment:

- High-dose oral steroids

Bullous Pemphigoid

- An autoimmune condition that affects mostly the elderly
- Less severe than pemphigus vulgaris
- Presents with hard and tense bullae that do not rupture easily
- Prognosis is much better than that of pemphigus vulgaris

 ***Bullous Pemphigoid***

Diagnosis:

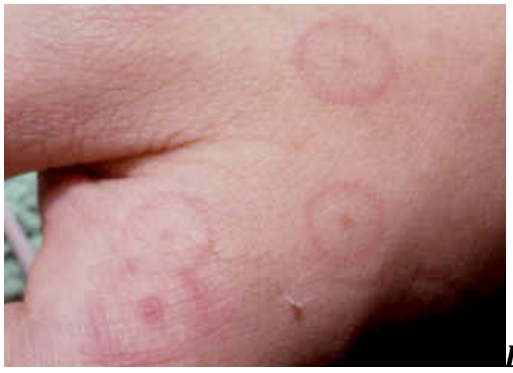
- Skin biopsy showing a linear band along the basement membrane on immunofluorescence
- Increased eosinophils found in the dermis

Treatment:

- Oral steroids

Erythema Multiforme

- Is a hypersensitivity reaction to drugs, infections, or systemic disorders
- Presents with diffuse and erythematous target lesions that are highly differing in shape

 ***Erythema Multiforme***

Diagnosis:

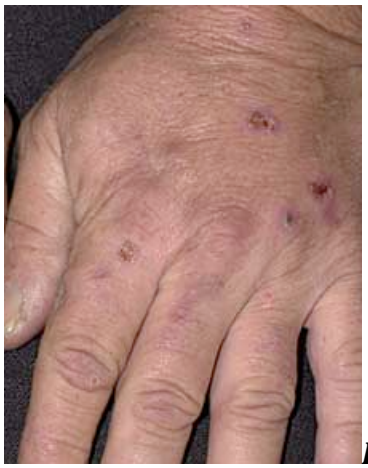
- Is clinical but a history of herpes infection makes this a likely diagnosis

Treatment:

- Stop offending causes and treat if there is a history of herpes with acyclovir

Porphyria Cutanea Tarda

- An autosomal disorder of impaired heme synthesis
- Get blisters on sun-exposed areas of the face and hands
- Differentiate this porphyria by the absence of abdominal pain

 ***Porphyria Cutanea Tarda***

Diagnosis:

- Wood's lamp of urine, where the urine fluoresces with an orange-pink color due to the increased levels of uroporphyrins

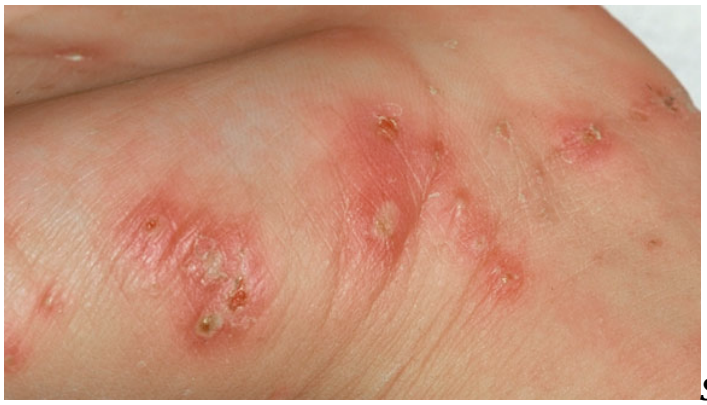
Treatment:

- Sunscreens used liberally
- Phlebotomy
- Chloroquine
- Avoidance of alcohol

Parasitic Infections of the Skin

Scabies

- Presents as severely itchy papules and burrows that are located along the webs of the fingers
- Highly contagious



Scabies infection

Diagnosis:

- Identify the Sarcoptes Scabiei mite from a skin scraping under the microscope

Treatment:

- Permethrin 5% create to the entire body for those infected and close contacts for 8-10hr, repeated in another week
- Wash all linens and bedding in hot water the same day as cream application

Pediculosis Capitis (Head Lice)

- Itching and swelling of the scalp
- Common in school-aged children

Diagnosis:

- Microscopic exam of the hair shaft will show lice attached to shaft

Treatment:

- Permethrin shampoo or gel to the scalp
- Repeat applications are commonly required

Pediculosis Pubis (Crabs)

- Extremely itchy papules in the pubic region
- May also be along the axilla, buttocks, eyelashes, eyebrows, and periumbilical area

Diagnosis:

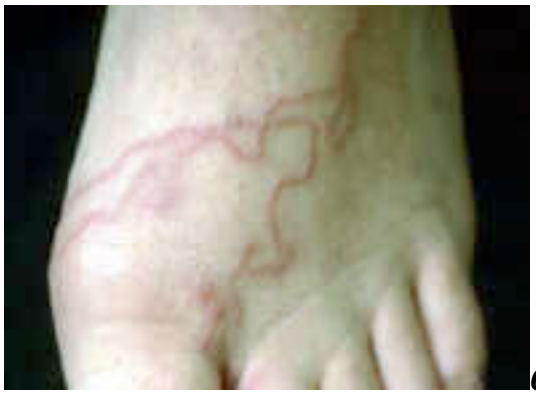
- Microscopic identification of lice

Treatment:

- Permethrin shampoo left on for 10 minutes, and repeated again within a week

Cutaneous Larva Migrans

- Is a snake-like, thread-like lesion that marks the burrow of the nematode larvae
- Often seen on the hands, back, feet, and buttocks
- Is caused by hookworms, namely → Ancylostoma, Necatar, and Strongyloides

 ***Cutaneous Larva Migrans***

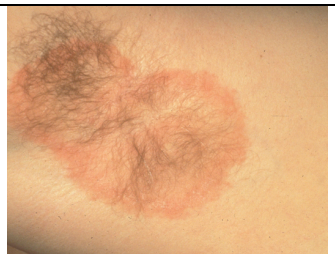
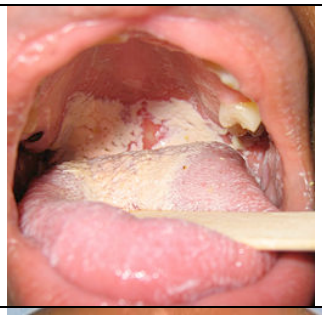
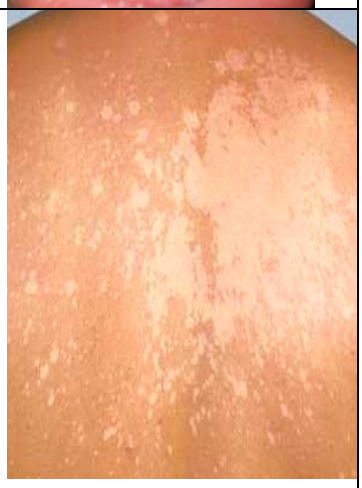
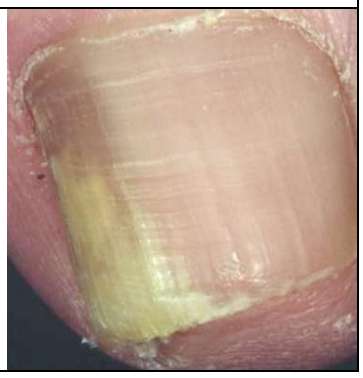
Diagnosis:

- A history of skin being exposed to moist soil or sand
- Presence of classic lesion

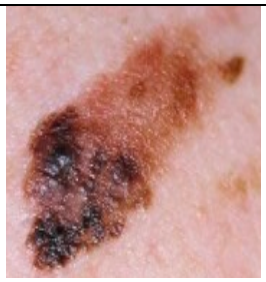
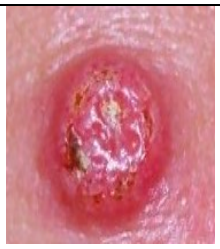
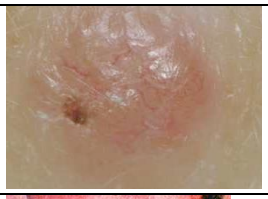
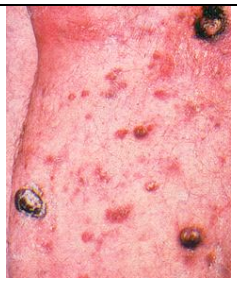
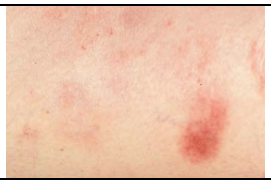
Treatment:

- Ivermectin orally or Thiabendazole topically

Fungal Cutaneous Disorders

Disease	Signs and Symptoms	Diagnosis	Treatment	
Tinea	Itchy, scaly, well demarcated plaques. Black dots seen on scalp	KOH Prep	Topical Antifungals	
Candida	Itchy, scaly plaques, usually in skin fold areas	KOH prep showing budding yeasts + pseudohyphae	Topical Nystatis or Oral Fluconazole	
Tinea Versicolor	Pityrosporum Ovale. Sharply demarcated hypopigmented macules on face and trunk in the summer. Macules do not tan	KOH prep shows the classic "Spaghetti and meatball appearance"	Selenium Sulfide shampoo on affected area for 7 days.	
Onychomycosis	Thickened, yellowing of the fingernails and toenails.	Clinical and/or KOH prep	Fluconazole or Itraconazole	

Skin Cancer

Cancer Type	Image	Signs and Symptoms	Treatment	Prognosis
Malignant Melanoma		Seen MC in light-skin people with increased sun exposure. Fits all ABCDE criteria	Excision + chemotherapy if there is metastasis	Poor with metastasis
Squamous Cells Carcinoma		Common in elderly. On sun exposed areas. Ulcerations and crusted	Excision + radiation	Moderate prognosis (better than melanoma but worse than basal cell)
Basal Cell Carcinoma		MC and looks like a pearly papule with translucent borders	Excision	Great prognosis – almost never metastasizes
Kaposi's Sarcoma		Red/purple plaques. Caused by HHV8. Almost exlusively in AIDS patient	Chemotherapy and HIV medications	Good unless there is associated organ damage
Cutaneous T-cell Lymphoma		Total body rash that is very itchy	Radiation, chemotherapy	7-10 yr survival with no treatment.

Chapter 19

High-Yield Preventative Medicine

- Female patients > 65 yrs of age should receive a one-time DEXA bone scan for osteoporosis
- Any female who is at least 60yr old and has 1 risk factor for osteoporosis
- The T-score is used to assess bone density
- A T-score between [-1.5 to -2.5] is considered to be osteopenia
- A T-score < -2.5 is osteoporosis
- A patient with a T-score of <-1.5 plus risk factors for osteoporosis (smoking, poor calcium vitamin D levels, lack of weight bearing exercise in the history, use of alcohol), should receive preventative medications, such as oral bisphosphonates or Raloxifene
- Raloxifene is the only SERM that is FDA approved to prevent osteoporosis
- Kids with cystic fibrosis should receive normal vaccinations in addition to a few additions → yearly influenza, pneumococcal boosters
- Patients with an egg allergy should avoid the influenza and yellow fever vaccines, and should be cautious with MMR as well
- The 1st step in management of increased LDL is lifestyle management, if LDL >100mg/dL and the patient has risk factors, they should be started on lifestyle modifications + statin drugs
- The drug of choice for increasing the HDL is Gemfibrozel and nicotinic acids
- It is recommended that any male who is an active smoker or former smoker and aged between 65-75 be given a one-time abdominal ultrasound to evaluate for a AAA
- Patients with a chronic liver disease should receive a number of vaccines (Tetanus every 10yrs + Hep A&B yearly, and pneumococcal vaccine)

Adult Vaccine Recommendations:

- Tetanus and Diptheria every 10yrs after 18yrs of age
- Influenza to all adults >50yr or to adults with chronic diseases (DM, CHF, etc)
- Pneumococcal vaccine given to all adults >65yr of age or to adults with chronic diseases

- Screening for cervical cancer with a pap smear should start at 21yr of age or 3yrs after the onset of sexual activity (whichever comes first)
- If 2-3 normal pap smears are done in a row and the woman is in a monogamous relationship, you can increase the time between pap smears to every 2-3 years.
- Screening can stop at 70yrs of age
- Bupropion is FDA approved for smoking cessation and must be used in conjunction with counseling and nicotine replacement

- Hepatitis A vaccine is given to men who have sex with men
- Meningococcal vaccine is given to those who live in close quarters with others
- Routine cholesterol screening should begin in patients at risk at 35yr of age in men and 45yr of age in women, then every 5yrs after that
- The most common vaccine-preventable disease is hepatitis A
- When CD4 count drops below 200/250, start PCP prophylaxis with TMP-SMX
- When CD4 count drops below 50, prophylax for mycobacterium avium complex with amoxicillin
- Routine screening for Chlamydia is now recommended for all sexually active females who are ≤ 24 yrs of age
- Mammograms should be done every 1-2 yrs starting at 40yr in females with an average risk of breast cancer. There is no clear time when they should stop, but it should be no sooner than 70yr of age
- Pneumococcal vaccine is a capsular polysaccharide of the 23 most common types of pneumococcus, which yield a B-cell response only, it is T-cell independent
- Any female with a history of being treated for CIN2/3 should have pap smears with or without colposcopy and curettage every 6 months until three negative results are obtained (patients may resume standard screen after meeting this milestone)