Study Questions

Bone Pathologies:

* **Bisphosphonate-Associated Osteonecrosis:**
	+ BON is associated with **2nd generation bisphosphonates** because 2nd gen bisphophonates have **very long half lives** (i.e. half life of fosamax is 12 years) and get incorporated into the skeleton
	+ Factors that increase risk of BON in the mandible include
		- Dental surgery, dental infections/disease, oral trauama, periodontitis
		- Concomitant **chemotherapy** or **corticosteroids**
		- **Age >65 years**
		- Presence of bony tori/exotoses
		- Diabetes, smoking/alcohol use, poor oral hygiene
		- Duration of Rx use for **>3 years**
	+ Etiology of BON:
		- Bisphosphonates decrease **osteoclast activity**, which normally includes the repair of microfractures and the resorption of foci with nonviable osteocytes, allowing the **accumulation of microfractures** and the persistence of **osteocytes past their 150 day lifespan**
	+ Management of BON includes **conservative** management where the goal of therapy is to **stop pain** and **infection** (if present)
		- **Asymptomatic** patients:
			* **Chlorhexidine** rinse daily
			* Smooth edges of rough bone
		- **Symptomatic** patients:
			* Treat with **chlorhexidine** and **antibiotics**
				+ Amoxicillin +/- metronidazole
				+ Clindamycin, azithromycin
* **4 genetic diseases of bone**:
	+ **Osteogenesis imperfect** – **AD** (**type I**) or **AR** mutation of **chromosome 7** and **17**
	+ **Cleidocranial dysplasia – AD** mutation of **chromosome 6**
	+ **Osteoporosis** (infantile and adult)
	+ **Cherubism**
	+ **Note**: **Gardner’s syndrome** is an **autosomal dominant** genetic mutation of **chromosome 5** that clinically produces **osteomas**
* **Osteogenesis imperfect**
	+ OI is a disorder of **collagen maturation**
	+ All 4 types of OI produce the following **craniofacial,** **teeth** and **ocular findings**
		- **Class III malocclusion** (due to **maxillary hypoplasia**)
		- (sometimes) **florid osseous dysplasia-like bone changes**
		- Clinical and radiographic evidence of **dentinogenesis imperfect** in primary and permanent teeth
			* Produces opalescent teeth
		- Classic ocular presentation of OI includes **blue sclera**
* **Cleidocranial dysplasia**
	+ CCD is a condition that affects **osteoblastic differentiation**, affecting primarily membranous bones (clavicle and skull)
	+ Clinical features of CCD include:
		- **Clavicles**:
			* **Hypoplastic** or **aplastic**
			* **Unilateral** or **bilateral**
			* Increased **shoulder mobility**
		- **Skull bones**:
			* **Sutures** and **fontanels** remain open
			* **Wormian** bones (extra sutural bones) develop in suture lines
			* **Frontal** and **parietal** bone bossing
			* Produces large heads
			* **Hypertelorism** (increased length between 2 organs), **high palate**, **depressed nasal bridge**, **absent maxillary sinus**
		- **Teeth**:
			* many **unerupted** permanent and supernumerary teeth
			* often **misshapen** teeth
			* **retention of primary teeth** into adulthood
			* **absent cementum**
* **Osteopetrosis:**
	+ A condition that affects **osteoclasts**, preventing their function and producing **osteosclerosis**
* **Paget’s Disease of the Bone**
	+ PDB is a condition characterized by increased bone turnover/metabolism, affecting **white men** (esp. England) **over age of 40**
	+ Clinically characterized by:
		- **Pain**, especially near joints
		- **Bowing** of long bones
		- **Enlarged** skull
		- Usually **polystotic**
	+ **17%** of patients experience **jaw involvement** (primarily the body and alveolus) where the **maxilla** becomes **larger** than the **mandible**, there can be **sinus obstruction,** and there can be **bone expansion**
	+ Radiographic findings:
		- **Early** phase: often **osteoclastic**
			* **Decreased bone density** and **increased radiolucency**
		- **Late** phase: **osteoblastic** stages with **patchy sclerosis** becoming **confluent** (“**cotton wool**”)
	+ Histologically, the **active phase** presents with **alternating** phases of **resorption/osteoclastic activity** and **formative/osteoblastic activity** on the same bone trabeculae and very **vascular fibrous CT** replacing bone marrow
	+ Diagnosis involves:
		- **Blood** test: **increased alkaline phosphatase** (a marker of osteoblastic activity) with **normal** calcium and phosphorous
		- **Urine** test: **increased hydroxyproline** and other markers of **bone resorption**
	+ **Osteosarcomas** can develop (1-13% of patients), usually in the lower extremities and pelvis
* **Idiopathic osteosclerosis**
	+ A **common** condition (5% of population) that affects **teens and young adults** (20’s)
	+ Usually an incidental finding; **asymptomatic** with **no bone expansion**
	+ Radiographically presents as a **well-defined**, **rounded** or **triangular radiodensity** that is **uniformily opaque** with **no lucent portions**
	+ **No treatment**
* **Focal osteoporotic bone marrow defect**:
	+ A condition which affects **women (3:1)** and is often found in the **posterior mandible** (70%)
	+ It is **asymptomatic** and does **not expand bone**
	+ Radiographically presents as a **radiolucency** with **ill-defined borders** and **fine trabeculations**
	+ **No treatment**
* **Langerhan’s Cell Histiocytosis**
	+ A condition with the monoclonal proliferation of **histiocyte-like cells (Langerhan’s Cells)** with polyclonal infiltration of **MNGC**, **eosinophils,** and other **chronic inflammatory cells**
	+ 3 forms of LCH:
		- **Eosinophilic granuloma of bone**: can be monostotic or polystotic
		- **Chronic disseminated histiocytosis/Hand-Schuller-Christian disease**:
			* Affects **bone, skin and viscera**
			* Presents as a **triad** of symptoms:
				+ **Bone lesions**
				+ **Exophtalamos**
				+ **Diabetes insipidus**
		- **Acute disseminated histiocytosis/Letterer-Siwe disease**
			* Prominent **visceral, bone marrow, and skin** involvement
			* Usually in **infants**
	+ LCH is a **childhood** disease, with **50%** of patients being **<10 years old**
	+ **Jaw** lesions develop in **10-20%** of cases, usually in patients **>20 years old**
	+ Radiographic presentations:
		- **Well-defined, non-corticated, punched out radiolucencies**
		- In jaws, is sometimes **ill-defined** due to **destruction of alveolar bone** from the crest
			* Mimics severe periodontal disease
	+ Histologically, LCH is diagnosed when samples present as:
		- **CD1a** - positive
		- **S100** - positive
		- **Peanut agglutinin** – positive
		- **Birbeck granules** – positive
* **Central Giant Cell Granuloma**
	+ Clinically presents in:
		- **Wide** age range, though **60%** of patients are **<30 years old**
		- Presents more often in **women**
		- Usually located in the **mandible**, **anterior to 1st permanent molar**
		- **Asymptomatic** and grows **slowly**, though the more aggressive course of the disease causes rapid growth, bone expansion, pain, root resorption, parasthesia, and perforation of the cortical plate
	+ Radiographically presents as a **unilocular or multilocular radiolucency** with **well-defined**, **possibly corticated** margins
	+ Histologically, CGCG presents with **MNGC**’s, which are mostly likely **osteoclasts**
	+ **Normal** lesions are treated with surgery
		- **Curettage** leads to **20% recurrence rate**, but responds well
	+ **Aggressive** lesions are treated with **intralesional** or **systemic chemotherapy** with **corticosteroids or calcitonin** (respectively)
* **Cherubism**:
	+ A condition that begins to present in patients between **ages 2-5** and **stabilizes at puberty**
	+ Clinically present with an **expanded mandible** and **orbital bone**, which produces a “cherubic” facial appearance
	+ Radiographically, the lesion presents as **bilateral, multifocal, multilocular radiolucencies**
	+ Diagnosis is made utilizing information gained from **radiography**, **family history**, and **biopsy**
		- need to **rule out** **hyperparathyroidism** and  **CGCG** (which are found in this disease’s histology)
* **Traumatic Bone Cyst**:
	+ AKA:
		- **simple bone cyst**
		- **solitary bone cyst**
		- **idiopathic bone cyst**
	+ Not a true cyst, but is rather an **empty or fluid-filled bone cavity**
	+ TBC’s are reported in **almost all bones** of the body
	+ **Jaw** lesions develop in **10-20 year olds** (**rare** to find in patients **>35 years old**) with a slight **male predilection**
		- Radiographically presents as a **unilocular radiolucency** with **variable definition** and **scallops between roots** (without devitalizing or resorbing them)
	+ **Rare** for TBC’s to produce **pain, parasthesia or bone expansion**
	+ **Surgical exploration** of the cavity often causes repair of the lesion
* **Aneurysmal Bone Cyst:**
	+ An **intrabony accumulation of blood** with growth of **reactive fibrous CT and bone**
	+ ABC’s are more common in **long bones** and **vertebrate**, but presents in the **posterior mandible** of **children and young adults**
	+ ABC’s cause **pain, rapid swelling, ballooning expansion of bone**, but **rarely parasthesis, tooth mobility or root resorption**
	+ Treat by **surgically curetting** out the “**blood-soaked sponge**” lesion
		- The defect usually heals within **6 months**
		- **Cryosurgery** may be needed
* **Benign Fibro-osseous Lesions of the Jaw**:
	+ Contains 3 categories of diagnoses:
		- **Fibrous dysplasia**
		- **Cemento-osseous dysplasia**
		- **Ossifying fibroma**
	+ **Fibrous dysplasia**:
		- Clinically appears as **tumorous swellings of bone(s) and skin with endocrine abnormalities**
		- Usually diagnosed in **teenage years**
	+ **Cemento-osseous dysplasia** (in jaws only)
		- 3 types:
			* **Periapical** COD
			* **Focal** COD
			* **Florid** COD
	+ Radiographic presentations:
		- **FD**:
			* **Poorly defined uniform radioppacity with “ground glass” trabeculae appearance**
		- **CODs:**
			* **PA-COD**: **radiolucency** at the apex of **vital** teeth; identical looking to a periapical granuloma/cyst
			* **Florid COD**: adjacent **radiolucencies** fuse to form a **continuous lucency at the apex of several teeth**
			* **Mature** lesions: **mixed** **radiolucent rim with radioopaque center**; no fusion of teeth
		- **OF:**
			* **Early** lesions: **well-defined unilocular radiolucency** or a **mixed lesion with varying radioopacity**; some have **sclerotic borders**
			* **Later** lesions: can become **large**, causing **root divergence** and **bowing of inferior border of mandible**
	+ **Fibrous dysplasias** and **ossifying fibromas** can cause **bone expansion**, but **all COD’s** do **not**
	+ Locations of lesions:
		- **FD: mandible** is the most common location
			* Usually **below** the **inferior alveolar canal**, thus displacing it superiorly during bone expansion
			* If found on the **maxilla**, it is found **below** the **maxillary sinus**, thus displacing it upward to possibly obliterate it
		- **CODs:** only occur on the **jaws**, in **tooth-bearing areas**
		- **OF:** most common in the **posterior mandible**
	+ Histologically:
		- FD:
			* A haphazard mix of **immature/woven bone** in a **fibrous CT stroma**
			* “**Chinese-character**” trabeculae
		- COD:
			* **Fibrous CT**, **metaplastic immature/woven bone** with **cementum-like droplets**
		- OF:
			* Very similar, if not identical to CODs
	+ Etiology:
		- FD: Found in **teens**
		- COD:
			* PA-COD: found in **40 year old African-American women**
			* Florid COD: found in **50 year old AA females**
			* Focal COD:found in **40 year old African-American women**
		- OF: Found in **females** in their **20’s-30’s**
	+ Treatments:
		- FD: **Comestic shave down** once lesion is **mature**
		- COD: no treatment; surgery can cause **osteomyelitis**
		- OF: **enucleation**
* **Osteomas:**
	+ Clinically presents as a **periosteal (on bone)** or an **endosteal (in bone)** lesion
	+ Radiographically presents as **well-defined radiolucencies** either **in** or **on bone (endosteal, periosteal,** respectively)
* **Gardner Syndrome**:
	+ Oral presentation includes:
		- **osteomas** in any part of the **mandible,** but most often at the **angle of the mandible**
		- **odontomas**
		- **supernumerary teeth**
		- **impacted teeth**
	+ Oral manifestations of GS is **not** the most important determinant in the patient’s overall health because GS is primarily a disease of **familial colorectal polyposis**
* **Osteoblastoma**:
	+ A **rare benign bone neoplasm** that is **very painful**
	+ Radiographically presents as a **mixed** lesion with **varying amounts of patchy opacity**
		- >**2cm** large
* **Cementoblastoma**:
	+ A **neoplasm of cementum** that causes **pain and swelling** (66% of patients) in the **posterior mandible** of patients **<30 years old**
	+ Radiographically presents with the lesion **fused to the roots** with a **radiolucent rim** at the periphery
* **Hemangioma of the Bone**
	+ Usually found in patients that are **10-20 years old**, though it can occur in adults as well
	+ If the lesion is symptomatic, it will present as **pain, swelling and spontaneous bleeding from the sulcus**, which can mimic severe periodontal disease/abscess
* **Osteosarcoma**:
	+ The most common **primary malignancy of bone** in patients **<40 years old**
	+ Radiographic appearance, unique to osteosarcomas:
		- **Spiking root resorption**
		- **Sunburst periosteal reaction** (to cortical infiltration)
		- **Symmetric widening of PDL**
	+ Histologic appearance:
		- **Pleomorphic round or spindle cells** that are producing **bone, fibrous CT, or chondroid**
	+ Treatment includes **radical resection** and possible pre- and post-op chemotherapy
		- **30-50%** survival rate
* **Chondrosarcoma**:
	+ **Half** as common as an osteosarcoma
	+ Radiographic appearance:
		- Much like an osteosarcoma:
			* **Spiking root resorption**
			* **Sunburst periosteal reaction**
			* **Symmetric widening of PDL**
		- However, the lesions **vary in density** and have **poorly-defined borders**
	+ Histologically, lesions have varying degrees of **cartilage** and **cellular atypia/pleomorphism**
	+ Treatment includes **radical resection**, with chemotherapy as a last resort for unresectable tumors
		- Survival data is unreliable due to possible **late recurrences** (**>15 years** after surgery)
* **Metastatic Malignancies of the Jaws**:
	+ The most common carcinomas to metastasize to the mandible are:
		- **Breast**
		- **Lung**
		- **Thyroid**
		- **Prostate**
		- **Kidney**
	+ Most malignancies are most often **ill-defined radiolucencies**, except **breast and prostate malignancies**, which can present with **mixed lucent/opaque** lesions
* **Ewings Sarcoma**:
	+ ES most often occurs in **white** **teenagers**
	+ Clinically, ES can occur in **any bone**, including the **mandible** (which is **uncommon)**
		- Symptoms include **parasthesia, loose teeth, pain, swelling, soft tissue breakthrough, fever, elevated WBC count**
	+ Radiographically, ES presents with **ill-defined radiolucencies**, with rare **onion skin periosteal reactions**
	+ Treatment for ES includes **surgery and chemotheraphy** with possible radiation therapy
	+ **40-80% survival rate**