ORAL PATHOLOGY

Legend			
Major Topic	**Abbreviation**	**Major Topic**	**Abbreviation**
Abnormalities of Teeth	Abn of Tth	Odontogenic Cysts	Odont Cyst
Connective Tissue Lesions	Con Tis Les	Odontogenic Tumors	Odont Tum
Diseases of the Blood	Bld Dis	Pigmented Lesions of the Oral Cavity	Pig Les of Oral Cav
Inflammatory Jaw Lesions	Infl Jaw Les	Pseudocyst	Pseudocyst
Metabolic & Genetic Jaw Diseases	M & G Jaw Dis	Red-Blue Lesions	R-B Les
Miscellaneous	Misc.	Salivary Gland Tumors	SG Tum
Neoplasms	Neo	Terms	Terms
Nerve & Muscle Disorders	Ner & Mus Disord	Ulcerative-Conditions	Ulc Cond
		Verrucal-Papillary Lesions	V-P Les
Non-Odontogenic Cysts	N-O Cysts	Vesiculo-Bullous Diseases	V-B Dis
Non-Odontogenic Tumors	N-O Tum	White Lesions	W Les

A developmental abnormality characterized by the **total absence** of teeth is called:

- Hypodontia
- Anodontia
- Diphyodontia
- Hypsodontia

• **Anodontia**

Two forms of anodontia:

1. **Complete true** → is a rare condition in which **all of the teeth** are missing. It may involve both the primary and permanent dentitions. It is usually associated with **hereditary ectodermal dysplasia.**

2. **Partial anodontia** *(commonly referred to as* **congentially missing teeth***)*→ is rather common. Teeth usually affected include the third molars *(maxillary more often than mandibular),* maxillary lateral incisors and mandibular second premolars. **Note:** As a general rule, if only one or a few teeth are missing, the absent tooth will be the **most distal tooth** of any given type *(if molar, then it would be the third molar).*

Other terms to be familiar with include:

• **Oligodontia** → refers to the congenital absence of many, **but not all,** teeth.

• **Hypodontia** → refers to the absence of **only a few** teeth.

All of the following statements concerning **dentinogenesis imperfecta** are true, **except:**

- It is an inherited disorder of the dentin
- It is also known as hereditary opalescent dentin
- It affects only the teeth
- It is an extremely common disorder
- It is sometimes linked to osteogenesis imperfecta
- The presence of blue sclera or a history of bone fractures are signs of osteogenesis imperfecta

- **It is an extremely common disorder**

***This is false;** it is a **rare disorder** found in about 1:7,000 children.

Teeth affected with dentinogenesis imperfecta have an amber, gray, or purple **opalescence** *(translucence)* or discoloration. The pulp chamber may be **completely obliterated.** This is due to the continued deposition of dentin. The crowns are generally short and bulbous in appearance, while the roots are narrow. The enamel can chip away within 2-4 years following eruption. This exposes the dentin underneath, which is soft and wears rapidly. **Note:** These effects can be seen in both the deciduous and permanent dentition. **See picture #4 in booklet.**

Three types of dentinogenesis imperfecta:

- **Type I** → dentin abnormality occurs in patients that have **osteogenesis imperfecta** *(characterized by blue sclera or a history of bone fractures).*
- **Type II** → most common, only the dentin abnormality exists with no bone involvement.
- **Type III** *(Brandywine Type)* → like Type II, only the dentin abnormality exists **however** there are clinical and radiographic variations in this type. They include multiple pulp exposures in the deciduous dentition.

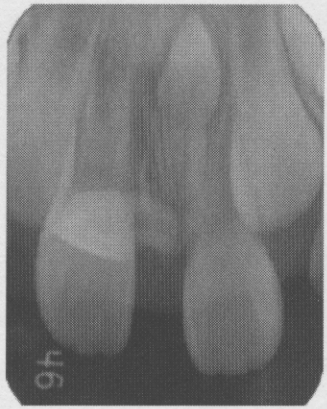
Important: Clinically, dentinogenesis imperfecta is usually easily detected and identified. The teeth exhibit a **translucent or opalescent appearance**. **See picture #5 in booklet.** Another clinical feature is the abnormal constriction at the enamel-cementum junction that is detected by exploration. **Note:** The enamel in these teeth is structurally and chemically normal.

ORAL PATHOLOGY

Abn of Tth

The permanent maxillary centrals in the x-ray below are **vital.** What is the most probable interpretation of the condition illustrated?

- Mesiodens
- Concrescence
- Fusion
- Dens in Dente



- **Mesiodens**

Mesiodens is the **most common supernumerary** tooth, appearing singly or in pairs as a small tooth with a cone-shaped crown and a short root between the maxillary central incisors; it may be erupted, impacted or even inverted.

Mesiodens appear situated in the maxilla near the midline and almost always posterior to the normal central incisors. Many of them, therefore, are bypassed by the permanent incisors which are permitted to erupt into their normal position in the arch.

Remember:

- **Fusion** → is a developmental union of two or more teeth in which the dentin and one other dental tissue are united *(may be the root)*. **See picture #1 in booklet.**
- **Concrescence** → is a condition in which only the cementum of two or more teeth becomes united. **See picture #2 in booklet.**
- **Dens in dente** *(also called dens invaginatus)* → means **"tooth within a tooth,"** it is caused by a deep invagination of the enamel organ during formation, most likely found associated with a maxillary lateral incisor. **See picture #3 in booklet.**

Excessive formation of **cementum** on the surface of the root is referred to as:

- Cemental pearls
- Hypercementosis
- Condensing osteitis
- Secondary cementum

• **Hypercementosis**

Hypercementosis is often confined to the apical half of the root but, in some instances, may involve the entire root. In large majority of instances, it affects **vital** teeth, is not associated with any one particular systemic disease and may be regarded as a dental anomaly. It may be seen when a tooth has lost its antagonist or when there is chronic inflammation of the tooth. The **premolars** are most frequently involved. Next in frequency are the first and second molars. **See picture #39 in booklet.**

Hypercementosis produces no **significant clinical signs or symptoms** indicative of its presence. It is seen radiographically as a bulbous enlargement that has surrounding it a **continuous and unbroken periodontal membrane space** and a normal lamina dura.

There is a form of hypercementosis which is a common feature in **Paget's disease** that involves the jaws. On the x-ray in this case, there is **complete absence** of the periodontal membrane space and lamina dura surrounding the hyperplastic cementum.

Note: Hypercementosis is also seen in acromegaly.

An enamel defect resulting from the **incomplete formation** of the enamel matrix is called:

- Enamel pearls
- Hypercementosis
- Enamel hypoplasia
- Dentinal dysplasia

• **Enamel hypoplasia**

Enamel hypoplasia is a developmental defect in which the enamel of the teeth is hard in context but thin and deficient in amount. It results from **incomplete formation of the enamel matrix with a deficiency** in the cementing substance. Enamel hypoplasia affects both the deciduous and permanent teeth. It is usually due to illness or injury during tooth formation or due to a genetic disorder. **Note:** The genetic forms of enamel hypoplasia are generally considered to be types of amelogenesis imperfecta.

The clinical appearance of enamel hypoplasia includes **1)** the lack of contact between teeth **2)** the rapid breakdown of occlusal surfaces **3)** a yellowish-brown stain that appears where the dentin is exposed. **Note:** If only one permanent tooth is affected, it is usually caused by physical damage to the primary tooth that this permanent tooth replaced.

Remember: Enamel **hypocalcification** is a hereditary dental defect in which the enamel is soft and undercalcified in context yet **normal in quantity**. It is caused by the defective maturation of ameloblasts *(there is a defect in the* **mineralization** *of the formed matrix).* The teeth are chalky in consistency, the surfaces wear down rapidly and a yellow to brown stain appears as the underlying dentin is exposed. This condition affects both the deciduous and permanent teeth as well.

The **fusion** of the **alveolar bone** to a tooth is known as:

- Hypercementosis
- Gomphosis
- Ankylosis
- Gemination

- **Ankylosis**

Ankylosis may be initiated by an infection or trauma to the periodontal ligament. The ankylosed tooth has lost its periodontal ligament space and is truly fused to the alveolar process or bone. **Note:** There is a change in the continuity of the occlusal plane which is caused by the continued eruption of non-ankylosed teeth and the growth of the alveolar process.

Remember:

- **Gemination** *(also called twinning)* is a division of a single tooth germ by invagination. This results in the incomplete formation of two teeth. Frequently occurs in the incisor region.

- **Hypercementosis** is the excessive formation of cementum around the root after the tooth has erupted. It may be caused by trauma, metabolic dysfunction or periapical inflammation.

- **Gomphosis** is a type of fibrous joint in which a conical process is inserted into a socket-like portion, such as the styloid process in the temporal bone or the teeth in the dental alveoli.

All of the following statements concerning **amelogenesis imperfecta** are true, **except:**

- It is an inherited condition which is transmitted as a dominant trait
- It only affects the permanent teeth
- It causes the enamel of the teeth to be soft and thin
- The teeth appear yellow, because the dentin is visible through the thin enamel
- The teeth are easily damaged and susceptible to decay

• **It only affects the permanent teeth**

***This is false;** amelogenesis imperfecta **affects all teeth,** both deciduous and permanent

Amelogenesis imperfecta is a hereditary **ectodermal** defect, unlike dentinogenesis imperfecta which is a hereditary mesodermal defect. There are three types of amelogenesis imperfecta: hypoplastic, hypocalcified and hypomaturation.

The crowns of the teeth may or may not show discoloration. If present, it varies depending upon the type of the disorder, ranging from **yellow to dark brown**. Contact points between teeth are often open and occlusal surfaces and incisal edges frequently are severely abraded.

The radiographic findings are frequently distinctive and **pathognomonic. See picture #63 in booklet.** When the enamel is totally absent, the **radiographic appearance makes the diagnosis obvious.** When some enamel is present, thin radiopaque coverings on the proximal surfaces of the teeth are noted. When the anatomic crown forms are normal or nearly normal, the softness of the defective enamel may not be easily distinguished from the dentin. **In all cases,** however, the dentin, pulp and cementum are unaffected by the disease process itself *(unlike dentinogenesis imperfecta).*

Exception: Amelogenesis imperfecta will only show pulp obliteration if there is advanced abrasion with secondary dentin formation.

The **abnormal** or **pathological** wearing away of tooth substance is referred to as:

- Erosion
- Attrition
- Abrasion

- **Abrasion**

Types of abrasion:

1. **Toothbrush** abrasion → most often results in V-shaped wedges at the cervical margin in the **canine and premolar areas.** It is caused by the use of a hard toothbrush and / or a horizontal brushing stroke and/or a gritty dentifrice.
2. **Occlusal** abrasion → results in flattened cusps on all posterior teeth and worn incisal edges. It results from the chewing or biting of hard foods or objects and chewing tobacco.

Remember:

- **Attrition** is the wearing away of enamel and dentin due to the **normal function** or most commonly, due to the excessive grinding or gritting together of teeth by the patient *(referred to as* **bruxism***)*. The most noticeable effects of attrition are **polished facets,** flat incisal edges, discolored surfaces of the teeth and exposed dentin. Facets usually develop on the **linguoincisal** of the maxillary central incisors, the **facioincisal** of the mandibular canines and the **linguoincisal** of the maxillary canines.

- **Erosion** is the loss of tooth structure from **non-mechanical means.** It can result from drinking acidic liquids or eating acidic foods. It is common in **bulimic** individuals as a result of regurgitated stomach acids. It affects smooth and occlusal surfaces.

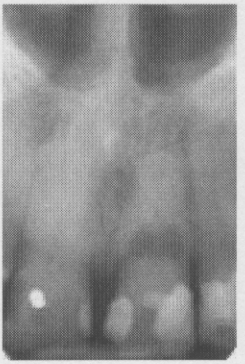
All of the following can cause **intrinsic staining** of teeth, **except:**

- Dentinogenesis imperfecta
- Erythroblastosis fetalis
- Porphyria
- Fluorosis
- Diabetes mellitus
- Pulpal injury
- Internal resorption
- Tetracyclines

- **Diabetes mellitus**

Cause	Intrinsic stain
Dentinogenesis imperfecta	Translucent or opalescent hue, usually gray to bluish-brown
Erythroblastosis fetalis	Bluish-black, greenish-blue, tan or brown
Porphyria	Red or brownish
Fluorosis	White opacities or light brown to brownish-black
Pulpal injury	Starts pink and usually becomes orange-brown to bluish-black
Internal resorption	Pinkish
Tetracycline	May vary from light gray, yellow or tan to darker shades of gray

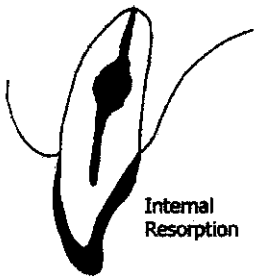
What is the **most probable diagnosis** of the x-ray below?



• **Internal resorption**

Internal resorption is an unusual form of tooth resorption that begins centrally within the tooth. Resorption of the dentin of the pulpal walls may be seen as part of an inflammatory response to pulpal injury, or it may be seen in cases in which no apparent trigger can be identified. Most cases of internal resorption present no early clinical symptoms. The **first evidence of the lesion** may be the appearance of a pink-hued area on the crown of the tooth, which represents the hyperplastic, vascular pulp tissue filling the resorbed areas.

If the condition is discovered before perforation of the crown or root has occurred, **endodontic therapy** may be carried out with the expectation of a pretty high success rate.



Which type of **amelogenesis imperfecta** is characterized by having enamel so soft that it can be removed during a prophylaxis?

- Hypoplastic type
- Hypocalcified type
- Hypomaturation type

• **Hypocalcified type**

There are **three types** of amelogenesis imperfecta:

1. **Hypoplastic** type → the enamel **has not formed** to full normal thickness *(or in some cases may be completely absent)* on newly erupted developing teeth. It results from the **defective formation of the enamel matrix.**
2. **Hypocalcified** type → the **quantity of enamel is normal** but the enamel is so soft that it can be removed during a prophylaxis. It results from the **defective mineralization of the enamel matrix.**
3. **Hypomaturation** type → the enamel can be pierced by an explorer tip under firm pressure and can be chipped away from the normal-appearing dentin. It is characterized by **immature crystallites.**

Remember:

1. Amelogenesis imperfecta is a hereditary **ectodermal** defect, unlike dentinogenesis imperfecta which is a hereditary **mesodermal** defect.

2. In all three types of amelogenesis imperfecta, the dentin, pulp and cementum are **unaffected** by the disease process itself *(unlike dentinogenesis imperfecta).*

Which of the following are characteristics of **dentin dysplasia:**

- Normal enamel
- Atypical dentin
- Pulpal obliteration
- Defective root formation
- A tendency towards multiple periapical radiolucencies and early exfoliation of teeth
- All of the above

- **All of the above**

There are two types of dentin dysplasia *(which is also called* **rootless teeth***)*. Type I and Type II. This appears to be a hereditary disease, transmitted as an autosomal dominant characteristic. This condition has not been associated with any systemic connective tissue disorder.

Type I (Radicular)	Type II (Coronal)
• **More common type** • Both dentitions involved • Normal morphology and color *(deciduous and permanent)* • Mobile teeth • Premature exfoliation • Short roots *(rootless teeth)* • Obliterated pulp chambers *(deciduous)* • Crescent-shaped pulpal remnant *(permanent)* • Periapical radiolucencies • Coronal dentin okay • Root dentin disoriented	• Both dentitions involved • Bluish-gray opalescent appearance *(deciduous)* • Normal clinical appearance *(permanent)* • Obliterated pulp chambers *(deciduous)* • Thistle tube pulp chambers and stones *(permanent)* • Normal coronal dentin in both dentitions • Amorphous and atubular dentin in radicular portion of deciduous teeth • Pulp stones or true denticles in permanent teeth

The **etiology** of leukemia is:

- Viral
- Bacterial
- Fungal
- Unknown

- **Unknown**

However, the following agents are believed to be closely **associated** with the development of leukemia:

- **Ionizing** radiation → increased incidence of leukemia among atomic bomb survivors and radiologists – usually **myelogenous**.
- **Viruses** → shown to cause leukemia in fowl and rodents. Herpes-like viral particles have been cultured from patients with various types of leukemia and leukemic patients have high antibody titer to the **Epstein-Barr Virus**.
- **Genetic Mutations** → **Philadelphia chromosome** *(translocation of chromosomal material from chromosome 22 to chromosome 9)* is present in 90% of patients with chronic **myelogenous leukemia;** also, higher incidence of acute leukemia in patients with **Down syndrome** *(mongolism)* in which there is trisomy 21.
- **Other** → chronic exposure to benzol, aniline dyes and related chemicals has been associated with the development of leukemia.

Remember:

- Leukemia is classified by the dominant cell type and by duration from onset to death

- Leukemia can **modify** the inflammatory reaction

All of the following statements concerning **Plummer-Vinson syndrome** are true, **except:**

- It is a rare disorder associated with severe and chronic iron-deficiency anemia
- It occurs chiefly in adolescent teenagers
- Systemic symptoms include weakness, pallor, difficulty swallowing and difficulty in breathing
- Oral symptoms include angular stomatitis and a smooth, red, painful tongue with atrophy of the papillae

• **It occurs chiefly in adolescent teenagers**

***__This is false;__ Plummer-Vinson syndrome occurs **chiefly in women in their thirties and forties.**

Because of the predisposition to the development of carcinoma of the oral mucous membranes, it is essential that the diagnosis be established early so that treatment can be given ASAP. This includes administration of iron, vitamin B complex and a high protein diet. **Note:** The dysphagia *(difficulty swallowing)* results from an esophageal stricture or web.

Aplastic anemia is a form of anemia in which the capacity of the bone marrow to generate red blood cells is defective. **Two types:**

1. **Primary** → unknown cause, affects young adults. The signs and symptoms include pallor, weakness, malaise, dyspnea *(difficulty breathing)*, headache and vertigo. Oral symptoms include spontaneous bleeding, bruising *(petechiae)* and gingival infections. **It is usually fatal.**
2. **Secondary** → caused by exposure to toxic agents, such as radiation, chemicals or drugs *(for example,* **chloramphenicol***)*. It can occur at any age. Symptoms are the same as primary. Prognosis is good once you remove the cause.

***Aplastic anemia is the **most serious** and **life-threatening blood dyscrasia** associated with drug toxicity.

A chronic, usually fatal, inherited form of anemia marked by **crescent-shaped red blood cells** and characterized by fever, leg ulcers, jaundice and episodic pain in the joints is called:

- Aplastic anemia
- Thalassemia
- Sickle-cell anemia
- Pernicious anemia

• **Sickle-cell anemia**

Sickle-cell anemia *(also called sickle-cell disease)* is the result of the production of **abnormal hemoglobin** *(hemoglobin S)* due to a genetic defect. It is found primarily in African-Americans. It is more common in females and usually clinically manifests itself before the age of 30. The typical signs of anemia are present. The patient is weak, short of breath and easily fatigued. Muscle and joint pains are common.

Dental radiographs are often of diagnostic value → marrow spaces are markedly enlarged because of the loss of many trabeculae; the trabeculae, which are present, are often abnormally prominent. Occasionally, osteosclerotic areas are noted in the midst of large radiolucent marrow spaces. **However,** the lamina dura and the teeth are unaffected.

See picture #25 in booklet.

A condition characterized by **hemorrhages in the skin** and mucous membranes that result in the appearance of purplish spots or patches is called:

- Addison's disease
- Purpura
- Aplastic anemia
- Hemophilia

- **Purpura**

Major kinds of purpura:

- **Thrombocytopenic purpura** *(also known as* **Werlhof's disease***)* is a bleeding disorder characterized by a deficiency in the **number of platelets.** This results in multiple bruises, petechiae and hemorrhage into the tissues. **Note:** May be caused by heparin therapy.

- **Thrombotic thrombocytopenic purpura** *(TTP)* is a severe and frequently fatal form characterized by a low platelet count in the blood and thrombosis in the terminal arterioles and capillaries of many organs.

Oral manifestations of thrombocytopenic purpura:

- Severe and profuse **gingival hemorrhage**
- Petechiae occur commonly on the palate

Important: Tooth extractions are **contraindicated** due to the tendency for excessive bleeding.

All of the following statements concerning **acute leukemia** are true, **except:**

- It has an insidious onset
- Acute myelogenous leukemia is more common in adults
- Acute lymphocytic leukemia is largely confined to children
- Symptoms include severe anemia, hemorrhages, and slight enlargement of the lymph nodes or the spleen

• **It has an insidious onset**

***This is false;** acute leukemia has an **abrupt onset.**

The clinical symptoms and course of the various types of **acute leukemia** are similar.

- **Abrupt onset** *(few months)* with fever, weakness, malaise, severe anemia and generalized lymphadenopathy; bone and joint pain common in children.
- Principal organs involved are bone marrow, spleen *(splenomegaly)* and liver *(hepatomegaly)*. **Lymph node enlargement is common in acute lymphocytic leukemia.**
- **Petechiae and ecchymoses** in skin and mucous membranes, hemorrhage from various sites; bacterial infections common.
- **Laboratory findings** → leukocytosis 30,000-100,000 per cu. mm. with immature forms *(myeloblasts and lymphoblasts)* predominating; anemia and thrombocytopenia; bleeding and coagulation times may be prolonged; tourniquet test usually positive.
- In 75% of the cases of acute **lymphocytic** *(lymphoblastic)* leukemia, the lymphocytes are neither B nor T cells and are called **"null"** cells.
- **Untreated patients die within 6 months;** with intensive therapy *(chemotherapy, radiation and marrow transplants)* remissions lasting up to 5 years may be obtained; death is usually due to hemorrhage *(usually of the brain)* or a superimposed bacterial infection.

Histologically, which of the following is pathognomonic of **agranulocytosis?**

- The ulcerated areas exhibit **an intense** polymorphonuclear reaction due to the bacteria in the tissues
- The ulcerated areas exhibit **no** polymorphonuclear reaction due to the bacteria in the tissues
- Agranulocytosis **does not exhibit anything** histologically that is pathognomonic of this disease

- **The ulcerated areas exhibit no polymorphonuclear reaction due to the bacteria in the tissues**

Agranulocytosis is an abnormal condition of the blood, characterized by a **severe reduction in the number of granulocytes** *(particularly neutrophils)*. It may be caused by the **antithyroid drugs** *(i.e., propylthiouracil, methimazole, and carbimazole)*.

Remember: One important aspect of agranulocytosis is that there is little or no apparent inflammatory cell infiltration around the lesions. These lesions *(referred to as* **necrotizing ulcerations***)* appear on the gingiva and the palate. This disease is most commonly caused by **ingestion of a drug.** The treatment involved is to find out the causative drug and eliminate it. The administration of antibiotics to control the infection is critical.

Note: Cyclic neutropenia is an unusual form of agranulocytosis. These patients typically exhibit **severe gingivitis.** The severe ulcerations usually seen in agranulocytosis usually do not occur.

A form of leukemia in which the abnormal cells are thought to be the **precursors** of lymphoblasts, myeloblasts, or monoblasts is called:

- Aleukemic leukemia
- Subleukemic leukemia
- Stem cell leukemia

- **Stem cell leukemia**

***The cells are **too immature** to classify.

Other leukemia **terms** to know:

- **"Aleukemic"** leukemia is a term to describe a form of leukemia in which there are leukemic cells present in the bone marrow, but the circulating white blood cells are **neither immature nor increased in number.**

- **"Subleukemic"** leukemia is a term to describe a form of leukemia in which leukemic cells appear in the blood but there is **no significant increase** in the number of circulating white blood cells.

- A **"leukemoid"** reaction is a term to describe a marked increase in the number of circulating granulocytes. This condition is seen in a variety of disorders including chronic infections and neoplasms.

An inability to absorb adequate amounts of **vitamin B_{12}** from the digestive tract may result in:

- Thalassemia
- Pernicious anemia
- Aplastic anemia
- None of the above

• **Pernicious anemia**

Pernicious anemia is a relatively common, chronic, progressive, **megaloblastic anemia.** It is caused by the lack of secretion of the **intrinsic factor** in normal gastric juice. This factor is necessary for adequate absorption of vitamin B_{12}, which is necessary for the **maturation of erythrocytes.** As a result, they produce fewer erythrocytes than normal. This disease is often characterized by the presence of a **triad of symptoms:** generalized weakness; **a sore, painful tongue** *(atrophic glossitis);* and numbness and tingling of the extremities.

See picture #65 in booklet.

Remember: Thalassemia major and minor are **hemolytic anemias** that result from a genetic defect. Both are characterized by a **low level of erythrocytes** and abnormal hemoglobin.

Oral manifestations of thalassemia:
• Oral mucosa may exhibit the characteristic **anemic pallor**
• **Flaring of the maxillary anterior teeth** with malocclusion

A severe hemolytic disease of the fetus or newborn caused by the **production of maternal antibodies** for fetal red blood cells is called:

- Sickle-cell anemia
- Erythroblastosis fetalis
- Erythrocyanosis
- Aplastic anemia

- **Erythroblastosis fetalis**

It usually involves Rh incompatibility between the mother and fetus *(it is also called* **hemolytic disease of the newborn)*

This condition of erythroblastosis is characterized by an **excessive destruction of erythrocytes.** It is caused by an antigen-antibody reaction in the bloodstream of the infant resulting from the placental transmission of maternally formed antibodies against the incompatible antigens of the fetal blood. In Rh factor incompatibility, the hemolytic reaction only occurs when the **mother is Rh negative and the infant is Rh positive.**

Oral manifestations of erythroblastosis fetalis:

- **Teeth** appear to have a green, blue or brown hue due to the deposition of blood pigment in the enamel and dentin.

- **Enamel hypoplasia** may occur. If it does, it affects the incisal edges of the anterior teeth and the middle portion of the deciduous cuspid and the first molar crown.

All of the following statements concerning **chronic leukemia** are true, **except:**

- It has an abrupt onset
- The skin is frequently involved in chronic leukemia and may manifest as petechiae or ecchymoses
- Massive splenomegaly is characteristic of chronic **myelogenous** leukemia
- Lymph node enlargement is the main pathologic finding in chronic **lymphocytic** leukemia

- **It has an abrupt onset**

***This is false;** chronic leukemia **has an insidious onset.**

Clinical features of **chronic** leukemia:

- **Insidious onset** with weakness and weight loss: chronic leukemia may be detected during examination for some other condition *(for example anemia, unexplained hemorrhages or recurrent intractable infections).*
- **Organ involvement similar to acute type**→ massive splenomegaly is characteristic of chronic **myelogenous** leukemia; lymph node enlargement is main pathologic finding in chronic **lymphocytic** leukemia. **Note:** Lymphocytic leukemia may be complicated by autoimmune hemolytic anemia.
- **Petechiae and ecchymoses** are common along with recurrent hemorrhages and bacterial infections: anemia.
- **Laboratory findings** → leukocytosis **above** 100,000 per cu. mm. with mature forms *(granulocytes and lymphocytes)* predominating. The Philadelphia chromosome and low levels of leukocyte alkaline phosphatase are common findings in **chronic myelogenous** *(myelocytic)* leukemia.
- **Median survival time** for patients with chronic **myelogenous** leukemia is 4 years with death due to hemorrhage or infection; chronic **lymphocytic** leukemia runs a variable course; older patients may survive years even without treatment.

Oral lesions are most likely to be observed in:

- Myelogenous leukemia
- Lymphocytic leukemia
- Monocytic leukemia

• **Monocytic leukemia**

Important: Oral lesions may be the **initial manifestation** of the disease. The oral lesions include gingivitis, gingival hemorrhage, generalized gingival hyperplasia, petechiae, ecchymoses, and ulcerations.

Classifications of leukemia:

- Myelogenous → involves the granulocytes and megakaryocytes
- Lymphocytic → involves the lymphocytes
- Monocytic → involves the monocytes

Leukemia is a cancerous condition in which an uncontrolled proliferation of leukocytes leads to a diffuse and almost total replacement of the red bone marrow with leukemic cells.

Important points about leukemia:

1. All types of leukemia occur in an **acute or chronic form**, but about 50% are acute.
2. Acute myelogenous leukemia is the **most malignant type** and chronic lymphocytic is the **least malignant**.
3. Acute lymphocytic *(lymphoblastic)* is the most common type of leukemia in **children**.
4. Chronic monocytic leukemia is **very rare.**

All of the following statements concerning **polycythemia vera** are true, **except:**

- It is characterized by excessive erythrocyte production
- It usually occurs within the age range of 10-35, with 20 being the mean age of onset
- Polycythemia vera is considered to be one of the chronic myeloproliferative disorders
- Clinical features include headaches, weakness, weight loss, and pruritus
- Hemorrhage and thrombosis may be evident at any time

- **It usually occurs within the age range of 10-35, with 20 being the mean age of onset**

***This is false;** Polycythemia vera usually occurs within the age range of **20-80**, with 60 being the mean age of onset.

Polycythemia is the condition of **too many red blood cells** in the circulation. The blood can be too thick to pass easily through the small blood vessels of the body. This in turn leads to clot formation and blockage of the small vessels which can lead to a stroke. There are **two types** of polycythemia:

1. **Primary polycythemia** *(also called* **polycythemia vera** *or erythemia)* occurs when excess erythrocytes are produced as a result of tumorous abnormalities. This occurs in the tissues that produce blood cells. Usually accompanied by leukocytosis. **Splenomegaly**, as a result of vascular congestion, is seen in 75% of patients.
2. **Secondary polycythemia** is an increase in the total number of erythrocytes due to another condition. For example, chronic tissue hypoxia of advanced pulmonary disease, high altitude *(Osker's disease)* or the secretion of erythropoietins by certain tumors.

Oral manifestations of polycythemia:

- Oral mucous membranes *(especially the gingiva and tongue)* appear deep purplish-red.
- The **gingiva** are very **swollen and bleed** very easily.
- Submucosal petechiae *(purplish spots),* ecchymoses *(same as petechiae, but bigger)* and hematomas are common.

An **acute condition** characterized by pronounced **leukopenia** with a severe reduction in the number of **polymorphonuclear** leukocytes is called:

- Infectious mononucleosis
- Agranulocytosis
- Sickle-cell anemia
- Porphyria

• **Agranulocytosis**

Agranulocytosis is a toxic effect of certain drugs *(for example,* **antithyroid** *drugs).* It can occur at any age, but it somewhat more common in adults, particularly women.

The white blood cell count is often **below 2,000** with an almost complete absence of polymorphonuclear leukocytes *(neutrophils).* **Note:** The normal WBC is between 4,000-10,000 and neutrophils are usually 50-70%.

This condition commences with a high fever, accompanied by chills and sore throat. The patient suffers from malaise, weakness and prostration. The skin appears pale and anemic. The **most characteristic feature** of this condition is the presence of **infection**, particularly in the **oral cavity.** The signs and symptoms develop very rapidly, usually within a few days, and death may occur soon afterward.

The oral lesions are an important phase of the clinical aspects of agranulocytosis. They appear as necrotizing ulcerations of the oral mucosa, particularly the gingiva and palate. These lesions appear as ragged necrotic ulcers covered by a gray membrane. One important aspect is that there **is little or no apparent inflammatory cell** infiltration around the lesions.

The leukemic cells of more than 95% of patients having **chronic myelogenous leukemia** *(CML)* have a *(an)*:

- Missing chromosome
- Extra chromosome
- Philadelphia chromosome
- New York chromosome

• **Philadelphia chromosome**

***The Philadelphia chromosome is the result of a reciprocal translocation between chromosomes 9 and 22 which results in a **shortened chromosome 22**

Chronic myelogenous leukemia *(CML)* is one of a group of diseases called the **myeloproliferative disorders.** Other related entities include polycythemia vera, myelofibrosis and essential thrombocythemia. CML is characterized by uncontrolled proliferation of immature granulocytes. CML accounts for 20% of all leukemias in adults. It typically affects middle-aged individuals. Although uncommon, the disease also occurs in younger individuals.

The typical symptoms of **CML** include:
• **spongy bleeding gums**
• fatigue
• fever
• weight loss
• moderate splenomegaly
• joint and bone pain
• **repeated infections**

Note: Acute myeloid leukemia *(AML)* is a malignant disease of the bone marrow in which hematopoietic precursors are arrested in an early stage of development. AML is distinguished from other related blood disorders by the presence of greater than 30% blasts in the blood and/or bone marrow. These blasts *(myeloblasts)* contain **Auer rods** in their cytoplasm.

The **oral traumatic neuroma** usually appears as a small nodule or swelling of the mucosa typically:

- Near the mental foramen
- On the alveolar ridge in edentulous areas
- On the lips
- On the tongue
- All of the above

• **All of the above**

***The **most common site** is over the mental foramen in edentulous mouths. **However**, they may occur **wherever a tooth has been removed.** Extraction sites in the anterior maxilla and the posterior mandible are relatively common sites.

A **traumatic neuroma** is a lesion caused by trauma to the peripheral nerve. In the oral cavity, the injury may be in the form of trauma from a surgical procedure such as a tooth extraction, from a local anesthetic injection or from an accident. It is usually a very small nodule *(less than 0.5 cm in diameter).* It is **painful when palpated**. Pressure applied to the neuroma elicits a response often described as an **"electric shock".** The treatment is surgical excision.

Remember: Multiple neuromas discovered on the lips, tongue or palate may indicate the possibility that the patient has **MEN III** *(multiple endocrine neoplasia syndrome).*

The most common site in the oral cavity for a **lymphangioma** is the:

- Palate
- Tonsils
- Floor of the mouth
- Tongue

• **Tongue**

***A lymphangioma is a benign, yellowish-tan tumor, composed of a mass of dilated lymph vessels.

Lymphangioma			
Histogenesis *(Etiology)*	**Clinical Characteristics**	**Microscopic Characteristics**	**Treatment and Prognosis**
Endothelial cells, connective tissue origin	Painless, nodular, vesicle-like swelling, Effects both sexes equally. The intraoral lymphangioma most commonly occurs on the tongue, but also appears on the lips and neck. The superficial lesions are manifested as papillary lesions, which are grayish-red. On the tongue, considerable enlargement may occur (**macroglossia**). The papillary lesions may contain fluid. They are often present at birth or arise in early life. **Less common than hemangioma.**	**Three types:** (1) Simple (2) Circumscriptum (3) Cystic – **closely related to cystic hygroma,** contains serous fluid.	Surgery, cryosurgery; may recur due to their lack of encapsulation

Which of the following is most often caused by **retained foreign material** *(i.e., bone, tooth fragment, etc.)*?

- Pyogenic granuloma
- Pregnancy tumor
- Epulis ganulomatosum
- Fibroma

- **Epulis granulomatosum**

Entity	Cause	Location	Clinical	Microscopic	Treatment
Pyogenic granuloma	Minor trauma provides pathway for non-specific organisms, calculus	Gingiva most common, lips, buccal mucosa	Elevated mass, often ulcerated, bleeds easily, greater with females, may recu	Exuberant granulation tissue	Surgical excision, may recur
Pregnancy tumor*	Possible secondary to altered endocrine state during pregnancy *(first trimester)*	Gingiva most common, lips buccal mucosa	No justification for continued use of term as it is a pyogenic granuloma*	Exhuberant granulation tissue	Don't remove until after pregnancy
Epulis granulomatosum	Retained foreign material *(bone, tooth, fragment)* **"Iatrogenic"**	Post-extraction **socket**, almost always within 10 days of extraction	Soft, non-painful, bleeds easily	Granulation tissue within which one may find bone, dentin, cementum or foreign material	Curettage
Fibroma *(also called "Irritation Fibroma" or "Traumatic Fibroma")*	Reactive, most common tumor seen in oral cavity	Buccal mucosa lateral border of tongue and lower lip	Elevated, smooth pink, painless, well demarcated mass. Same color or lighter than normal mucosa	Bundles of collagen interspersed with fibroblasts and small blood vessels	Conservative surgical excision

*Term is used for a **pyogenic granuloma in a pregnant patient**

All of the following statements concerning the **peripheral ossifying fibroma** are true, **except:**

- It is most common in elderly people
- It usually presents as a well-demarcated focal mass of hyperplastic tissue on the gingiva with a sessile or pedunculated base
- It is usually the same color as normal mucosa or slightly reddened
- It may demonstrate bone radiographically and often demonstrates bone formation histologically

- **It is most common in elderly people**

***This is false;** it can occur at any age, although it appears to be **somewhat more common in children and young adults.**

The **peripheral ossifying fibroma** is a subtype or variant form of the **peripheral fibroma.** Both are seen more frequently in **young adult females** and the gingiva, anterior to the permanent molars is most frequently affected. They both appear to originate from an interdental papilla.

The **peripheral ossifying fibroma** is a gingival mass in which calcified islands, **presumed to be bone,** are seen. The surface is often ulcerated. The lesion is quite characteristic, **histologically**, in its high degree of cellularity *(usually exhibiting bone formation)* in contrast to the peripheral fibroma. Vascularity is not a prominent feature as it is in the **pyogenic granuloma.** Treatment is also local excision. **Note:** These lesions tend to recur, **as opposed to** the peripheral fibroma, which does not.

The **peripheral fibroma** also presents as well-demarcated focal mass of hyperplastic tissue with either a sessile or pedunculated base. It is similar in color to the surrounding connective tissue. It may be ulcerated. The treatment for a peripheral fibroma is local excision. **Recurrence is rare.**

Note: Other variant forms of the **peripheral fibroma** include:

- The **peripheral odontogenic fibroma:** which is gingival mass composed of a well-vascularized, non-encapsulated fibrous connective tissue.
- The **giant cell fibroma:** which is a fibrous hyperplasia composed of multi-nucleated connective tissue cells.

The **multiple endocrine neoplasia syndromes** are a group of syndromes characteized by:

- Tumors of the CNS
- Tumors of the skin
- Tumors of the various endocrine glands
- Tumors of the salivary glands

• **Tumors of the various endocrine glands**

***These tumors occur in association with a variety of other pathologic features.

The **multiple endocrine neoplasia syndromes** *(also called* **MEN Syndrome***)* have been classified into three groups:

- **Men I** → consists of tumors or hyperplasias of the pituitary, parathyroids, adrenal cortex, and of the pancreatic islets.
- **Men II** *(also called* **Sipple's Syndrome***)* → is characterized by parathyroid hyperplasia or adenoma, but no tumors to the pancreas. **However**, in addition, these patients have pheochromocytomas of the adrenal medulla and medullary carcinoma of the thyroid gland.
- **Men III** → is characterized by **mucocutaneous neuromas,** pheochromocytomas of the adrenal medulla and medullary carcinoma of the thyroid gland.

Important: The most constant feature of **Men III** is the presence of neuromas, particularly of the **oral cavity.** These are most common on the lips, tongue, and buccal mucosa.

Note: The most important aspect of this syndrome is the **medullary carcinoma of the thyroid** because of its ability to metastasize and cause death. Therefore, the detection of the mucosal neuromas may alert the clinician for early diagnosis and treatment.

ORAL PATHOLOGY

Con Tis Les

Which of the following soft tissue tumors is most likely to be found over the **mental foramen in an edentulous patient**?

- Traumatic neuroma
- Neurilemoma *(Schwannoma)*
- Neurofibroma

- **Traumatic neuroma**

Tumor	Etiology	Clinical Characteristics	Treatment and Prognosis
Traumatic Neuroma	Trauma to a peripheral nerve	Most common site over **mental foramen** in edentulous mouths; nodule or swelling, which may be painful to digital pressure	Excision with small proximal portion of involved nerve; recurrence uncommon
Neurilemoma *(Schwannoma)*	It is derived from proliferation of **Schwann cells** of the neurolemma that surrounds peripheral nerves	Encapsulated mass that presents as an asymptomatic lump. The **tongue** is the most common location. Bony lesions may cause pain or paresthesia.	Conservative excision; recurrence rare
Neurofibroma	Some investigators say it is derived from the Schwann cell; others say the perineural fibroblast	**Two forms:** 1. **Solitary neurofibroma** – asymptomatic nodule, occurs on tongue, buccal mucosa and vestibule 2. **Multiple** lesions as part of the syndrome neurofibromatosis	1. **Solitary**: surgical excision 2. **Neurofibromatosis:** removal is impractical. Watch for high rate of **malignant transformation**

The most outstanding feature of **Von Recklinghausen's** disease is:

- Vertigo
- Hemorrhage
- Neurofibromatosis
- Osteoporosis

• **Neurofibromatosis**

***This is a condition of **multiple tumors of nerve tissue origin.**

Von Recklinghausen's disease is inherited as an autosomal dominant trait. It is relatively common and is characterized by multiple neurofibromas, cutaneous café-au-lait macules, bone abnormalities and CNS changes. The presence of **six or more café-au-lait macules** greater than 1.5 cm in diameter is generally regarded as being **indicative** of this disease until proven otherwise.

There is no satisfactory treatment. The importance of the lesions is the high risk of **malignant transformation.**

Remember: The single neurofibroma presents at any age as an uninflamed, asymptomatic nodule that commonly occurs on the **tongue, buccal mucosa and vestibule.** It is removed by surgical excision and rarely recurs.

The most frequently encountered intraoral benign neoplasm of **connective tissue origin** is:

- A papilloma
- A fibroma
- A lipoma
- A nodular melanoma

• **A fibroma**

***Also called an **"irritation fibroma"** or a **"traumatic fibroma."**

Fibromas occur in people of all ages and with equal frequency in both sexes. They may arise from almost any soft tissue in the mouth, although they are found most commonly on the buccal mucosa, lateral border of the tongue and the lower lip. They usually present as pink, painless, smooth, elevated, well-demarcated masses. The history is often helpful in establishing a diagnosis, since, in most cases, the tumor is reported to have been present for months or years and to have a **slow-growing** behavior pattern.

See picture #7 in booklet.

Note: Some investigators feel that **"true fibromas"** of the oral cavity are rare and that in reality, they are simply examples of localized hyperplasia, resulting from **long-standing irritation or trauma** *(hence the term* **"irritation fibroma"** *or* **"traumatic fibroma"***)*. These fibrous nodules are comparable with the hyperplasias from denture irritation *(termed* **"epulis fissuratum"***)*. The only difference between the **"true fibroma"** *(which is a true neoplasm)* and the **"irritation fibroma"** *(which is not a true neoplasm)* is that the hyperplastic tissue **may regress after removal** of the irritant, while the true fibroma will not regress. Either way, the treatment is conservative surgical excision.

Remember: A papilloma is the most common benign neoplasm of **epithelial tissue origin.**

ORAL PATHOLOGY

Con Tis Les

Oral radiographs of a patient with **scleroderma** would show:

- A "sun-ray" appearance of the bone
- An obscure lamina dura
- An abnormal widening of the periodontal ligament
- Vertical bone loss

• An abnormal widening of the periodontal ligament
*****See picture #11 in booklet.**

The space is created by a thickening of the periodontal-membrane as a result of an increase in size and number of collagen fibers. The enlarged space is almost **uniform in width,** surrounds the entire root of the tooth and makes the tooth appear as if it is being extruded rapidly from its socket. **Note:** Other oral radiographic features may include bilateral resorption of the angle of the ramus of the mandible or complete resorption of the condyles and/or coronoid process of the mandible.

Remember: The abnormal widening of the periodontal-membrane space is also a radiographic finding in **osteosarcomas**.

Scleroderma is a relatively rare autoimmune disease affecting the blood vessels and connective tissue. It is characterized by hardness and rigidity of the skin and subcutaneous tissue. The continuous deposition of collagen in major organs may lead to dysfunction and potentially to failure of these organs.

Clinical features → systemic scleroderma appears usually during middle age *(30-50 years old)*, predominantly in **females** *(4:1)*. The skin is usually affected first and becomes indurated.

Treatment → other than supportive therapy, there is no satisfactory treatment for scleroderma.

The **congenital epulis of the newborn** is composed of cells that are identical to those of:

- A traumatic neuroma
- A schwannoma
- A granular cell myoblastoma
- A lipoma

• **A granular cell myoblastoma** *(also called a granular cell tumor)*

The **congenital epulis of the newborn** *(also called congenital gingival granular cell tumor)* usually appears on the **anterior gingiva** of newborns. It presents as a non-inflamed, pedunculated or broad-based mass. The **maxillary gingiva** is more often involved than the mandibular gingiva, and **females** are affected more than males. The treatment is surgical excision with little possibility of recurrence.

The **granular cell myoblastoma** is an uncommon neoplasm of unknown etiology. It presents as an uninflamed, asymptomatic mass. The most common location in the head and neck region is the **tongue**. It may affect any age group and females seem to be affected more than men.

Important: Both of these lesions are identical histologically. They both contain **granular cells,** however, the congenital epulis of the newborn **does not exhibit** overlaying pseudoepitheliomatous hyperplasia. The pseudoepitheliomatous hyperplasia of the overlying epithelium is frequently seen in the **granular cell myoblastoma.**

A periapical abscess **usually** arises as a result of:

- Orthodontics
- Trauma
- Infection of the pulp of a tooth
- Periodontal disease

- **Infection of the pulp of a tooth**

***This infection follows the **carious** involvement of the tooth. The cellular debris and / or infection which caused the tooth pulp to die, slowly filters out of the tip of the root and produces an inflammatory reaction around the root tip.

Note: A periapical abscess can also occur after traumatic injury to a tooth, which results in necrosis of the pulp, and in cases of irritation of the periapical tissues, either by mechanical manipulation or by the application of chemicals in endodontic procedures.

Clinical features:

- If **acute**, presents as an **abscess:**
  - Tooth is extremely painful to percussion
  - May feel slightly extruded from its socket
  - Tooth will exhibit mobility
- If **chronic**, presents as a **granuloma or cyst.** There are usually **no clinical** features or symptoms

Radiographic features:

- If **acute**, only a slight thickening of the periodontal membrane is noticeable
- If **chronic** *(granuloma or cyst),* there will usually be a radiolucent area at the apex of the involved tooth

Treatment: Establish **drainage** either by opening the pulp chamber or extracting the tooth.

Note: If a periapical abscess is **not treated,** it can lead to serious complications such as osteomyelitis, cellulitis and bacteremia.

The **inflammation or infection** of the bone marrow and adjacent bone is called:

- Osteopetrosis
- Osteoporosis
- Osteomyelitis
- Osteonecrosis

• **Osteomyelitis**

It is usually caused by **bacteria** *(staphylococci)* introduced by trauma or surgery, by direct extension from a nearby infection, or via the bloodstream. It often presents the following signs and symptoms: **pain, redness and swelling** in the infected area. Fever and general malaise are usually evident. **Radiographically**, you may see a poorly circumscribed radiolucency with a central sclerotic nidus. **See picture #15 in booklet.**

Remember:

1. **Osteonecrosis** is the death of bone.

2. **Osteoporosis** is a reduction of total skeletal mass due to increased bone resorption, which results in predisposition to pathologic fractures. It is common especially in **thin, elderly, white women.** It is caused by calcium or hormone *(estrogen)* deficiencies over a long period of time. The treatment includes estrogen therapy, calcium supplements and vitamin D.

3. **Osteopetrosis** is a congenital disease that prevents formation of bone marrow and results in abnormal bone development, blindness, stunted growth, abnormal dental development and fragile bones. It results from a defect in osteoclasts which are necessary for the formation of bone marrow. Also called Albers-Schonberg disease and marble bone disease.

All of the following statements concerning **condensing osteitis** are true, **except:**

- It most often occurs in young patients
- The mandibular second molar is the tooth most commonly involved
- It is most often seen associated with a long-standing periapical infection
- There may be no signs or symptoms of the disease other than mild pain associated with an infected pulp

• **The mandibular second molar is the tooth most commonly involved**

***This is false; the **mandibular first molar** is the tooth most commonly involved.

Chronic focal sclerosing osteomyelitis *(a fancy name for condensing osteitis)* is an unusal reaction of bone to infection, occurring in instances of extremely high tissue resistance, or in cases of low-grade infection.

The periapical x-ray demonstrates the pathognomonic, **well-circumscribed radiopaque mass** of sclerotic bone surrounding and extending below the apex of one or both roots. The **entire root outline is almost always visible,** an important feature in distinguishing it from the **benign cementoblastoma,** which radiographically, it may resemble. The tooth with this lesion may be treated or extracted, since the pulp is infected and the infection has spread past the immediate periapical area. The sclerosing bone constituting the osteomyelitis is not attached to the tooth, and remains after the tooth is treated or removed.

See picture #37 in booklet.

Lateral clefting of the lip results from the failure of:

- The maxillary processes to merge
- The palatine processes to merge
- The maxillary and frontonasal processes to merge
- None of the above

• **The maxillary and frontonasal processes to merge**

Cleft lip occurs during the **fifth to sixth week** of embryonic life. It may be bilateral or unilateral. Clefts of the lip are more frequent in **males**. Lip cleft involvement is more frequent on the **left side** than the right.

Cleft palate occurs in the **sixth to eighth week** of embryonic life. Isolated clefts of the palate are more common in females. It is characterized by a fissure in the midline of the palate, resulting from the failure of the two sides to fuse during embryonic development. The most severe handicap imposed by cleft palate is an impaired mechanism **preventing normal speech and swallowing.**

Note: Speech problems associated with both of the above are usually the result of the **inability of the soft palate to close airflow into the nasal area.**

An uncommon hereditary disorder characterized principally by the **overgrowth** and denseness of **bones** is called:

- Osteopetrosis
- Osteogenesis imperfecta
- Achondroplasia
- Fibrous dysplasia

• **Osteopetrosis**

Long bones become dense and hardened to an extent that the marrow is obliterated; anemia along with spleen and liver enlargement, blindness and progressive deafness occur. It begins in infancy. It is also called **Albers-Schönberg disease** and **marble bone disease.**

Remember:
Osteogenesis imperfecta is a genetic disorder also known as **"brittle bones."** This disorder is rare but does demonstrate the effect of inadequate osteoid production. It is characterized by bones that break easily often from little or no apparent cause. The teeth are poor because of the malformation of dentin *(***Remember: Type I dentinogenesis imperfecta** *is associated with osteogenesis imperfecta).*

Achondroplasia is the most common type of dwarfism. Clinically the child has the following appearance: very short *(50 inches is average)*, fingers are stubby, legs are bowed, bulging of the forehead and bossing of the frontal bones are present. The nose has a **saddle-like** appearance and the **mandible exhibits prognathism.**

Fibrous dysplasia is characterized by normal bone being replaced by fibrous tissue. There are three classifications depending on the extensiveness of the skeletal involvement:

1. **Monostotic** → one bone 2. **Polyostotic** → more than one bone and 3. **Polyostotic with associated endocrine disturbances** *(called* **Albright's syndrome***)* → pathologic fractures are often the presenting complaint.

Osteogenesis imperfecta is caused by a genetic defect that affects the body's production of:

- Vitamin K
- Collagen
- Reticulin
- Elastin

• **Collagen**

Collagen is the major protein of the body's connective tissue. In osteogenesis imperfecta, a person has either less collagen than normal, or a poorer quality of collagen than normal – **leading to weak bones that fracture easily.**

The characteristic features of osteogenesis impefecta **vary greatly** from person to person and **not all characteristics** are evident in each case. The chief clinical characteristic of osteogenesis imperfecta is the extreme fragility and porosity of the bones, with a **proneness to fracture.** Other features include: pale blue sclera, deafness due to otosclerosis, abnormal teeth *(***Remember:** *Type I dentinogenesis imperfecta is associated with osteogenesis imperfecta)*, loose joints and low muscle tone, a triangular face, and a tendency toward spinal curvature.

Clinically the teeth have:

- Crowns that are bulbous; with a cervical constriction.
- Pulps that are obliterated, either partially or completely.
- Roots that are narrower and shorter.

***The deciduous teeth are **more severely affected** that the permanent dentition.

There is **no known** cure for osteogenesis imperfecta. Treatment is directed toward preventing or controlling the symptoms.

Hypophosphatasia is one of several disorders that resembles:

- Acromegaly
- Paget's disease
- Osteogenesis imperfecta
- Ewing's sarcoma

• **Osteogenesis imperfecta**

Hypophosphatasia is an inherited metabolic *(chemical)* bone disease that results from low levels of an enzyme called **alkaline phosphatase**. This enzyme is essential to the calcification of bone tissue. The severity of hypophosphatasia is remarkably variable from patient to patient. Some patients have blue sclera that resembles osteogenesis impecta. There may be deformity of the arms, legs and chest. Frequent bouts of pneumonia can occur as well as recurrent fractures. In general, patients are categorized as having:

- **"Perinatal"** hypophosphatasia → fail to form a skeleton in the womb and are stillborn
- **"Infantile"** hypophosphatasia → is manifested by severe rickets, hypercalcemia, and bone abnormalities. Most cases are lethal.
- **"Childhood"** hypophosphatasia → there is the premature exfoliation of deciduous teeth, increased infection, numerous skeletal abnormalities and those children that survive are dwarfs.
- **"Adult"** hypophosphatasia → is manifested by spontaneous fractures, prior history of rickets and osseous radiolucencies.

Important: The loosening and premature loss of the deciduous teeth is usually characteristic. These teeth also exhibit hypocalcification. Radiographically the teeth display **large pulp chambers,** as well as **alveolar bone loss.**

All of the following statements concerning **acromegaly** are true, **except:**

- It is a hormonal disorder that results when the pituitary gland produces excess ACTH
- It most commonly affects middle-aged adults and can result in serious illness and premature death
- Soft tissue swelling of the hands and feet is often an early feature, with patients notic ing a change in ring or shoe size
- Gradually, bony changes alter the patient's facial features *(i.e., the brow and lower jaw protrude, the nasal bone enlarges, and spacing of the teeth increases)*

- **It is a hormonal disorder that results when the pituitary gland produces excess ACTH**

***This is false;** it is a hormonal disorder that results when the pituitary gland produces excess **growth hormone.**

In over 90% of acromegaly patients, the overproduction of GH is caused by a benign tumor of the pituitary gland, called an adenoma. Whether or not the epiphyses of the long bones have fused with the shaft is the main determinant of whether gigantism or acromegaly will occur when there is oversecretion of growth hormone by the pituitary gland.

Remember:

- **Gigantism** → tumor prior to adolescence *(non-fusion of epiphyses)*
- **Acromegaly** → tumor after adolescence *(fusion of epiphyses)*

Oral manifestations of acromegaly and gigantism include: enlarged tongue, mandibular prognathism, teeth are usually tipped to the buccal or lingual side, owing to enlargement of the tongue. Roots may be longer than normal.

Note: Dwarfism *(pituitary dwarfs)* is characterized by arrested growth. Frequently these people have limbs and features not properly proportioned or formed. It is caused by undersecretion of growth hormone. **Oral manifestations** include: eruption rate and the shedding of the teeth are delayed, clinical crowns appear smaller as do the roots of the teeth, the dental arch as a whole is smaller causing malocclusion and the **mandible is underdeveloped.**

A five-year-old child **lacks sweat** and **sebaceous glands.** He has **fine, sparse hair** and exhibits heat intolerance. His radiographs **do not** reveal tooth buds of either the primary or permanent dentition. **These findings are all consistent with:**

- Pierre Robin syndrome
- Ectodermal dysplasia
- Cleidocranial dysostosis

• **Ectodermal dysplasia**

Ectodermal dysplasia is a hereditary condition characterized by abnormal development of the skin and associated structures *(hair, nails, and teeth, and sweat glands).* It involves all structures which are derived from the **ectoderm.** It affects males more than females. Common clinical findings include **hypothrichosis** *(decrease in hair)*, **anhidrosis** *(no sweat glands, leading to heat intolerance),* **anodontia** or **oligodontia** *(complete or partial absence of teeth),* **depressed bridge of nose, lack of salivary glands** and the **child appears much older than what he or she is**. There is no treatment for the disease, however dentures can be fabricated for these patients. Keep in mind that they will need to be replaced periodically to accommodate the patient's jaw growth.

Cleidocranial dysostosis is an inherited disorder of bony development characterized by absent or incompletely formed collar bones, a characteristic facial appearance *(a heavy protruding jaw and wide nasal bridge)*, and dental abnormalities which include **malaligned** teeth, the presence of **multiple supernumerary** teeth and **unerupted** teeth. **Important:** The dentition itself, as observed by radiographs alone, often suggests the diagnosis. **See picture #62 in booklet.**

Pierre Robin syndrome is an inherited disorder that presents the following in the neonate: **micrognathia** *(smallness of the jaws)*, **glossoptosis** *(downward displacement or retraction of the tongue),* breathing problems, and **cleft palate**.

An **increased serum alkaline phosphatase** level is clinically significant and aids in the diagnosis of:

- Hyperparathyroidism
- Paget's disease of bone
- Myasthenia gravis
- Prostate cancer

- **Paget's disease of bone**

***Paget's disease of bone is also called **Osteitis Deformans.**

Paget's disease of bone is a chronic bone disorder in which bones become enlarged and deformed. The bone may become dense, but fragile, because of excessive breakdown and formation of bone. The disease affects both genders and is rarely found in people under the age of 40. The cause is unknown *(it appears to be familial).* The signs and symptoms include pain in the affected area, deformity of the bone in the affected area, susceptibility to fractures in the affected area, and headache and hearing loss if the affected area is the skull. **Note:** These symptoms develop **slowly**.

Other important features of this disease include:

- Patients may also give a history of progressively increasing size of hats or new dentures being made at progressively more frequent intervals. **Note:** This is due to bony changes.
- Bones are warm to touch due to increased vascularity.
- X-rays of the skull and the jaws demonstrate the typical **"cotton-wool"** appearance. The teeth have pronounced hypercementosis and, often, the loss of lamina dura.
- Lab tests: Drastically **increased serum alkaline phosphatase**. Serum phosphate and calcium are normal. Urinary calcium and hydroxyproline are increased.
- Treatment: Calcitonin decreases bone resorption. Antimetabolites may also be used.

Important: These patients are predisposed to developing **osteosarcomas**.

See picture #64 in booklet.

All of the statements concerning **cherubism** are true, **except:**

- It is an autosomal dominant inherited disease
- The expansion of the jaws gives the children a very rounded face reminding one of cherubs in paintings
- Histologically, the lesions bear a close resemblance to those seen in central giant cell granulomas
- The treatment is radiation therapy

• **The treatment is radiation therapy**

***This is false;** the treatment is **cautious waiting** as it tends to regress in early adulthood.

Cherubism is a benign inherited disease of the maxilla and mandible, usually found in children by 5 years of age *(it affects males 2:1)*. The vast majority of cases occur in the **mandible**. The jaws are firm and hard to palpation and regional lymphadenopathy may be present. There are **no associated** systemic manifestations. The deciduous dentition may be spontaneously shed prematurely, beginning as early as three years of age. There is often **delayed eruption** of the permanent dentition which is often defective with the absence of numerous teeth and displacement of those present.

Radiographically, the lesions characteristically appear as multiple, well-defined, **multilocular radiolucencies** of the jaw.

Histologically, the lesions bear a close resemblance to those seen in **central giant cell granulomas.** The histology shows a giant cell lesion with some reactive bone formation. However, **perivascular collagen cuffing** is regarded as pathognomonic for cherubism.

The tumors tend to cease growing shortly **after puberty.** With increase in age and size of the patient, the deformity produced becomes less noticeable.

All of the following are dental manifestations of hypoparathyroidism, **except:**

- Delayed eruption of teeth
- Small tooth crowns with short crown / root ratio
- Blunted root apices
- Enamel hypoplasia

- **Small tooth crowns with short crown / root ratio**

Hypoparathyroidism results from a deficiency of parathyroid hormone. Without sufficient amounts of parathyroid hormone, **the calcium level in the blood falls** and produces signs and symptoms. A common symptom of a low plasma calcium level is tingling of the extremities. In addition, skeletal muscle twitching may occur. If the plasma calcium level is low enough, skeletal muscle spasms *(tetany)* may occur. Hypoparathyroidism may be caused by a genetic disorder, congenital absence of the parathyroid glands, accidental removal or injury of the parathyroid glands during surgery to remove the thyroid gland, massive radiation to the thyroid gland or magnesium deficiency.

Important: The **dental manifestations** of hypoparathyroidism *(i.e., delayed eruption, enamel hypoplasia and blunted root apices)* may be prevented by early treatment with **vitamin D.**

Remember: The term **muscular dystrophy** refers to a group of genetic diseases marked by the progressive weakness and degeneration of the skeletal, or voluntary, muscles, which control movement. **Oral manifestations** include an increase in dental disease if oral hygiene is neglected, weakness in the muscles of mastication leading to decreased maxillary biting force and a higher-incidence of mouth breathing and open bite.

The symptoms of **hyperthyroidism** include all of the following, **except:**

- Nervousness
- Irritability
- Increased perspiration
- Constipation
- Fine brittle hair
- Muscular weakness
- Weight loss despite a good appetite

- **Constipation**

***Constipation is a symptom of **hypothyroidism.**

The term **hyperthyroidism** refers to any condition in which there is too much thyroid hormone *(thyroxin)* in the body. This most commonly results from a generalized overactivity of the entire thyroid gland, a condition also known as **diffuse toxic goiter** or **Graves' disease.** Alternatively, one or more nodules or lumps in the thyroid may become overactive, a condition known as **nodular toxic goiter** or **Plummer's disease.** The primary role of thyroxin is to stimulate cellular metabolism, growth and differentiation of all tissues. In excess, therefore it leads to high basal metabolism, fatigue, weight loss, excitability, elevated temperature and generalized osteoporosis. Oral manifestations are not too remarkable, but if the disturbance begins in the early years of life, the premature eruption of the teeth and the premature loss of the deciduous dentition are common findings.

Comparison of the **two forms** of hyperthyroidism:

- **Graves' disease** → is the **most common** form, occurs most frequently in women under 50. **Exophthalmos** is common. Multi-systems are affected primarily CNS, cardiovascular and musculoskeletal.

- **Plummer's disease** *(nodular toxic goiter)* → affects both genders usually over 50. **Exophthalmos is rare**. Often uni-system. May present with only cardiac disease.

The symptoms of **hypothyroidism** include all of the following, **except:**

- Feeling run down
- Depression
- Increased weight
- Increased perspiration
- Dryness and brittleness of hair
- Dry and itchy skin
- Constipation
- Muscle cramps

• **Increased perspiration**

***Increased perspiration is a symptom of **hyperthyroidism.**

Hypothyroidism refers to a condition in which the amount of thyroid hormone in the body is below normal. This is the **most common** form of thyroid function abnormality, and is far more common than hyperthyroidism. This condition is considerably more common in **women** than in men. The most common cause of hypothyroidism is **Hashimoto's thyroiditis**. The second most common cause is the treatment of hyperthyroidism. Hypothyroidism is characterized by puffiness of the face and eyelids and swelling of the tongue and larynx. The skin becomes dry and rough and the hair becomes sparse. The individual has a **low basal-metabolic rate** and a low body temperature. The affected individuals also have poor muscle tone, low strength and get tired very easily. Mentally they are very sluggish. The treatment of hypothyroidism is straight-forward and consists of administering thyroid hormone *(thyroxin)*.

Severe hypothyroidism in a child is called **cretinism**. Due to a lack of thyroid hormone, there is a retardation of growth and an abnormal development of bones. Mental retardation is caused by the improper development of the CNS. If this condition is recognized early, it can be markedly improved with the use of thyroid hormones. **Note: Extreme hypothyroidism** in adults is called **myxedema**.

Note: Dental findings in a child with hypothyroidism include an **underdeveloped mandible** with an overdeveloped maxilla, **enlarged tongue** which may lead to malocclusion, delayed eruption of teeth and deciduous teeth being retained longer.

All of the following statements concerning **primary hyperparathyroidism** are true, **except:**

- Primary hyperparathyroidism is a metabolic disorder in which one or more of the parathyroid glands produce too much parathyroid hormone
- It can result in the loss of bone tissue
- It is seen twice as often in men than in women
- The exact cause of primary hyperparathyroidism is not known

• **It is seen twice as often in men than in women**

***__This is false__; it is seen twice as often in **women** than in men.

Hyperparathyroidism is a metabolic disorder in which the parathyroid glands produce too much parathyroid hormone. Too much parathyroid hormone causes too much calcium to be released from bone. When one parathyroid gland becomes enlarged, the condition is called **adenoma**. When more than one becomes enlarged, the condition is called **hyperplasia**. The **symptoms include:** loss of appetite, increasing thirst, frequent urination, lethargy and fatigue, muscle weakness, joint pain and constipation. **Important: pathologic fracture** *(due to the marked resorption of bone)* may be the first symptom of the disorder.

Intraorally, there is diffuse bone loss causing malocclusion and shifting of the teeth. **Radiographically**, there are cyst-like radiolucencies *(central giant cell lesions)* found posteriorly in the jaw that have a **"ground glass"** appearance and the **lamina dura** around the teeth may be lost.

Important points:

1. Fibrous dysplasia also demonstrates the typical **"ground-glass"** appearance of bone. ***If this ground-glass appearance is seen, further tests should include a skull radiograph and **blood chemistries** to aid in the diagnosis.
2. Any patient who has a lesion diagnosed as a **central giant cell lesion** should be evaluated medically to rule out the possibility of hyperparathyroidism. If hyperparathroidism is present, the **serumcalcium** will be elevated. Conditions in which central giant cell lesions are found include fibrous dysplasia, the central and peripheral giant cell granuloma and Paget's disease.

Note: In Paget's disease, serum levels of **alkaline phosphatase** will be increased.

All of the following statements concerning **osteomalacia** are true, **except:**

- Osteomalacia means hardening of the bones
- The signs and symptoms include pain in the bones of the arm, legs, spine, and pelvis
- It is the adult form of Rickets
- One of the most common causes is a problem of fat malabsorption called steatorrhea

• **Osteomalacia means hardening of the bones**

***This is false;** osteomalacia means **softening** of the bones.

This softening of the bones occurs because the bones contain osteoid tissue which has failed to calcify due to the **lack of Vitamin D.** Steatorrhea is a condition in which the body is unable to absorb fats, and they are passed directly out of the body in the stool. The result of this problem is that Vitamin D, which is usually absorbed with fat, and calcium are poorly absorbed. All bones are effected, specifically their epiphyseal growth plates. Osteomalacia appears to be more common in women. This condition may be asymptomatic until fracture occurs.

Rickets is osteomalacia in children. It causes skeletal deformities. It is usually accompanied by listlessness, irritability and generalized muscular weakness. A child with rickets may have bowlegs and develop a pigeon breast and a protruding stomach. The **teeth** in a child with rickets are affected as follows: delayed eruption, malocclusion and developmental abnormalities of the dentin and enamel along with a higher caries rate.

Which of the following are **oral manifestations** of patients with **cerebral palsy?**

- They have a higher incidence of periodontal disease, caries, bruxism and malocclusion
- They are prone to gingival hyperplasia if dilantin is used to control seizures
- They are more susceptible to trauma, particulary to the maxillary anterior teeth
- All of the above

• **All of the above**

Cerebral palsy is a term used to describe a group of disorders affecting body movement and muscle coordination. It is due to an insult to or anomaly of the brain's motor control centers. This damage interferes with messages from the brain to the body, and from the body to the brain. The effects vary widely from individual to individual. Cerebral palsy is characterized primarily by spastic paralysis or impairment of control or coordination over voluntary muscles and is often accompanied by mental retardation, seizures and disorders of vision and communication. **Note:** No intraoral anomalies are unique to persons with cerebral palsy. **However,** several conditions *(those listed on the front of the card)* are more common or more severe than in the normal population.

Remember: Down syndrome is a congenital defect caused by a chromosomal abnormality *(trisomy 21)*. It is marked by various degrees of mental retardation and characteristic physical features such as a short, flattened skull, slanting eyes, **a thickened tongue** *(fissured)*, broad hands and feet and other anomalies. **Other oral manifestations include** mandibular prognathism, increased incidence of periodontal disease, delayed eruption of teeth, higher incidence of congenitally missing teeth, malocclusion, and enamel dysplasia.

All of the following statements concerning **cystic fibrosis** are true, **except:**

- It is a hereditary disease of the exocrine glands, primarily affecting the GI and respiratory systems
- It is the most common inherited disease leading to death among white people in the United States
- It is more common in boys than girls
- It is usually characterized by COPD, exocrine pancreatic insufficiency, and abnormally high sweat electrolytes
- A high percentage of children with cystic fibrosis have teeth that are dark in color, ranging from yellowish gray to dark brown

- **It is more common in boys than girls**

***This is false; it is equally common** in boys and girls.

Cystic fibrosis is a congenital metabolic disorder that causes the exocrine glands *(which are glands that secrete fluids into a duct)* to **produce abnormal secretions**, resulting in several symptoms, the most important of which affect the digestive tract and the lungs. In some glands, such as the pancreas and those in the intestines, the secretions are thick or solid *(an excessively* **viscous mucous***)* and may block the gland completely. The mucous-producing glands in the airways of the lungs produce abnormal secretions that clog the airways and allow bacteria to multiply. The sweat glands secrete fluids that have a high sodium and chloride content. **Note:** The staining of the teeth is most likely due to the fact that patients with cystic fibrosis are usually subjected to large amounts of **tetracyclines** during childhood.

Symptoms of CF include:
- Poor growth despite good appetite
- Malabsorption and foul, bulky stools → steatorrhea
- Chronic bronchitis *(COPD)* with cough
- Recurrent pneumonia → respiratory infections
- Clubbing of fingers and toes
- Barrel-chested appearance

Important: There is a significantly **reduced caries rate** in patients with cystic fibrosis. This is **probably** the result of alterations in saliva and the long-term use of antibiotics.

Which form of hepatitis is caused by an RNA enterovirus and is usually transmitted by the **fecal-oral route?**

- Hepatitis A
- Hepatitis B

• **Hepatitis A**

***Hepatitis A is also known as **viral** or **infectious** hepatitis.

Hepatitis A is an infectious disease of the liver. It most often occurs in young adults and is prevalent in areas with inadequate sewerage. Shellfish from contaminated waters is also a prime source of Hepatitis A. The initial symptoms of viral Hepatitis appear after an **incubation period of 3-6 weeks**. They include fever, abdominal pain, and nausea, followed by jaundice.

The damage to liver cells results in increased serum levels of enzymes, such as transaminases, normally active in liver cells. The detection of the increased serum levels of these enzymes is used in diagnosing this disease. In most cases of Hepatitis A, the infection is **self-limiting** and recovery occurs within 4 months.

Notes:

1. The presence of **surface antigen** *(A or B)* in a patient's serum indicates that the patient is potentially infectious for Hepatitis *(known as the* **carrier state***).*

2. These Hepatitis viruses are **very heat-resistant** *(more so than the AIDS virus).* However, autoclaving properly **will kill** them.

Which type of hepatitis is also known as **"Serum Hepatitis"?**

- Hepatitis A
- Hepatitis B

• **Hepatitis B**

Hepatitis B is an infectious disease of the liver that produces liver inflammation and necrosis. It is caused by a **DNA virus.** The principal means of transmission involves exposure to **contaminated blood or serum**. There is a high rate of transmission among drug addicts, who frequently use contaminated needles. Hepatitis B also can be transmitted sexually or by blood transfusions.

The signs and symptoms are similar to Hepatitis A *(fever, abdominal pain, nausea, etc.)* but there is a **longer incubation period** *(2-3 months)*. The symptoms are slower in developing but are of a **longer duration.** Most patients recover fully, however, some develop chronic liver disease.

Important: Hepatitis B transmission is of major concern to members of the dental profession. They have **at least three times higher risk** than the general population of acquiring this virus. **Note:** For this reason **always use universal precautions.**

HB vaccine is recommended for all health care personnel. A series of **three doses** is required; the second and third doses are given **one and six months** after the first, respectively. The injection is given IM, preferably in the deltoid.

Which of the following is one of the most important substances that **influences** the rate of healing of wounds in the oral cavity?

- Fat
- Protein
- Iron
- Calcium

- **Protein**

***Nutritional factors** such as the amount of **protein** a patient is consuming is one of the most important factors which may effect the speed of wound healing. Hypoproteinemia has been shown to delay wound healing, while having a high protein diet has been shown to accelerate wound healing. **Vitamins**, especially Vitamin C, have been shown to be important in proper wound healing.

Other factors that influence the rate of healing:

- **Location of the wound** → wounds in an area in which there is a good vascular bed heal considerably more rapidly than wounds in an area which is relatively avascular.
- **Physical factors** → severe trauma to tissue is a deterrent to rapid wound healing. The local temperature in the area of a wound influences the rate of healing. In environmental hyperthermia, wound healing is accelerated; while in hypothermia, healing is delayed.
- **Circulatory factors** → anemia and dehydration have been found to delay the healing of wounds.
- **Age of patient** → wounds in younger persons heal considerably more rapidly than wounds in elderly persons.
- **Infection** → bacterial invasion will retard healing.
- **Hormonal factors** → **ACTH and cortisone** are substances that have been known to interfere with the healing of wounds. **Diabetes mellitus** *(insulin deficiency)* is one of the most widely recognized diseases in which there is significant, clinically evident, retardation in repair of wounds.

Which of the following terms refers to a **small section** of tissue that is removed for examination?

- Excisional biopsy
- Incisional biopsy
- Neither of the above

• **Incisional biopsy** *(also called diagnostic biopsy)*

This is done when lesions are **too large to excise** initially without having established a diagnosis or are of such a nature that excision would be inadvisable.

The **total excision** of a small lesion for microscopic study is called excisional biopsy. This is preferred if the size of the lesion is such that it may be removed along with a margin of normal tissue and the wound can be closed primarily. **Example:** A 1-cm exophytic mass *(which is a lesion that grows outward from an epithelial surface)* on the cheek.

Notes:

1. **Biopsy** is the most reliable technique to diagnose soft tissue lesions.
2. The **scalpel** is the instrument of choice since it cleanly removes the tissue and does not dehydrate it as cautery or the high-frequency cutting knife may.
3. The fixative of choice is **10% formalin.**
4. The rationale for surgical removal and biopsy of a large periapical lesion suspected to be of inflammatory origin is that a clinical diagnosis can be confirmed microscopically.

Remember: This is the only way to distinguish between a granuloma and a cyst.

Infectious mononucleosis is caused by the:

- Pox virus
- Epstein-Barr virus
- Rubella virus
- Paramyxovirus

• **The Epstein-Barr virus**

The Epstein-Barr virus is a member of the **herpesvirus** group. It causes infectious mononucleosis and has been associated with the subsequent development of two forms of cancer: **Burkitt's lymphoma** and **nasopharyngeal carcinoma.**

Notes:

1. There are no specific oral manifestations of infectious mononucleosis, although secondary lesions do occur.
2. Neck swellings are characteristic of infectious mononucleosis, Hodgkin's disease and tuberculosis.

Remember:

- **Rubella viruses** cause German measles *(rubella)*, which present with a characteristic rash *(flat, pink spots on the face which spreads to other body parts).* Oral manifestations of rubella **may include** swollen and congested tonsils and red macules.
- **Paramyxoviruses** cause measles *(rubeola)* and mumps. Rubeola is characterized by the formation of **Koplik spots** in the oral cavity. These spots are small bluish-white lesions which are surrounded by a red ring. They cannot be wiped off and occur opposite the molars. **Mumps** causes the enlargement of the parotid glands. Serious complications include **deafness** in children and **orchitis** *(which is the inflammation of the testis)* in males past puberty.

All of the following are characterized by granulomatous inflammation, **except:**

- Sarcoidosis
- Tuberculosis
- Pneumonia
- Histoplasmosis
- Tertiary syphilis

- **Pneumonia**

***Pneumonia is charactized by **acute inflammation** in which there is purulent exudate in response to bacterial infection.

Granulomatous inflammation is a subtype of chronic inflammation characterized morphologically by **granulomas**.

Granulomatous infections include:
- Mycobacterial diseases → tuberculosis and leprosy
- Fungal diseases → histoplasmosis, blastomycosis, and coccidioidomycosis
- Parasitic diseases → schistosomiasis
- Syphilis and cat-scratch disease
- Silicosis and berylliosis are granulomatous lung diseases produce by the occupational exposure to dusts
- Sarcoidosis is a chronic disorder of unknown etiology characterized by the formation of tubercles of non-necrotizing epithelioid tissue

***Granulomas are nodules of **epithelioid cells** *(which is a modified macrophage with abundant eosinophilic cytoplasm)* usually 2 mm or less in diameter. Lymphocytes, plasma cells, and fibroblasts surround the nodule of epithelioid cells.

Ludwig's angina refers to cellulitis of the floor of the mouth in which the infection spreads to the:

- Submental space
- Sublingual space
- Submandibular space
- All of the above

• **All of the above**

Ludwig's angina often results from an odontogenic infection. As a result, the bacteriology of theses infections generally involves oral flora, particularly anaerobes. Other recognized etiologies of Ludwig's angina include poor oral hygiene, IV drug abuse, trauma, and tonsillitis.

It is characterized by:
• Rapid onset
• Brawny induration *(tissues are board-like)* and no fluctuance is present
• The three facial spaces are involved **bilaterally**
• Typical **"openmouthed"** appearance
• Drooling, dyspnea, trismus, and fever

Airway management, massive antibiotic coverage *(IV),* and surgical incision and drainage are the mainstays of treatment.

Very Important: The **most serious complication** of Ludwig's angina is **edema of the glottis** *(which is a slit-like opening between the true vocal cords)*

The **erythrocyte sedimentation rate** *(ESR)* is a nonspecific test that monitors:

- Bleeding rate
- The progression of disease
- Prothrombin time
- Platelet function

• **The progression of disease**

The ESR is the rate at which red blood cells settle out in a tube of unclotted blood, expressed in millimeters per hour. Blood is collected in an anticoagulant and allowed to sediment in a calibrated glass column. At the end of one hour, the lab technician measures the distance the erythrocytes have fallen in the tube. Elevated sedimentation rates are not specific for any disorder but **indicate the presence of inflammation.** Inflammation causes an alteration of the blood proteins which makes the red blood cells aggregate, becoming heavier than normal. The speed with which they fall to the bottom of the tube corresponds to the degree of inflammation.

ESR rises during:
• Inflammation
• Tissue degeneration
• Suppuration
• Necrosis

Note: Certain non-inflammatory conditions, such as pregnancy, are also categorized by high sedimentation rates.

Which of the following are symptoms of **acute fluoride** poisoning?

- Nausea and abdominal pain
- Excessive salivation
- Vomiting
- Diarrhea
- Convulsions
- Hypotension
- All of the above

• **All of the above**

The treatment for acute fluoride poisoning includes: 1) Call poison control center, 2) Monitor vital signs, 3) Initiate basic life support as needed and 4) Get patient to the hospital.

Fluoride poisoning may be acute *(caused by a single large dose of fluoride)* or chronic *(caused by long-term ingestion of fluoride)*. The characterisitic signs of chronic fluoride poisoning are:

1. **Osteosclerosis** of the bones → which results from long-term ingestion of water with 10 to 25 ppm of fluoride.
2. **Dental fluorosis** *(enamel hypoplasia)* → which is due to fluoride intake during the **calcification stage** of tooth development. This can occur in permanent and deciduous teeth.

Note: It has been estimated that the average American diet contains about 0.2 to 0.3 mg of fluoride per day. If 1 ppm of fluoride is added to the drinking water, about 1 to 2 mg of fluoride will be added to the diet daily. Balance studies have shown that when quantities of fluoride ingested **do not exceed 4 to 5 mg daily, little is retained by the body**. The finding indicates the safety of the preventive dentistry programs based on the addition of fluoride to drinking water in concentrations of approximately 1 ppm. **Sodium sillicofluoride** is the type of fluoride often used for fluoridation of the communal water supply.

Important: Fluoride normally accumulates slowly in bones as a person ages. **However**, if ingested in very high amounts, it accumulates rapidly. The intake of calcium in high doses will reduce the absorption of dietary fluoride.

Xerostomia is caused by:

- Sialadenitis
- Sjögren's syndrome
- Medications
- Cancer therapy
- Nerve damage
- Conditions such as Alzheimer's disease or stroke, bone marrow transplants, endocrine disorders, stress, anxiety, depression and nutritional deficiencies
- All of the above

• **All of the above**

Xerostomia *(dry mouth)* is not a disease, however, it can be a symptom of certain diseases. Many times xerostomia is caused by failure of the salivary glands to function normally, but the sensation can also occur in people with normal salivary glands.

Xerostomia can cause health problems by affecting nutrition, as well as psychological health. At its most extreme, it can lead to **rampant tooth decay** and **periodontal disease.**

Notes:

1. **Sialadenitis** is an insidious inflammatory disease of the major salivary glands

2. **Anticholinergic** drugs are well know to cause xerostomia, these include atropine and scopolamine

3. **Antipsychotic** drugs also cause xerostomia, these include the phenothiazines, such as chlorpromazine and prochlorperazine

All of the following statements concerning **malignant melanoma** are true, **except:**

- It is a relatively benign form of skin cancer
- Currently about 1 in 100 persons in the United States can expect to develop this cancer in a lifetime
- Without treatment it has the tendency to become widely metastatic and result in the demise of the patient
- There are four general clinical types of melanoma: superficial spreading, nodular, acrolentiginous, and lentigo maligna

• **It is a relatively benign form of skin cancer**

***This is false;** it is a **serious form** of skin cancer

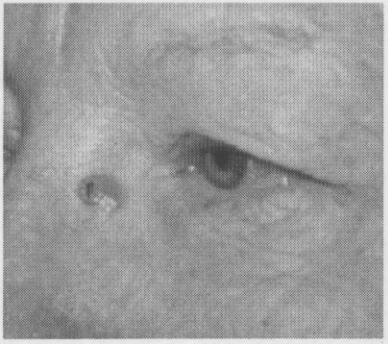
Malignant melanoma is a serious skin cancer in which the tanning cells in the skin that produce a dark-colored substance called "melanin" undergo uncontrolled growth. Melanoma may suddenly appear without warning, but can often develop from or near or a mole. It can occur anywhere on the skin. **See picture #53 in booklet.**

Excessive exposure to UV radiation from the sun may be the primary cause of melanoma. Malignant melanoma has been linked to both a lot of sun exposure over a lifetime and to painful sunburns during childhood.

Malignant melanoma is an **uncommon neoplasm of the oral mucosa.** It exhibits a definite predilection for **the palate and the maxillary gingiva/alveolar ridge.** Unfortunately, oral mucosal melanomas have a dismal prognosis.

Note: A **nevus** is a mole. Almost all moles are normal. **Atypical** *(dysplastic)* **nevi** are unusual moles that are generally larger than normal moles and are either flat or have a flat part. They have irregular borders and often are variable shades of color, particularly brown. The presence of dysplastic nevi may mark a greater risk of malignant melanoma developing on apparently normal skin. **See picture #52 in booklet.**

Name the malignant, epithelial cell tumor that characteristically begins as a papule and enlarges peripherally, developing a central crater that erodes, crusts and bleeds. **An example of this tumor is shown below.**



• **Basal Cell Carcinoma**

Metastasis is rare, but the local invasion by direct extension destroys the underlying and adjacent tissue. It frequently develops on the exposed surfaces of the skin, face and scalp in middle-aged or elderly persons. The primary cause of the cancer is excessive **exposure of the sun or to x-rays.** The treatment for basal cell carcinoma is eradication of the lesion, often by electrodessication or cryotherapy.

A malignancy of **plasma cells** is called:

- Histoplasmosis
- Multiple myeloma
- Osteomalacia
- Amyloidosis

- **Multiple myeloma** → also known as "Plasma Cell Myeloma"

Multiple myeloma is a primary malignant neoplasm of bone characterized by progressive destruction of the marrow with replacement by neoplastic plasma cells.

Clinical Features:

- **Men 2:1** → 40-70 years old. Vertebrae, ribs, and skull are most frequently involved; **pain in lumbar or thoracic region** is a common early symptom
- Jaws are rarely a primary site, but become involved in 70% of the cases, **molar-ramus area most common site.** Symptoms include swelling, pain, loosening of the teeth, and paresthesia

Radiographic features → variable; slight demineralization to extensive destruction, characteristic finding is multiple, small, discreet **"punched out"** radiolucencies in involved bones. In a patient suspected of having multiple myeloma, a **lateral skull** radiograph is best to confirm the diagnosis.

Laboratory findings → important in establishing diagnosis; **hypergammaglobulinemia** *(especially* **IgG** *in 70% of the cases).* **Bence Jones proteinuria** is present in 60-85% of the cases.

Treatment and Prognosis → chemotherapy, radiation; **prognosis poor** with median survival time 2-3 years.

Which of the following types of skin cancer is considered to be the **most severe?**

- Basal cell carcinoma
- Squamous cell carcinoma
- Malignant melanoma

• **Malignant melanoma**

It is a potentially serious skin cancer in which the tanning cells *(melanocytes)* in the skin that produce a dark-colored substance called melanin undergo uncontrolled growth. Melanoma may suddenly appear without warning, but can often develop from or near a mole *(nevus)*. It is fairly common in white people and can occur anywhere on the skin. Excessive exposure to UV radiation from the **sun** may be the primary cause of melanoma.

Four general clinical types of melanoma:

1. **Superficial spreading** → **most common form** *(65%)*, **radial growth** phase predominates
2. **Nodular** → much less common, **vertical growth phase predominates**
3. **Acrolentiginous** → occurs on the hands and feet
4. **Lentigo maligna** → more common among the elderly population

Notes:

1. Melanoma exhibits either **"radial"** *(horizontal)* or **"vertical"** growth phases within the skin.

2. Skin cancer is the **most common** malignancy in the United States.

When using the **TNM** method in assessing the prognosis and therapy of malignant neoplasms, the **N** represents:

- The presence of Nikolsky's sign
- The presence of nodules
- The presence of regional lymph node involvement
- The presence of erythroplakia

• **The presence of regional lymph node involvement**

Clinical staging of malignant neoplasms:

The **TNM** method of assessing the prognosis and therapy of malignant neoplasms is based upon **1)** the size of the primary tumor, **2)** the presence of regional lymph node involvement and **3)** the presence of distant metastases. This is represented as:

- **T** = size of the primary tumor
- **N** = presence of regional lymph node involvement
- **M** = presence of distant metastases

Which location of **squamous cell carcinoma** has been etiologically related to race, complexion and sunlight?

- Lip
- Tongue
- Floor of the mouth
- Gingiva and alveolar mucosa
- Buccal mucosa

• **Lip**

***Squamous cell carcinoma *(also called Epidermoid carcinoma)* is the **most common malignancy in the oral cavity.**

Location	Incidence	Etiology	Clinical Characteristics	Treatment
Lip	**Most common site**	Race, complexion, sunlight, pipe smoking	Men over 60, **vermillion of lower lip**, painless ulcer, keratotic plaque	Surgery or irradiation
Tongue *(lateral border and ventral surface)*	Most common **intraoral** site	Tobacco, alcohol, syphilis, Plummer-Vinson syndrome	Men over 60, posterior lateral border and middle third, painless ulcer, leukoplakia, erythroplakia	Surgery or irradiation
Floor of the mouth	Second most common intraoral site	Tobacco, alcohol	Men 40-60 years old, painless ulcer, leukoplakia, erthroplakia	Surgery or irradiation
Buccal mucosa	10% of all oral carcinomas	Tobacco, alcohol, denture irritation	Painless ulcer, exophytic mass, leukoplakia	Surgery or irradiation
Gingiva and alveolar mucosa	10-15% of all oral carcinomas	Tobacco, alcohol	Men over 60, mandibular mucosa, painless ulcer, plaque-like or exophytic or exophytic mass	Surgery

The most common form of **malignant melanoma** is:

- Superficial spreading
- Nodular
- Acrolentiginous
- Lentigo maligna

• **Superficial spreading**

Superficial spreading melanoma is the most common cutaneous melanoma in Caucasians. The lesion presents as a tan, brown, black or admixed lesion on sun exposed skin, especially the back. The cancer presumably begins at one focus in the skin at the dermo-epidermal junction. It initially grows in a **horizontal plane,** along, just above and below the dermo-epidermal junction. This is referred to as the **"radial"** growth phase of melanoma and is clinically macular or only slightly elevated. The **"vertical"** growth phase is characterized by an increase in size, a change in color, nodularity and, at times, ulceration.

Nodular melanoma is much less common and accounts for approximately 13% of cutaneous melanomas. In this tumor, there is presumably no **"radial"** growth phase *(it exists solely in the "vertical" growth phase).* It presents as a sharply delineated nodule with degrees of pigmentation. They may be pink *(amelanotic melanoma)* or black. They have a predilection for occurrence on the back, head and neck of men.

Lentigo maligna melanoma accounts for approximately 10% of cutaneous melanomas. It is more common among the elderly population. The lesion may grow for years in the "radial" growth phase before developing into the more aggressive "vertical" growth phase. This "radial" growth phase is known as lentigo maligna or melanotic freckle of Hutchinson. The "vertical" growth phase is known as lentigo maligna melanoma.

Acrolentiginous melanoma occurs on the hands and feet. It has a reputation for being ignored by the patient resulting in the development of metastatic disease.

Of the following types of squamous cell carcinomas, which is the **least common?**

- Squamous cell carcinoma of the palate
- Squamous cell carcinoma of the nasopharynx
- Squamous cell carcinoma of the oropharynx
- Squamous cell carcinoma of the maxillary sinus

- **Squamous cell carcinoma of the nasopharynx**

Location	Incidence	Etiology	Clinical Characteristics	Treatment
Nasopharynx	Less than 2% of all cancers in the U.S.	Tobacco, alcohol	Men between 30 and 40, roof or lateral wall is most common site, cervical mass, earache sore throat, nasal obstruction	Surgery and Irradiation
Palate	10% of all oral carcinomas	Tobacco, alcohol, denture irritation	Men over 60, soft palate is more common than hard palate, painful ulcer, leukoplakia, exophytic mass	Surgery and irradiation
Oropharynx	10% of all head and neck cancers	Tobacco, alcohol	Men over 50, sore throat, dysphagia, painful ulcer, cervical mass	Surgery and irradiation
Maxillary sinus	30% of all head and neck cancers	Unknown	Men over 40, chronic sinusitis, **bulging of palate,** loosening of teeth, paresthesia in cheek	Surgery and irradiation

The **initial growth** of a melanoma just above and below the dermo-epidermal junction is referred to as the:

- "Radial" growth phase
- "Vertical" growth phase

• **"Radial" growth phase**

***This refers to the initial growth of a melanoma in a **horizontal plane.** It is clinically macular or only slightly elevated.

The **"vertical"** growth phase is the phase that begins when neoplastic cells populate the underlying dermis. It is characterized clinically by an increase in size, a change in color, nodularity and, at times, ulceration. **Metastasis** is possible once the melanoma reaches this phase.

Remember: Malignant melanoma is an uncommon neoplasm of the oral mucosa. It exhibits a definite predilection for the palate and the maxillary gingiva / alveolar ridge. Unfortunately, oral mucosa melanomas have a dismal prognosis. The five-year survival rate for such tumors is approximately 7%.

The most common site for carcinoma of the **tongue** is the:

- Dorsum
- Tip
- Lateral border
- Sulcus terminalis

• **Lateral border**

Cancer of the **tongue** causes more deaths than do malignant lesions in other regions of the head and neck. The reasons for this are 1) The tongue is the **most frequent site for intraoral cancer** 2) It is a highly mobile organ that is **richly endowed with lymphatics and blood vessels** which facilitate metastases. It very rarely gives rise to skeletal metastasis.

Notes:
1. The **dorsum** of the tongue is the area least frequently involved
2. Squamous cell carcinoma of the tongue commonly **metastasizes** to the cervical lymph nodes.

Cancer of the lips → cancer is more common here than **intraorally**, most of which *(95%)* are found on the **lower lip**. 90 to 98% of cancers of the lower lip occur in males. Chronic exposure to the sun and **pipe smoking** have been implicated in the etiology.

Cancer of the floor of the mouth → occurs most commonly in the **anterior segment** on either side of the midline, near the orifices of the salivary glands. Premalignant lesions of squamous epithelium most often occur here. **Note:** Prognosis is **very poor** for lesions found here.

Cancer of the buccal mucosa → generally occurs along the **plane of occlusion,** midway anteroposteriorly.

Cancer of the gingiva → is more common in the mandible than in the maxilla, and posterior sites are seen more frequently than anterior.

Which neoplasm has strong evidence of a **viral etiology?**

- Ewing's sarcoma
- Burkitt's lymphoma
- Liposarcoma
- Osteoid osteoma

• **Burkitt's Lymphoma**

***Also called **African Jaw Lymphoma.**

Burkitt's lymphoma is a high-grade, **non-Hodgkin's lymphoma** that is endemic in Africa and occurs only sporadically in North America. It is manifested most often as a large osteolytic lesion in the jaw *(African form)* or as an abdominal mass *(Non-African form)*.

Burkitt's lymphoma is the first human cancer with **strong evidence** of a viral etiology. The Epstein-Barr virus *(a herpes-type virus)* has been isolated from cultures of tumor cells and patients with Burkitt's lymphoma have high titers of antibodies against EBV. Also, an antibody against a surface antigen on the tumor cells has been demonstrated. **Note:** The Epstein-Barr virus is also associated with **infectious mononucleosis** and **oral hairy leukoplakia.**

Two forms of Burkitt's lymphoma:

1. **African** → younger *(mean age 3)*, male predominance, typically involves the jaws.
2. **Non-African** → older *(mean age 11)*, no sex predilection, presents most often as an **abdominal mass.**

Note: The jaw lesions usually present as expanding intraoral masses with mobility of the involved teeth. **Radiographically**, there is a **moth-eaten**, poorly marginated destruction of bone.

A malignant tumor developing from bone marrow, usually in long bones or the pelvis of **adolescent boys** is called:

- Myeloma
- Ewing's sarcoma
- Osteogenic sarcoma
- Multiple myeloma

• **Ewing's sarcoma**

Ewing's sarcoma is an uncommon, highly lethal, malignant neoplasm of bone of uncertain origin. The most common sites for Ewing's sarcoma are the pelvis, the thigh, and the trunk of the body. The peak ages are between 10 and 20.

Pain, usually of an intermittent nature, and **swelling** of the involved bone are often the earliest clinical signs and symptoms of Ewing's sarcoma. Fever and leukocytosis are also present. **Histologically**, it is often difficult to distinguish this tumor from a neuroblastoma or a reticulum cell sarcoma, however, the cells of Ewing's sarcoma contain **glycogen**.

When the jaws are involved, there is predilection for the **ramus of the mandible**. There is usually pain followed by rapid swelling and loosening of the teeth.

Radiographically, the most characteristic appearance is that of a **moth-eaten, destructive radiolucency of medulla and erosion of the cortex with expansion.** A variable periosteal **"onion-skin"** reaction may also be seen.

Important: The most common osseous malignancies are **osteosarcomas**, followed by chondrosarcomas, fibrosarcomas and Ewing's sarcoma.

Which of the following bone lesions is **most likely** to be fatal?

- Osteochondroma
- Paget's disease
- Giant cell tumor
- Multiple myeloma
- Odontogenic myxoma

- **Multiple myeloma**

Multiple myeloma is characterized by an elevated blood level of **Bence Jones protein** and multiple radiolucent areas in the mandible and skull. It is a malignant neoplasm of the bone marrow. The tumor is composed mostly of **plasma cells,** which destroy osseous tissues. Patients are past the age of 45, and males are affected twice as frequently as females.
See picture #61 in booklet.

Remember:
- An **osteochondroma** is a benign tumor of bone and cartilage.
- **Paget's disease** is a common, nonmetabolic disorder of bone of unknown cause, usually affecting middle-aged and elderly people, characterized by excessive bone destruction and unorganized bone repair. It is treated with a high protein and high calcium diet.
- The **giant cell tumor** is a bone tumor of multinucleated **giant cells** that resembles osteoclasts scattered in a matrix of spindle cells. Myelomas of this kind may be benign or malignant and may cause pain, functional disability, and in some cases, pathologic fracture.
- The **odontogenic myxoma** is a rare tumor of the jaw. It is most often seen in the mandible.The patients are usually under 35 years of age. It is slow growing and usually asymptomatic, but eventually leads to localized expansion of the jaw. The treatment is curettage.

In the **African form** of Burkitt's lymphoma:

- The patients are generally older than in the non-African form
- The patients are generally younger than in the non-African form
- There is no gender predilection
- It presents most often as an abdominal mass

• **The patients are generally younger than in the non-African form**

Burkitt's lymphoma is a high-grade **non-Hodgkin's lymphoma** that is endemic in Africa and occurs only sporadically in North America. There are significant differences between the African and non-African forms.

In the **African form,** the patients are **younger** *(between 3 and 8 years of age),* and there is a 2 to 1 **male predominance.** It typically involves the **mandible, maxilla** and abdomen, with extranodal involvement of retroperitoneum, kidneys, liver, ovaries and endocrine glands.

In the **non-African form,** the patients generally are **older** *(between 9 and 12 years of age),* and there is **no gender predilection.** It presents most often as an **abdominal mass** involving the mesenteric lymph nodes or Peyer patches in the ileocecal region, often with an intestinal obstruction. Involvement of the gonads, retroperitoneum and other viscera is **less common.**

Note: Both forms are **histologically identical.** The **Epstein-Barr virus** has been implicated in this cancer.

All of the following statements concerning an **osteosarcoma** are true, **except:**

- It is a malignant bone tumor composed of anaplastic cells derived from mesenchyme
- Its peak incidence is after epiphyseal fusion, particularly the years between 50 and 60
- This tumor seems to arise in the long bones which show the greatest longitudinal growth
- Joint involvement is rare

- **Its peak incidence is after epiphyseal fusion, particularly the years between 50 and 60**

***This is false;** its peak incidence is **before** epiphyseal fusion, particularly the years between 10 and 25. **Note:** There is a later peak which is associated with **Paget's disease**, chronic osteo-myelitis and previous radiotherapy.

The histologic classification of an osteosarcoma is based on the **dominant type of tissue cell:**

- **Osteoblastic** → produces osteoid tissue
- **Chondroblastic** → produces cartilage tissue
- **Fibroblastic** → produces fibrous tissue

***A very important early radiographic feature of an **osteosarcoma of the jaw** is a **symmetrically widened PDL space** around one or more teeth.

Other **radiographic** features, depending on the degree of calcification:

- Sclerotic → excessive production of bone, **"sun-ray appearance"**
- Lytic → irregular radiolucency
- Mixed → most osteosarcomas **have this appearance**

Important: For **osteosarcoma of the jaw,** the differential diagnosis should include

- Chrondrosarcoma
- Metastatic carcinoma and Pindborg tumor
- Ossifying subperiosteal hemangioma
- Peripheral odontogenic fibroma
- Scleroderma
- Chronic osteomyelitis

Kaposi's sarcoma is an oral manifestation of:

- Diabetes
- Hypothyroidism
- AIDS
- Lichen planus

• **AIDS**

Kaposi's sarcoma is a malignant neoplasm originating in the skin. It is characterized by **abnormal vascular proliferation** *(it is a cancer of the lining of blood vessels)*. It occurs on multiple sites, especially the lower extremities. Initial lesions are small, red papules, which enlarge and fuse to form purple-to-brown, spongy nodules. It spreads to lymph nodes and internal organs. It is most commonly associated with AIDS.

Note: Intraorally, the **hard palate** is the most common location, followed by the gingiva and buccal mucosa.

Remember: AIDS is caused by the RNA retrovirus, **HIV** *(also called HTLV-III)*. The HIV infection is acquired by sexual contact *(homosexual and heterosexual)* or from contaminated blood products.

A **poorly differentiated** squamous cell carcinoma involving lymphoid tissue in the region of the tonsils and nasopharynx is called:

- A lipoma
- A lymphoepithelioma
- A lymphoma
- A nasopharynioma

• **A lymphoepithelioma**

It has a high frequency among young adults of East Asian extraction. The primary lesion is usually very small, often completely hidden. **Swelling of the lymph node** is the most common presenting symptom, followed by a sore throat, nasal obstruction, bloody nose and headache.

The **lymphoepithelioma** is composed of cells *(squamous or undifferentiated),* with a slight to moderate amount of fibrous stroma that contains numerous lymphocytes. **Most importantly,** this neoplasm shows **metastasis at an early stage** to the cervical lymph nodes.

The treatment of choice is x-ray radiation, **however**, the complicating factor lies in the relative inability to treat the widespread metastases in the various organs. The prognosis is poor, with a 30% five-year survival rate.

All of the following statements concerning **metastatic tumors of the jaws** are true, **except:**

- They may be completely asymptomatic
- The patient is usually aware of slight discomfort or pain
- The maxilla is affected far more frequently than the mandible
- The molar region is predominantly involved

• **The maxilla is affected far more frequently than the mandible**

***This is false;** the mandible is affected far more frequently than the maxilla.

The most common malignancy affecting skeletal bones is **metastatic carcinoma**. However, metastatic disease to the mandible and maxilla is unusal *(only about 1%)*. **Most Importantly,** a tumor of the jaws may be the first evidence of dissemination of a known tumor from its primary site.

Note: Metastases to the jaws most commonly originate from primary carcinomas of the **breast**, kidney, lung, colon, prostate and thyroid.

Clinical features of **metastatic** jaw lesions:

- They may be completely asymptomatic
- Usually, however, there is paresthesia or anesthesia of the lip or chin due to involvement of the mandibular nerve
- Teeth in the area are loose or extruded
- There can be swelling or expansion of the jaw
- Appears as an asymptomatic radiolucency

Explain what is meant by a carcinoma of the oral cavity having the following **TNM** designation:

T2, N1, M0

Clinical Staging of Carcinoma of the Oral Cavity

- **T = Size** of the primary tumor
  - TX: Primary tumor can not be assessed
  - TO: No evidence of tumor
  - Tis: Carcinoma in situ
  - T1: less than 2 cm in greatest diameter
  - **T2: 4 cm in greatest diameter**
  - T3: greater than 4 cm in greatest diameter
- **N =** Regional **lymph node** involvement
  - NX: Regional lymph nodes can not be assessed
  - NO: No clinically palpable lymph nodes, or lymph nodes palpable but metastases not suspected
  - **N1: Palpable homolateral lymph node(s), not fixed but metastases suspected**
  - N2: Palpable centralateral/bilateral lymph node(s), not fixed but metastases suspected
  - N3: Palpable lymph node(s), fixed metastases suspected
- **M = Distant** metastasis
  - MX: Presence of distant metastasis can not be assessed
  - **MO: No distant metastasis**
  - M1: Clinical and / or radiographic evidence of metastasis other than regional lymph nodes

The **most common primary** malignant tumor of bone is:

- An osteosarcoma
- A chondrosarcoma
- Ewing's sarcoma
- A myeloma

• **An osteosarcoma**

*** Also called an **Osteogenic Sarcoma.**

An osteosarcoma is a malignant bone tumor composed of anaplastic cells derived from mesenchyme. Osteosarcomas can be classified by site of origin into **1)** The conventional type, arising within the medullary cavity; **2)** The juxtacortical tumors, arising from the periosteal surface; and, **3)** The extraskeletal osteosarcomas, arising in soft tissue.

Common presenting symptoms of an **osteosarcoma of the jaws:**
- Tumor, swelling or mass
- Pain
- Loose teeth
- Paresthesia
- Bleeding

Notes:
1. The most common malignancy found in bone is **metastatic carcinoma.**

2. Metastatic carcinoma to the jaws is **least likely** to have originated in the brain.

All of the following statements concerning **verrucous carcinoma** are true, **except:**

- It is a rare form of squamous cell carcinoma that occurs in either the oral or laryngeal cavity
- There is a characteristic whitish, cauliflower, or coral appearance to the mass
- Typically, the lesion develops on the vocal cords of an elderly male who has been a heavy cigarette smoker
- It is known for its slow growth pattern and well-developed hyperkeratotic epithelial boundaries
- It usually shows rapid metastasis

- **It usually shows rapid metastasis**

***This is false; it **does not** usually metastasize.

A **verrucous carcinoma** is a well-differentiated squamous cell neoplasm of soft tissue of the oral or laryngeal cavity. The lesion may invade or infiltrate the borders of adjacent structures **but it rarely metastasizes**. Verrucous carcinoma may transform into an invasive form of carcinoma or coexist with other squamous cell carcinomas. It is often misdiagnosed histologically as a benign lesion.

Verrucous carcinoma of the **oral cavity:**
- **Etiology:** Tobacco chewing, snuff dipping
- **Clinical:** Men 60+ years; mandibular mucobuccal fold, alveolar mucosa and palate; whitish, cauliflower or coral papillary mass
- **Pathology:** Very well-differentiated
- **Treatment:** Surgery
- **Prognosis:** 60-70% 5-year survival rate

See picture #13 in booklet.

ORAL PATHOLOGY

Which type of cancer occurs most frequently in the **oral cavity?**

- Basal cell carcinoma
- Squamous cell carcinoma
- Angiosarcoma
- Pindborg tumor

• **Squamous cell carcinoma**

It is a malignant **epithelial** tumor. It is the **most common type of oral cancer**, accounting for over 90% of all malignant neoplasms of the oral cavity. It is two times more prevalent in males *(40-65 years of age)*. **See pictures #49, 50 and 51 in booklet.**

It is more common on the **lips** *(lower lip)* than intraorally. The most common site for **intraoral** squamous cell carcinoma is the **lateral border and ventral surface of the tongue** *(from this site, SCC often metastasizes to the cervical lymph nodes)*. The floor of the mouth is the second most common site for intraoral carcinoma and it has the least favorable prognosis. Squamous cell carcinoma of the palate is uncommon, and the dorsal surface of the tongue is almost never affected.

Risk factors identified include **smoking**, alcohol consumption, painful and ill-fitting dentures, chronic inflammation and the use of **smokeless tobacco.**

Important: Tobacco use is the **primary** risk factor.

Remember: The most reliable histologic criterion for a diagnosis of oral squamous cell carcinoma is **invasion**.

A patient in your operatory is observed to have a drooping mouth on one side and a watering eye. She complains of having a loss of taste sensation on the anterior portion of her tongue. This patient is **most likely** suffering from:

- Cerebral palsy
- Bell's palsy
- Tic douloureux
- Multiple sclerosis

- **Bell's Palsy**

Bell's palsy is a form of facial paralysis resulting from damage to the facial nerve. It can strike at any age; **however**, it disproportionately attacks pregnant women and people who have diabetes, influenza, a cold, or some other upper respiratory ailment.

Clinical signs include a unilateral paralysis of all facial muscles with loss of eyebrow and forehead wrinkles, drooping of the eyebrows, flattening of the nasiolabial furrow, sagging of the corner of the mouth and the inability to frown or raise the eyebrows. The upper and lower lips may also be paralyzed on the side affected.

After its sudden onset the paralysis begins to subside within two or three weeks, and gradual, complete recovery occurs in over 85% of patients.

Remember: While attempting to give an inferior alveolar nerve block, if you inject the anesthetic solution into the capsule of the parotid gland, you may cause a Bell's palsy like feeling for the patient by anesthetizing the facial nerve.

Which of the following is the **prime factor** that initiates the myofascial pain-dysfunction syndrome *(MPD)*?

- Trauma
- Muscle spasm
- Periodontal disease
- Tumor

• **Muscle spasm**

Such spasm may arise in one of three ways: muscular overextension, muscular overcontraction or muscle fatigue. The most frequent cause of the spasm seems to be **muscle fatigue.** This syndrome is seen predominantly in women, usually in the 20 to 40 age range, and generally occurs unilaterally.

There are **four cardinal signs** and symptoms of the syndrome:
1. Pain
2. Muscle tenderness
3. A clicking or popping noise in the TMJ
4. Limitation in jaw motion *(especially in the morning)*

The pain itself is usually **unilateral** and is described as a dull ache in the ear or preauricular area, which may radiate to the angle of the mandible, temporal area or lateral cervical area. The muscle most apt to exhibit tenderness is the **lateral pterygoid** muscle.

Note: There are **no radiographic** findings associated with MPD.

Treatment: Most cases are self-limiting. Soft diet, limited talking, no gum chewing, moist heat, NSAID's and Diazepam help relieve symptoms.

An unusual phenomenon, which arises as a result of damage to the auriculotemporal nerve and subsequent reinnervation of the sweat glands by parasympathetic salivary fibers is called:

- Postherpetic neuralgia
- Orolingual paresthesia
- Frey's syndrome
- Glossopharyngeal neuralgia

• **Frey's syndrome**

***It is also called the **auriculotemporal syndrome.**

The syndrome follows some surgical operation such as removal of a parotid tumor or ramus of the mandible, or an infection of the parotid that has damaged the auriculotemporal nerve *(a branch of V-3)*. **Important: Gustatory sweating** is the chief symptom of this syndrome. The patient typically exhibits flushing and sweating of the involved side of the face during eating. This syndrome is not a common condition.

Glossopharyngeal neuralgia refers to pain similar to that of trigeminal neuralgia, which arises from the glossopharyngeal nerve *(CN IX)*. It is not **as common** as trigeminal neuralgia, but the pain may be as severe when it does occur. It occurs in both sexes, in middle-aged or older persons and presents as a **sharp, sudden shooting pain in the ear,** the pharynx, nasopharynx, tonsils or posterior portion of the tongue. It is almost always **unilateral.**

Postherpetic neuralgia is a persistent burning, aching, itching and hyperesthesia along distribution of a cutaneous nerve following an attack of **herpes zoster.** It may last for a few weeks or many months. Involvement of **the facial nerve and geniculate ganglion** produces the **Ramsey Hunt Syndrome**, which is characterized by facial paralysis and otalgia *(earache)*.

A chronic condition characterized by **extreme muscle weakness** is called:

- Myasthenia gravis
- Myelofibrosis
- Multiple sclerosis
- Graves' disease

• **Myasthenia gravis**

Myasthenia gravis is an autoimmune disorder in which the body creates antibodies against its own nicotinic acetylcholine receptors in the neuromuscular junctions. The muscles are quickly fatigued with repetitive use. It is typical for a myasthenic patient to have a flattened smile and droopy eyes, with slow papillary light responses. Xerostomia and rampant caries may accompany myasthenia gravis. The acetylcholine that is necessary for the proper transmission of nerve impulse is destroyed, with the result that salivary glands **do not** receive adequate stimulation.

Multiple sclerosis is a chronic, often disabling disease that randomly attacks the CNS *(brain and spinal cord)*. It is believed to be due to an autoimmune response in which the immune system attacks a person's own tissue. Twice as many women as men have MS, with the onset of symptoms occurring most often between the ages of 20 and 40. Symptoms may range from tingling and numbness to paralysis and blindness. Patients with multiple sclerosis sometimes have facial and jaw weakness. In addition, both **Bell's palsy and trigeminal neuralgia** may develop more frequently in patients with MS.

Trigeminal neuralgia is also known as:

- Glossopharyngeal neuralgia
- Spasmotic torticollis
- Tic douloureux
- Bell's palsy

• **Tic douloureux**

Trigeminal neuralgia is an excruciating painful illness in which the afflicted feels **sudden stab-like pains** in the face. The pains usually last only moments, but are among the most severe pains humans can feel. The pain is provoked by touching a **"trigger zone"**, typically near the nose or mouth. It is caused by degeneration of the **trigeminal nerve** or by pressure being applied to it. Any of the three branches may be affected. The momentary bursts of pain recur in clusters, lasting many seconds. Paroxysmal episodes of the pains may last for hours.

The drug of choice for treating trigeminal neuralgia is **carbamazepine** *(Tegretol)*. It is an analgesic and anticonvulsant. It is also prescribed in the treatment of certain seizure disorders. When used for the treatment of trigeminal neuralgia, it usually relieves the pain within 48 hours after treatment is started.

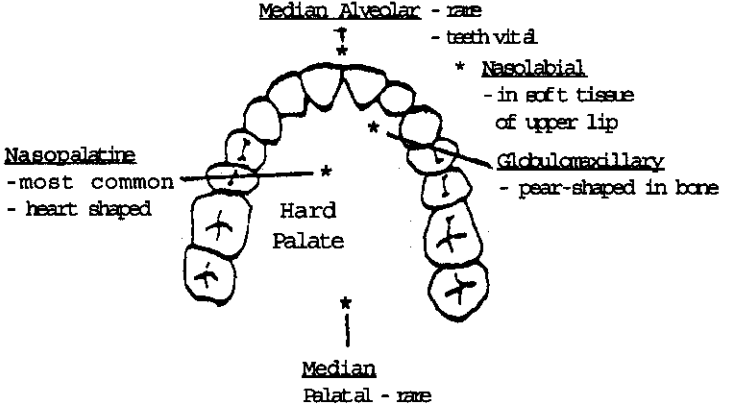
Which cyst is **extraosseous?**

- Median alveolar cyst
- Globulomaxillary cyst
- Nasiolabial cyst
- Nasopalatine cyst

• **Nasolabial cyst** → also called nasoalveolar cyst

Because this cyst is extraosseous, it is **not likely to be seen on a radiograph.**

Quick reference for developmental cysts:



All of the following cysts are **congenital, except:**

- Thyroglossal duct cyst
- Branchiogenic cyst
- Globulomaxillary cyst
- Dermoid cyst

- **Globulomaxillary cyst**

Congenital Cysts

- The **thyroglossal duct** cyst, which may arise from any portion of the thyroglossal duct. This cyst is therefore found in a **midline position** and is usually dark in color. It may be vascular as to resemble a hemangioma. One frequent important symptom is **hemorrhage into the mouth,** resulting from the rupture of the overlying veins. Complete excision of the tract to the base of the tongue, frequently including a portion of the hyoid bone, is necessary for a cure.

- The **branchiogenic cyst**, which arises from the persistence of the second branchial arch cleft. This cyst is characteristically located along the **anterior border of the sternocleidomastoid muscle** at any level in the neck. This type of cyst is lined with ciliated and stratified squamous epithelium and contains a milky or mucoid fluid. The treatment consists of complete surgical excision.

- The **dermoid cyst** is relatively uncommon in the oral cavity. This cyst frequently contains hair, sebaceous and sweat glands, as well as tooth structures. The most common site is the **floor of the mouth.** The treatment is the surgical removal of the entire tumor.

Note: The globulomaxillary cyst is a **developmental** cyst.

All of the following cysts are **developmental** *(or fissural)*, **except:**

- Nasopalatine cyst
- Nasolabial cyst
- Globulomaxillary cyst
- Branchiogenic cyst
- Median palatal cyst
- Median alveolar cyst

• **Branchiogenic cyst**

Developmental Cysts

1. **Nasopalatine** → "heart-shaped" radiolucency in midline, **most frequent** type of non odontogenic cyst. Usually asymptomatic or may produce an elevation in the anterior part of the palate. **Teeth are vital.** Treatment is enucleation.

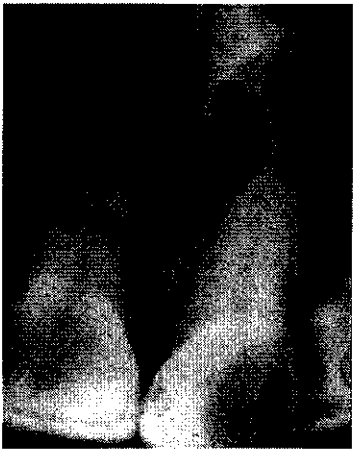
2. **Nasolabial** *(nasoalveolar)* → is superficially located in soft tissues of the upper lip. This is an **extraosseous** cyst. Treatment is surgical excision.

3. **Globulomaxillary** → "pear-shaped" radiolucency between maxillary lateral and cuspid. Asymptomatic, **all regional teeth are vital.** Treatment is enucleation.

4. **Median palatal** → rare, may occur anywhere along median palatal raphe. May produce swelling on palate. Treatment is enucleation.

5. **Median alveolar** → rare, occurs in bony alveolus between central incisors. Distinguished from periapical cyst by the fact that **adjacent teeth are vital.** Treatment is enucleation.

The **circular radiolucent** area seen in this radiograph is clinically seen as a **marked swelling** in the region of the palatine papilla. It is situated distal to the roots of the central incisors. The pulps of the anterior teeth in this patient tested **vital**. These findings would be compatible with what **diagnosis?**



• **Nasopalatine duct cyst** → also known as an incisive canal cyst

Of the cysts of the jaw, those that arise from epithelial remnants in the incisive canal are the **most common type** of maxillary developmental cyst. Histologically, this cyst is lined with vessels, nerves and mucous glands in the wall. They most often remain limited as to size and are **asymptomatic**. Some of them, however, become infected or show a tendency to grow extensively. When this occurs, surgical intervention is indicated.

Note: The **soft tissue** *(and far less common)* **variant** of the nasopalatine canal cyst is the cyst of the **palatine papilla**.

When making a diagnosis of this cyst, the following **two cysts** should be **ruled out:**

- The **globulomaxillary cyst** → usually appears between the roots of the lateral incisor and those of the canine. It is **"pear-shaped"** and often causes the roots of involved teeth to diverge. **See picture #89 in booklet.**

- The **median palatal cyst** → usually situated in the midline of the hard palate, posterior to the premaxilla. Clinically, this lesion presents as a firm swelling, which is usually painless. **Note:** Some investigators now believe that this cyst represents a **more posterior** presentation of a nasopalatine duct cyst, rather than a separate cystic degeneration of epithelial rests at the line of fusion of the palatine shelves. **See picture #88 in booklet.**

The **most common** developmental *(or fissural)* cyst is:

- The nasopalatine duct cyst
- The median palatal cyst
- The globulomaxillary cyst
- The nasiolabial cyst

- **The nasopalatine duct cyst**

Fissural Cyst	Histogenesis	Clinical Characteristics	Radiographic Characteristics	Treatment and Prognosis
Nasopalatine duct *(incisive canal cyst)*	Remnants of nasopalatine ducts *(within bone)*	Most common fissural cyst: usually asymptomatic. May complain of tender swelling of palate	Well-demarcated round, oval or heart-shaped radiolucency between and above maxillary central incisors; rarely just lateral to midline; lesio crosses midline; teeth vital	Surgical excision, prognosis excellent, don't confuse with enlarged palatine foramen
Median palatal	Epithelial remnants in line of fusion between palatine processes	Soft fluctuant or crepitant swelling in midline of hard palate	Well-demarcated radiolucency	Enucleation, prognosis excellent
Globulomaxillary	Epithelial remnants in line of fusion between globular and maxillary processes	Usually asymptomatic, occasionally produces swelling with or without pain	Inverted pear-shaped radiolucency between roots of maxillary lateral incisor and canine teeth, teeth vital but roots may diverge	Enucleation with preservation of teeth, prognosis excellent, don't confuse with lateral periodontal cyst

All of the following cysts occur **within bone, except:**

- The nasopalatine cyst
- The median palatal cyst
- The nasiolabial cyst
- The globulomaxillary cyst

• **The nasiolabial cyst**

***The nasiolabial cyst is also called the **nasoalveolar** cyst.

Histogenesis: Develops from epithelial remnants from the inferior and anterior portion of the nasolacrimal duct.

Clinical characteristics: Swelling just below or inside nostril. May present in the canine region. **It is a soft tissue cyst.**

Radiographic characteristics: Not visible, but may produce "cupping" of underlying bone. **Not within bone** *(extraosseous).*

Microscopic characteristics: Fibrous connective tissue forms the wall of the cyst. The epithelial lining is characteristically **pseudostratified columnar** type with numerous goblet cells.

Treatment and prognosis: Enucleation; prognosis excellent.

Which of the following soft tissue tumors is most likely to be found on the **tongue?**

- Traumatic neuroma
- Neurilemoma *(Schwannoma)*
- Neurofibroma
- None of the above

- **Neurilemoma** → also called a Schwannoma

Tumor	Etiology	Clinical Characteristics	Treatment and Prognosis
Traumatic Neuroma	Trauma to a peripheral nerve	Most common site over **mental foramen** in edentulous patients	Excision with small proximal portion of involved nerve; recurrence uncommon
Neurilemoma *(Schwannoma)*	It is derived from proliferation of **Schwann cells** of the neurolemma that surrounds peripheral nerves	Encapsulated mass that presents as an **asymptomatic lump. The tongue** is the most common location. Bony lesions may cause pain or paresthesia	Conservative excision; recurrence rare
Neurofibroma	Some investigators say it is derived from the Schwann cell; others say the perineural fibroblast	Two forms: 1. **Solitary** neurofibroma: asymptomatic nodule, occurs on tongue, buccal mucosa and vestiblule 2. **Multiple** lesions seen in neurofibromatosis	1. **Solitary:** surgical excision 2.Neurofibromatosis: removal is impractical. Watch for high rate of **malignant transformation**

All of the following are characteristics common to a child with **Albright's syndrome, except:**

- Irregular skin pigmentation
- Bone disease
- Heart disease
- Endocrine problems

• **Heart disease**

Albright's syndrome *(also called* **McCune-Albright** *syndrome)* is a disease of unknown cause affecting the bones and pigmentation of the skin, and causing premature sexual development. The extent to which each of these problems exist in those with the syndrome is quite variable. The hallmark of Albright's syndrome is **premature puberty in the female**. Early sexual development in the male is less common than the female.

The **triad** of symptoms includes:
1. **Polyostotic** type of fibrous dysplasia
2. **Café-au-lait** spots on the skin
3. **Endocrine abnormalities** → most common is precocious sexual development in females

***Pathologic fractures** are frequently associated with this syndrome

There is no specific treatment for this syndrome. Drugs that inhibit estrogen production, such as testolactone, have been tried with some success.

Important: An additional complication is the **malignant transformation potential** of both the polyostotic *(mainly)* and monostotic fibrous dysplasia into **osteosarcomas**.

All of the following statements concerning **eosinophilic granuloma** are true, **except:**

- It tends to start between the age of 50 and 70
- It affects males more than females
- It usually affects the bones but also affects the lungs
- Clinically, the lesions may present no physical signs or symptoms

• **It tends to start between the age of 50 and 70**

***This is false;** it tends to start between the age of 20 and 40 *(young adults)*

Eosinophilic granuloma is the most **benign** form of Histiocytosis X. It may be totally asymptomatic, however, there may be local pain or swelling particularly if fracture has occurred. In the mouth, the **mandible** is most likely affected, with teeth on the affected side being loose with signs of gingivitis. When the lungs are affected, the symptoms can include coughing, shortness of breath, fever, and weight loss. Pneumothorax is a common complication.

Radiographically, the lesions appear as irregular radiolucent areas usually involving superficial alveolar bone. Lesions in the jaw usually appear as single or multiple radiolucencies, which may be so well-circumscribed as to resemble cysts or periapical granulomas.

Bone lesions often resolve spontaneously and do not require treatment unless they cause symptoms. Curettage provides diagnostic biopsy material and is curative.

Remember: Eosinophilic granuloma, Hand-Schüller-Christian disease and Letterer-Siwe disease have traditionally been grouped under the generic term **Histiocytosis X**. These diseases occur as a result of metabolic defects in the reticuloendothelial system. They are characterized by the proliferation of histiocytes *(macrophages)* of loose connective tissue.

The **central giant cell granuloma** occurs predominantly in:

- Infants
- Children or young adults
- Middle aged adults
- Elderly people

• **Children or young adults** *(before the age of 20)*

The **central giant cell granuloma** is a benign process that occurs almost exclusively **within the jawbones.** It may be caused by trauma from a fall or blow, or even a tooth extraction.

It is somewhat more common in **females** than males. Either jaw may be involved, but the **mandible** is affected more often. The lesions are more common in the **anterior segments** of the jaws, and not uncommonly, cross the midline. Pain is not a prominent feature of this lesion. Slight to moderate bulging of the jaw due to expansion of the cortical plates occurs in the involved area, depending upon the extent of bone involvement. The radiographic feature of this granuloma consists of a multilocular radiolucency of bone. The margins are well-defined.

Note: It appears similar to an **ameloblastoma** and an **odontogenic keratocyst.**

Histologically, the central giant cell granuloma is made up of loose fibrillar connective tissue. **Multinucleated giant cells** are prominent throughout the connective tissue.

The **treatment** is curettage or surgical excision. These lesions will fill in with new bone after excision.

See picture #54 in booklet.

Condylar hyperplasia is:

- A common, bilateral enlargement of the condyle
- A rare, bilateral enlargement of the condyle
- A common, unilateral enlargement of the condyle
- A rare, unilateral enlargement of the condyle

• **A rare, unilateral enlargement of the condyle**

The cause of this condition is **unknown**, but many people feel it is the result of mild, chronic inflammation that stimulates the growth of the condyle or adjacent tissues.

Patients afflicted with condylar hyperplasia usually exhibit a **unilateral, slowly progressive** elongation of the face with deviation of the chin **away** from the affected side. The affected TMJ may or may not be painful, and there is usually severe malocclusion.

Condylar hypoplasia and aplasia can occur unilaterally or bilaterally. If it is unilateral, there is obvious facial asymmetry, and both occlusion and mastication may be altered. A shift of the mandible **toward the affected** side occurs during opening

Note: In bilateral cases this shift is not present.

All of the following statements concerning **craniofacial fibrous dysplasia** are true, **except:**

- This condition frequently causes expansion and deformity of the jawbone, as well as displacement of the teeth
- There is a characteristic thickening at the base of the skull, which can be seen radiographically
- Radiographically, the lesion is usually radiopaque, not well-circumscribed, and may have a "ground-glass" appearance
- The treatment of choice is radiation therapy
- It is also referred to as "monostotic" fibrous dysplasia of the jaw

- **The treatment of choice is radiation therapy**

Important: There has been documentation to support the possibility that **malignant transformation** can occur in patients who have been treated by radiation therapy.

The treatment usually consists of surgical removal of the lesion when possible. **However,** since these lesions **are not** usually well-circumscribed, surgical recontouring is often performed. This removes that portion of the lesion causing facial deformity.

Important points concerning monostotic fibrous dysplasia:
- It is the **most common form** and affects both sexes equally
- Jaw involvement is **common** *(especially maxilla)*
- **Radiographically**: The lesion has a **"ground glass"** appearance with poorly defined margins.
- The differential diagnosis of fibrous dysplasia of the jaws includes the **ossifying fibroma,** however, radiographically the ossifying fibroma has a **well-circumscribed appearance**

See picture #38 in booklet.

Mandibular tori most often appear:

- In the retromolar pad region
- On the lingual surface of the mandible, most often in the premolar region
- On the lingual surface of the mandible, inferior to the mylohyoid ridge
- Along the midline of the hard palate

• **On the lingual surface of the mandible, most often in the premolar region**

Mandibular tori *(also called torus mandbularis)* are bony, exophytic growths that occur along the lingual surface of the mandible superior to the mylohyoid ridge.

Mandibular tori may occur singly, however, there is a marked tendency toward **bilateral occurrence,** and the lesion is not necessarily confined to the premolar region. Unlike palatal tori, the mandibular tori are more readily demonstrated radiographically. **See picture #29 in booklet.**

Tori *(maxillary and mandibular)* are of no pathological significance and rarely are they of clinical significance while the normal teeth are still present. If, **however**, a complete denture needs to be made, they should be carefully removed.

All of the following statements concerning the **central ossifying fibroma** of bone are true, **except:**

- It is a slow-growing, painless, asymptomatic neoplasm which may occur in either jaw
- It may occur at any age, but it is far more common in elderly people
- The displacement of teeth may be an early clinical feature
- Because of this slow growth, the cortical plates of bone and overlying mucosa or skin are almost always invariably intact

• **It may occur at any age, but it is far more common in elderly people**

***__This is false;__ it may occur at any age, but is far more common in **young adults.**

This neoplasm presents an extremely variable radiographic appearance depending upon its stage of development. However, despite the stage of development, the lesion is **always well-circumscribed** and demarcated from surrounding bone, in contrast to fibrous dysplasia. In its early stage it appears as a radiolucent area. As the lesion matures, it eventually becomes a relatively uniform radiopaque mass. It most commonly involves only **one bone** *(as opposed to* **Paget's disease***)*.

See picture #32 in booklet.

There is a remarkable similarity in clinical features between this lesion and the **central cementifying fibroma**, a tumor accepted by most investigators as being odontogenic in origin. It has been suggested that these are two separate benign tumors, identical in nature **except** for the cell undergoing proliferation; the osteoblast with bone formation in one case, or the cementoblast with cementum formation in the other case.

Note: The treatment of this lesion is conservative excision, and recurrence is rare.

All of the following are characteristics of **Gardner's syndrome, except:**

- This syndrome is inherited in a dominant manner
- Thousands of polyps develop in the colon, as well as the stomach and duodenum
- This syndrome is also associated with bony tumors in the jaw and skull
- The polyps associated with this syndrome usually appear around age 15
- The polyps rarely undergo malignant transformation

• **The polyps rarely undergo malignant transformation**

***__This is false;__ the polyps **eventually** undergo malignant transformation.

Gardner's syndrome is a polyposis syndrome *(as are Familial multiple polyposis, Peutz-Jeghers syndrome and Turcot's syndrome)*. The most serious complication of Gardner's syndrome is the multiple polyps that affect the large intestine. The inevitable outcome of this disease is **colon cancer.**

The **oral findings** of Gardner's syndrome include:

- Multiple impacted and supernumerary teeth
- Multiple jaw osteomas which give a **"cotton-wool"** appearance to the jaws. These osteomas appear as dense, well-circumscribed radiopacities
- Multiple odontomas

***When Gardner's syndrome is suspected **based on oral findings,** the patient should be referred to a gastroenterologist for consultation. **Note:** Multiple desmoid tumors *(fibromatosis)* and epidermoid cysts of the skin are also characteristic of the disease.

Remember: Multiple impacted and supernumerary teeth are also seen in **Cleidocranial dysostosis.**

The most common form of **fibrous dysplasia** is:

- Monostotic
- Polyostotic
- Polyostotic with endocrine involvement *(Albright's syndrome)*

• **Monostotic**

Fibrous dysplasia is a rare, abnormal condition characterized by the fibrous displacement of the osseous tissue within the bones affected. The specific cause is unknown.

Three Distinct Forms are Recognized

1. **Monostotic** fibrous dysplasia → is the most common form *(80%)*. Affects children and young adults *(affects both sexes equally)*. This form of the disease affects **one bone** *(the ribs and the femur are common sites)*. The jaws are also affected frequently, mainly the **maxilla** *(it presents as a painless swelling or bulge)*. A panorex will show a radiopaque mass with irregular borders that has a **"ground glass"** appearance. **Note:** When several adjacent bones are affected, it is called **craniofacial fibrous dysplasia.**
2. **Polyostotic** fibrous dysplasia → usually displays a segmental distribution of the involved bones *(multiple)*. Occurs during childhood *(mainly females)*. Affects long bones, face, clavicles and pelvic bones. The initial signs may be a limp, a pain or a fracture of the affected side. Females who are affected often reach premature puberty.
3. **Albright's** syndrome → is the **most severe** form of polyostotic fibrous dysplasia *(involves multiple bones)*. Affects young people *(males and females equally)*. This syndrome is characterized by **irregular brown spots** on the skin *(called café-au-lait spots)* and **endocrine abnormalities** *(the most common of which is precocious sexual development in females)*.

All of the following diseases have traditionally been grouped under the generic term **Histiocytosis X, except:**

- Eosinophilic granuloma
- Tay-Sachs disease
- Hand-Schüller-Christian disease
- Letterer-Siwe disease

- **Tay-Sachs disease**

Histiocytosis X is a group of disorders in which abnormal scavenger cells called histiocytes and another immune system cell called the eosinophils proliferate, especially in the bone and lung, often causing scars to form.

- **Eosinophilic granuloma** → is the most **benign** form of Histiocytosis X. It is more common in males around 20 years old. May be totally asymptomatic, however, there may be local pain or swelling. In the mouth, the **mandible** is most likely affected, with teeth on the affected side being loose with signs of gingivitis.
- **Letterer-Siwe disease** *(also called the acute disseminated form)* → starts before age 3 and is usually **fatal** without treatment. The histiocytes damage not only the lungs but also the skin, lymph glands, bone, liver, and spleen. Pneumothorax may occur. Oral lesions are uncommon.
- **Hand-Schüller-Christian disease** *(also called the chronic disseminated form)* → usually begins in early childhood. It is more common in boys. There is a **triad of symptoms,** including exophthalmos, diabetes insipidus and bone destruction *(skull and jaws are affected).* **Oral signs** include bad breath, sore mouth and loose teeth.

Note: People with Hand-Schüller-Christian disease or eosinophilic granuloma may recover spontaneously. All three disorders may be treated with corticosteroids and cytotoxic drugs such as cyclophosphamide. The therapy for bone involvement is radiation. Death usually results from respiratory failure or heart failure.

The **nevoid basal cell carcinoma** syndrome may present which of the following abnormalities?

- Cutaneous anomalies
- Dental and osseous anomalies
- Ophthalmologic abnormalities
- Neurologic anomalies
- Sexual abnormalities
- All of the above

- **All of the above**

The nevoid basal cell carcinoma syndrome is also known as the basal cell nevus-bifed rib syndrome, the basal cell nevus syndrome and the Gorlin and Goltz syndrome.

Possible abnormalities include:

- **Cutaneous anomalies** → including **multiple basal cell carcinomas,** other benign dermal cysts and tumors, palmer pitting, palmer and plantar keratosis and dermal calcinosis.

- **Dental and osseous anomalies** → including odontogenic keratocysts *(often* **multiple***),* mild mandibular prognathism, rib anomalies *(often* **bifed***),* and vertebral anomalies.

- **Ophthalmologic abnormalities** → including hypertelorism with wide nasal bridge and congenital blindness.

- **Neurologic anomalies** → including mental retardation, dural calcification, agenesis of corpus callosum and congenital hydrocephalus.

- **Sexual abnormalities**→ including hypogonadism in males and ovarian tumors in females.

In most cases, what is the proper treatment for an **eruption cyst?**

- Incise and drain
- Prescribe antibiotics
- No treatment is necessary
- Extract the tooth associated with the eruption cyst

• **No treatment is necessary**

In a few rare cases, incision or even the removal of the overlying tissue may be necessitated by pain or tenderness associated with the lesion.

An eruption cyst is essentially a soft tissue variant of the dentigerous cyst. It is invariably associated with an erupting tooth *(usually* **primary** *but occasionally a permanent tooth)*. The effects are mostly limited to the overlying gingival tissues rather than bone.

Clinically, the lesion usually appears as a **smooth-surfaced,** reddish – pink or bluish – black, fluctuant, localized swelling on the **alveolar ridge** over the crown of an erupting primary or permanent molar tooth. The intense bluish color, which is often characteristic, is due to an accumulation of blood. Due to this appearance, it may be mistaken for a hemangioma or hematoma.

ORAL PATHOLOGY

Odont Cyst

Upon viewing a panorex of a 14-year-old patient, you see a large radiolucent area on the left side of the mandible apical to the premolars and first molar. No clinical symptoms are present. Teeth are not carious and respond normally to vitality tests. Medical history is unremarkable. Upon opening into the area, no fluid or tissue is evident. What is the **most probable diagnosis?**

- Dentigerous cyst
- Traumatic bone cyst
- Primordial cyst
- Residual cyst

• **Traumatic bone cyst**

Remember: This cyst may be completely devoid of solid or liquid material. It occurs most frequently in younger persons with no sex predilection. The usual location is in the **mandible** between the cuspid and ramus. The regional teeth are **vital**.

The **dentigerous** cyst contains a crown of an **unerupted tooth** or dental anomaly such as an odontoma. Enlarged dentigerous cysts can cause marked displacement of teeth. Pressure of accumulated fluid usually displaces the tooth in an apical direction.

The **primordial** cyst *(also called a* **follicular** *cyst)* differs from the periodontal and dentigerous cysts in that it contains no calcified structures. These cysts are lined by stratified squamous epithelium and may be either locular, multilocular or multiple.

The **residual** cyst refers to a situation in which a tooth associated with a **radicular** cyst is extracted but the cyst is left undisturbed, it persists within the jaw and this lesion is called a **residual** cyst. **Note:** You must curette the socket of a tooth with a radicular cyst after extraction.

Which of the following cysts is most commonly found in relation to a **developing third molar?**

- Lateral periodontal cyst
- Dentigerous cyst
- Fissural cyst
- Primordial cyst
- Traumatic cyst
- Residual cyst
- Gingival cyst

• **Dentigerous Cyst**

***It usually contains a crown of an **unerupted** tooth. If a tooth with a dentigerous cyst begins to erupt, the bulging which the cyst produces on the ridge is called an **eruption** cyst.

The **lateral periodontal** cyst is inflammatory in origin and forms along the lateral surface of the tooth. If at the apex, they are termed **radicular** cysts.

The **fissural** cysts *(which are also called* **developmental** *cysts)* are non-dental in origin, they include nasoalveolar, median palatal, nasopalatine, and globulomaxillary cysts.

The **primordial cyst** *(also called* **follicular** *cyst)* contains **no** calcified structures.

The **traumatic bone** cyst may contain blood, fluid, debris or be **completely empty.** Commonly found in young persons, in the mandible between the cuspid and ramus.

The **residual** cyst is often found in edentulous areas. This cyst refers to a situation in which a tooth with a radicular cyst associated with it was extracted, and the socket wasn't curetted. The radicular cyst persists in the jaw as a **residual** cyst.

The **gingival** cyst is a rare, circumscribed swelling of the gingiva,which is usually seen in the canine and bicuspid areas of the mandible. Usually limited to the gingiva but larger ones may erode the bone. They are easily excised.

Follicular and dentigerous cysts that contain **keratinizing** material are known as:

- Residual cysts
- Traumatic bone cysts
- Keratocysts
- Granulomas

• **Keratocysts**

Keratocysts differ from other odontogenic cysts in their microscopic appearance and clinical behavior. They may resemble periodontal, primordial or follicular cysts. Usually they **cannot** be distinguished radiographically.

Keratocysts increase in size principally by a process of epithelial cell multiplication. The treatment of choice is the excision of overlying mucosa in the area where the cyst wall is adhered.

Important: The most remarkable feature of keratocysts is their **great tendency toward recurrence.**

Which cyst is found in **place of a tooth** rather than directly associated with one?

- Residual cyst
- Traumatic bone cyst
- Primordial cyst
- Periodontal cyst

- **Primordial cyst**

Cyst	Clinical	Radiograph	Histological
Primordial Cyst	**Found in place of tooth** Arises from epithelium of the enamel organ Affects males and females under 25 equally Located **inmandibular third molar space**	Well-defined oval RL lesion	Stratified squamous epithelium No rete pegs
Residual Cyst	Any age, male and females equally Usually asymptomatic Located in edentulous space History o**prior extraction** in area	Well-defined RL **not associated** with tooth Usually solitary	Same **asradicular** *(apical periodontal cyst)*, stratified squamous epithelium lining the lumen Represents **aradicular** cyst which was left in jaw when associated **tooth was extracted**

ORAL PATHOLOGY

Odont Cyst

Which of the following odontogenic cysts is **always** associated with the crown of an **unerupted or developing tooth**?

- Lateral periodontal cyst
- Dentigerous cyst
- Odontogenic keratocyst

- **Dentigerous cyst** → also called follicular cyst

Cyst	Clinical	Radiograph	Histological
Dentigerous *(follicular cyst)*	Children and teenagers Mandibular third molar and maxillary canine area (**70% in** mandible) Associated with impacted or unerupted teeth Second most common odontogenic cyst	Well-defined usually unilocular RL in association with the crown of an unerupted tooth	Lined by non-keratinized stratified squamous epithelium Rete pegs are absent
Odontogenic keratocyst	Usually occurs between the ages of 10-30 Often associated with an **impacted tooth** 50% mandibular third molar area **Over 30% recurrence rate**	Well-circumscribed RL with smooth margins and thin radiopaque borders	Thin layer of corrugated parakeratin Uniform thin stratified squamous lining Distinct cuboidal to columnar basal layer Varying amounts of keratin debris may be seen in lumen
Lateral Periodontal cyst	95% mandibular cuspid-bicuspid area Apposition with root of vital tooth Usually symptomless	Well-defined, round or teardrop-shaped RL with an opaque margin along lateral surface of tooth	Thin lining of non-keratinized epithelium

A dental granuloma and a radicular cyst can be **differentiated:**

- Based on symptoms
- Radiographically
- Histologically
- By an Electric Pulp Tester

• **Histologically** → and only histologically

Lesion	Usual Location	Clinical and Radiographic Features	Histologic Features	Treatment
Dental granuloma (It is one of the **most common** of all sequelae of pulpitis)	Apex of tooth	• Asymptomatic • Circumscribed radiolucency at apex of tooth • Tooth is **non-vital** • May be sensitive to percussion	• Fibrous connective tissue, with macrophages, lymphocytes, cells and capillaries • Also present is stratified squamous epithelium	Root canal treatment or extraction of involved tooth
Radicular cyst *(Also called apical periodontal cyst and periapical cyst)*. It is the **most common** odontogenic cyst	Apex of tooth	• Asymptomatic • Circumscribed radiolucency at apex of tooth • Tooth is **non-vital** • May be sensitive to percussion	• Exhibits a lumen *(true cyst)* that is invariably lined by stratified squamous epithelium • The wall is made up of condensed connective tissue which contains plasma cells, lymphocytes and PMN leukocytes	Root canal treatment with apicoectomy or extraction with curettage of socket

The **radicular** or **periapical** cyst develops within a **pre-existing** periapical granuloma *(also called a dental granuloma)*.

Note: Increased osmotic pressure in the cyst lumen is important in the pathogenesis of a radicular cyst.

Which form of **ameloblastoma** is the more aggressive one and requires more extensive treatment?

- Solid *(multicystic)*
- Unicystic
- Extraosseous

• **Solid** *(multicystic)*

Ameloblastoma consists entirely of odontogenic epithelium, which – at sites – shows the differentiation of the familiar, histologic layers of the enamel organ. It is most often seen in adolescents in the **mandibular** *(retro)* **molar** area. It is usually benign but often shows a highly expansive and locally invasive mode of growth. Enlargement of the tumor may expand the buccal, lingual, or palatal bone plates. Various histologic types have been identified such as follicular, cystic, acanthomatous, plexiform, granular cell and desmoplastic. They are also sometimes classified as multicystic versus unicystic.

Treatment: Each case should be considered on its own basis. The **solid** *(multicystic)* lesion requires surgical excision. Resection should be reserved for larger lesions. **Unicystic** lesions usually require only enucleation. They should not be over treated. Recurrence is common if inadequately treated.

Which odontogenic tumor has a **high rate of recurrence** if it is inadequately treated?

- Odontogenic myxoma
- Odontogenic fibroma

- **Odontogenic myxoma**

Tumor	Histogenesis	Clinical Characteristics	Radiographic Characteristics	Treatment and Prognosis
Odontogenic myxoma	Dental papilla, dental sac, or periodontal ligament	Usually 3rd-4th decade; mandible most common site; painless swelling	Poorly defined multilocular radiolucency; may be associated with unerupted and/ or displaced teeth; it is an aggressive tumor	Thorough curretage with cautery; recurrence common if inadequately treated
Odontogenic fibroma	Dental papilla, dental sac, or periodontal ligament	Children and young adults; mandible; painless swelling	Multilocular or unilocular radiolucency; may be associated with unerupted and/ or displaced teeth	Enucleation; Recurrence rare

Which odontogenic tumor usually affects **males** under the age of 25 years old?

- Benign cementoblastoma
- Gigantiform cementoma

- **Benign cementoblastoma**

Tumor	Histogenesis	Clinical Characteristics	Radiographic Characteristics	Microscopic Characteristics	Treatment and Prognosis
Benign Cementoblastoma *(true cementoma)*	Periodontal ligament	**Males, under 25,** mandibular bicuspids or molars; usually solitary; may cause expansion of cortical plates; **tooth vital**	Well-demarcated, mottled or densely radiopaque mass with radiolucent periphery; attached to the tooth root; root resorbed	**Cementum-like tissue** with conspicuous reversal lines; variable amount of fibrous connective tissue with sheets of uncalcified "cementoid", especially at periphery	Extraction of involved tooth
Gigantiform Cementoma *(familial multiple cementomas)*	Periodontal ligament	**Middle-aged black women;** multiple, often symmetrical; may cause expansion of jaw	Large, dense, often lobulated radiopaque masses	Large sheets of tissue resembling **secondary cementum**	Conservative excision

Which odontogenic tumor requires **no treatment** at all?

- Cementifying fibroma
- Cementoma

- **Cementoma**

***Also known as **periapical cemental dysplasia**

Tumor	Histogenesis	Clinical Characteristics	Radiographic Characteristics	Treatment and Prognosis
Cementifying fibroma	Periodontal ligament	Adult, mandible, painless swelling	Well-defined radiolucency with scattered radiopaque foci	Through curettage, recurrance is rare
Cementoma *(periapical cemental dysplasia)*	Periodontal ligament	Affects women over the age of 30, **mandibular incisor region,** frequently multiple, teeth vital, sometimes seen exclusively in blacks	Small sharply circumscribed radiopacity attached to or adjacent to apices of teeth, early lesions radiolucent, then central opacity, finally densely radiopaque	None, simply recognize the condition and periodically observe

Which of the following tumors is also called the **Pindborg** tumor?

- Adenomatoid odontogenic tumor
- Calcifying epithelial odontogenic tumor
- Squamous odontogenic tumor

• Calcifying epithelial odontogenic tumor

Tumor	Histogenesis	Features	Clinical Characteristics	Treatment and Prognosis
Adenomatoid Odontogenic tumor *(Adenoameloblastoma)*	Enamel organ, lining of dentigerous cyst, reduced enamel epithelium, Rests of Malassez	Second decade *(teens)*, Anterior maxilla most common site, asymptomatic or painless swelling	Unilocular radiolucency associated with crown of unerupted tooth, may be tiny radiopaque foci	Enucleation; recurrence extremely rare
Calcifying epithelial odontogenic tumor *(Pindborg tumor)*	Reduced enamel epithelium	Fourth decade *(30%)*, not likely in children and adolescents, mandibular molar-bicuspid area, painless swelling, rarely extraosseous	Radiolucent-radiopaque area associated with unerupted tooth, associated with amyloid production	Resection of affected area; recurrence common if inadequately treated
Squamous odontogenic tumor	Rests of Malassez	May be asymptomatic, a painless swelling, or associated with mobile teeth	Triangular or circumscribed radiolucency associated with unerupted or erupted tooth	Conservative excision and close follow-up; recurrence rare

Remember: These tumors all are of ectodermal origin *(purely epithelial)*.

Which of the following odontogenic tumors have a **mixed origin** *(ectodermal-mesodermal)*?

- Ameloblastic fibroma
- Ameloblastic fibro-odontoma
- Ameloblastic odontoma
- All of the above

- **All of the above**

Odontogenic Tumors with a mixed origin *(ectodermal and mesodermal components of tooth germ)*				
Tumor	**Clinical Features**	**Radiographic Features**	**Microscopic Features**	**Treatment and Prognosis**
Ameloblastic fibroma ***often mistaken for **ameloblastoma**	Below age 20, **mandibular** bicuspid-molar area, painless swelling Compared to ameloblastoma: Younger age Slower growth Does not infiltrate	Well defined radiolucency frequently associated with **unerupted** tooth	Nests and strands of odontogenic epithelium and young cellular fibrous connective tissue resembling dental papilla	Conservative excision, recurrence rare
Ameloblastic fibro-odontoma	Below age 20, similar to ameloblastic fibroma cut occurs with equal equal frequency in maxilla and mandible.	Similar to ameloblastic fibroma but may show foci of calcification	Similar to ameloblastic fibroma but with dentin and enamel *(induction effect)*	Similar to ameloblastic fibroma
Ameloblastic odontoma	Usually below age 20, **maxillary or mandibular** bicuspid-molar area, painless swelling	Well defined radiolucency with foci of calcification, may or may not be associated with erupted tooth	Simple ameloblastoma in combination with composite odontoma	Resection of affected area, **may recur** if incompletely excised

The most common **epithelial** odontogenic tumor is:

- The adenomatoid odontogenic tumor *(adenoameloblastoma)*
- The ameloblastoma
- The calcifying epithelial odontogenic tumor *(Pindborg tumor)*
- The squamous odontogenic tumor

• **The ameloblastoma**

Histogenesis: Lining of **dentigerous cyst**, oral epithelium, dental lamina or enamel organ, and possibly remnants of Hertwig's sheath. **Key point:** Consists entirely of **odontogenic epithelium,** which-at sites-shows the differentiation of the histologic layers of the enamel organ.

Clinical features: It is most often seen in adolescents in the **mandibular** *(retro)* **molar** area. Clinically it is the **most aggressive** odontogenic tumor. It is usually benign but often shows a highly expansive and locally invasive mode of growth.

Radiographic characteristics: Multilocular or unilocular radiolucency, teeth vital. **Note:** In the mandible it appears similar to the **central giant cell** granuloma. **See picture #55 in booklet.**

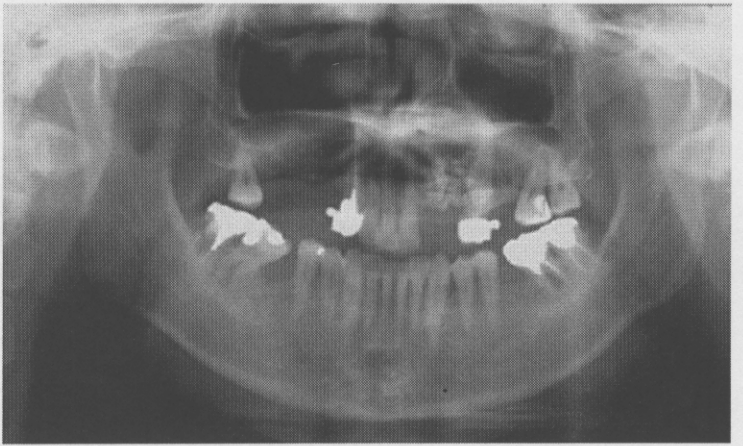
Microscopic features: Various histologic types have been identified such as follicular, plexiform, acanthomatous, granular cell and desmoplastic. All are **non-encapsulated.** Ameloblastomas have been further subdivided into two forms – solid *(multicystic)* and unicystic.

Treatment and prognosis: Varies depending on subtype, **recurrence common if inadequately treated,** very rarely metastasizes.

Note: You need to know the ameloblastoma **extremely well** for the exam!!!

Remember: All of the other tumors on the front of the card are also odontogenic tumors of **ectodermal** *(epithelial)* origin.

How would you refer to the group of small **radiopacities** between the maxillary premolar and maxillary central incisor in the panorex below?



- **Compound odontoma**

Type of odontoma	Histogenesis	Clinical Features	Radiographic Features	Microscopic Features	Treatment & Prognosis
Complex	Ectodermal and mesenchymal components of tooth germ	Second and third decades; **mandibular bicuspid-molar** area; **Asymptomatic**; may cause delayed eruption of permanent teeth	Well-defined radiopaque mass surrounded by narrow radiolucent zone; may or may not be associated with erupted tooth	Conglomerate mass of dental tissues *(dentin, enamel, and cementum)*	Enucleation; does not recur
Compound	Ectodermal and mesenchymal components of tooth germ	Second and third decades; **maxillary incisor-cuspid** area; **Asymptomatic**; may also appear in the mandible near canine-cuspid area; may cause delayed eruption of permanent teeth	Multiple small **tooth-like structures**	Multiple small, **malformed teeth**; composed of dentin, enamel and cementum	Enucleation; does not recur

See picture #43 in booklet for picture of complex odontoma.

Cementomas *(periapical cemental dyplasia)* occur most frequently in the:

- Mandible
- Maxilla

• **Mandible** → specifically, the anterior periapical region

Periapical cemental dysplasia *(cementoma)* represents a reactive rather than a neoplastic process. The term "cementoma" is a misnomer as the opacities are not cementum **but bone.** While they appear to arise from the teeth, the lesions apparently arise within the bone instead. It appears to be an unusual response of the periapical bone to some local factor *(for example,* **traumatic occlusion** *or* **infection***).* **See picture #28 in booklet.**

Clinical features:

- Occurs at the apex of **vital** teeth
- Affects **women over the age of 30** *(especially black women)* more than men.
- Asymptomatic, usually multiple, small periapical areas of radiolucency in the mandibular incisor area.

Note: Depending on stage, it may appear mixed radiolucent and radiopaque or totally radiopaque.
Important: Age, gender, location, radiographic appearance, and tooth vitality considered together are **diagnostic of this condition.**

Three stages:

1. **Osteolytic** stage → **radiolucency** appears on radiograph.
2. **Cementoblastic** stage → beginning of calcification in the radiolucent area *(***mixed radiolucent** *and* **radiopaque** *appearance).*
3. **Mature** stage→ **radiopacity** appears on radiograph with a thin radiolucent line around area. **Note:** No treatment is required for cementomas. Once this stage is reached, the lesion stabilizes and causes no complications.

All of the following statements are true concerning **Peutz-Jeghers syndrome, except:**

- It is an inherited disorder, transmitted as an autosomal dominant trait
- It is also called Hereditary Intestinal Polyposis Syndrome
- Multiple intestinal polyps are usually distributed through the entire intestine, especially in the jejunum
- Melanin pigmentation of the lips and oral mucosa is usually present at birth
- The tongue almost invariably shows this melanin pigmentation
- When this syndrome is suspected on the basis of oral pigmentations, other conditions to be considered in the differential diagnosis are Addison's disease and Albright's syndrome

• The tongue almost invariably shows this melanin pigmentation

***This is false;** the tongue **seldom shows** this melanin pigmentation.

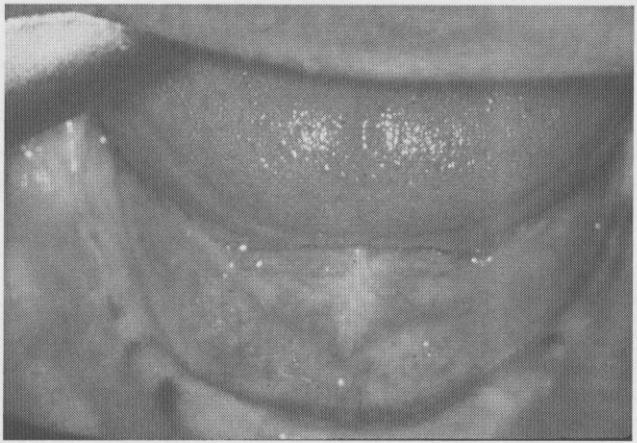
Peutz-Jeghers syndrome *(also called hereditary intestinal polyposis syndrome)* is an unusual condition which is of interest to the dentist because of the oral findings. The pigmentations usually appear at an early age, often during the first decade of life and at this time are restricted to the oral region. **Intraorally**, these pigmentations may be located anywhere on the mucosa, but are most frequently seen on the **buccal mucosa**, gingiva and hard palate. The mucosal surface of the **lower lip** is almost invariably involved. These spots or macules, while of variable intensity, may range through shades of brown, blue and black. During the succeeding decades of the patient's life, pigmentations may arise elsewhere on the skin, especially on the extremities. **It should be noted** that the pigmentations of Peutz-Jeghers syndrome may occur without demonstrable evidence of polyps and, also that multiple polyps may be encountered without pigmentations.

Important: It is significant to note that although the oral pigmentations per se are harmless, their presence is important in that they indicate a necessity for investigating the possible presence of multiple polyposis which may prove harmful. There is a strong tendency for these **multiple polyps of the colon to undergo malignant change.**

See picture #10 in booklet.

Pig Les of Oral Cav

What **most likely** caused the discoloration of the gingiva as seen in the picture below?



• **Amalgam**

An **amalgam tattoo** is a common finding in dental practice today. The tattoo has been mistaken for a melanin-pigmented lesion. The most common locations for amalgam tattoos are the gingiva, buccal mucosa and alveolar mucosa.

Other lesions in the mouth that are due to **chemical injuries** include dilantin hyperplasia, an aspirin burn, and the ingestion of the heavy metal bismuth.

- **Dilantin** is an anticonvulsant drug used extensively in the control of epileptic seizures. An unfortunate side effect of its use is fibrous hyperplasia of the gingiva which is called dilantin hyperplasia.
- **An aspirin burn** occurs when patients place the tablet against an aching tooth, allowing the cheek or lip to hold it in position while it dissolves slowly. Within a few minutes a burning sensation of the mucosa will be noted and the surface becomes blanched or whitened in appearance. The caustic action of the **drug causes necrosis** of the oral mucosa, with subsequent sloughing of the necrotic epithelium.
  See picture #60 in booklet.
- The use of the heavy metal bismuth is still common in treating certain dermatologic disorders as well as various other diseases. Bismuth pigmentation appears as a **"bismuth line,"** a thin, blue-black line in the marginal gingiva which is sometimes confined to the gingival papilla.

The most common location for an intraoral **congenital nevi** *(birthmark)* is the:

- Buccal mucosa
- Tongue
- Hard palate
- Alveolar mucosa

• **Hard palate**

Moles *(nevi)* are small, usually dark, skin growths that develop from pigment-producing cells in the skin *(called melanocytes).* While **nevi** are fairly common on the skin, intraorally they are **quite uncommon**. When present, they are usually on the hard palate, but can also be seen on the gingiva and lips. **Congenital nevi** *(commonly known as* **birthmarks***)* are usually large *(greater than 10 cm)* and with the passage of time, may change from flat, pale tan macules to elevated, verrucous, hairy lesions. Approximately 15% occur on the skin of the head and neck. Congenital nevi have a **higher incidence of malignant transformation** *(as opposed to acquired nevi).*

Acquired nevi are microscopically classified into five subtypes:

1. Intramucosal nevus → most common in oral cavity
2. Blue nevus
3. Compound nevus → rare in oral cavity
4. Junctional nevus → rare in oral cavity
5. Intradermal nevus → is the **most common lesion of skin,** known as the common mole

Note: Acquired nevi are much more common than congenital nevi both **intraorally and extraorally.**

Important: The **B-K mole syndrome** and the **dysplastic nevus syndrome** are both characterized by having numerous large, pigmented atypical nevi which have a high risk for developing **malignant melanoma.**

All of the following conditions demonstrate **pigmentation** of the intraoral mucous membranes, **except:**

- Addison's disease
- Albright's syndrome
- Cushing's syndrome
- Peutz-Jeghers syndrome

• **Cushing's syndrome**

Addison's disease *(also called chronic adrenocortical insufficiency)* results from hypofunction of the adrenal cortex. It is characterized by bronzing of the entire skin. **Oral signs** consist of diffuse pigmentation of the gingiva, tongue, hard palate and buccal mucosa. Although cutaneous pigmentation will most likely disappear following therapy, **pigmentation of the oral tissues tends to persist.**

Albright's syndrome *(also called McCune-Albright syndrome)* is a severe form of **polyostotic fibrous dysplasia,** involving nearly all bones in the skeleton. In addition to the bone lesions there are brown patches of cutaneous pigmentation *(called* **café-au-lait spots***)* and endocrine dysfunction, especially precocious puberty in girls. **Important:** There is an increased incidence of **osteosarcoma** seen with polyostotic fibrous dysplasia.

Peutz-Jeghers syndrome *(also known as Hereditary Intestinal Polyposis Syndrome)* is an inherited disorder that is characterized by having **multiple intestinal polyps** and intraoral **melanin pigmentations**. These pigmentations usually appear during the first decade of life and at this time are restricted to the oral region. They may be located anywhere on the mucosa, but are most frequently seen on the **buccal mucosa,** gingiva and hard palate. The mucosal surface of the **lower lip** is almost invariably involved.

Note: Cushing's syndrome is a hormonal disorder caused by prolonged exposure of the body's tissues to high levels of the hormone cortisol. It is relatively rare and most commonly affects adults aged 20-50. The symptoms vary, but most people have upper body obesity, rounded face, increased fat around the neck *("buffalo hump")*, and thinning arms and legs.

All of the following statements concerning **Addison's disease** are true, **except:**

- It is a rare endocrine disorder which occurs when the adrenal glands do not produce enough cortisol
- It is sometimes called chronic adrenal insufficiency or hypocorticolism
- It most commonly affects teenagers
- It afflicts men and women equally
- It is characterized by weight loss, muscle weakness, low blood pressure, and darkening of the skin in both exposed and unexposed parts of the body

- **It most commonly affects teenagers**

***This is false;** it occurs in **all age** groups.

Addison's disease occurs when the adrenal glands do not produce enough cortisol *(a glucocorticoid)*. Cortisol's most important function is to help the body respond to stress.The failure to produce adequate levels of cortisol can occur for different reasons. The problem may be due to a disorder of the adrenal glands themselves *(primary adrenal insufficiency)* or to inadequate secretion of ACTH by the pituitary gland *(secondary adrenal insufficiency)*. The symptoms of Addison's disease usually begin **gradually**. **These include:**

- Muscle weakness
- Loss of appetite
- Weight loss
- Skin changes with areas of hyperpigmentation covering exposed and nonexposed parts of the body. This darkening of the skin is most visible on scars, skin folds, pressure points such as elbows, knees, knuckles, and toes as well as the **oral mucous membranes** → there is diffuse pigmentation of the gingiva, tongue, hard palate, and buccal mucosa.
- Nausea, vomiting and diarrhea
- Low blood pressure

Laboratory tests show:

- Low blood concentrations of sodium and glucose
- Increased serum potassium
- Decreased urinary output of certain steroids

Important: The main concern when performing dental procedures on a patient with Addison's disease is that the adrenal cortex has **no capacity** to put out extra cortisol as a response to stress.

Which of the following may occur at any age and presents as a single or multiple small, **flat brown** asymptomatic lesion occurring mostly on the **lower lip?**

- Blue nevus
- Compound nevus
- Focal melanosis
- Lipoma

- **Focal melanosis**

A focal melanosis is a term that is used for two similar lesions that **differ in their location.**

- The **labial melanotic macule** is a lesion that appears on the lips *(most frequently on the lower lip)* and almost always occurs near the midline. Most lesions measure 5 mm or less in diameter.

- The **oral melanotic macule** is a lesion that appears intraorally on the gingiva, buccal mucosa and palate. Most lesions are under 1 cm in diameter.

The recommended treatment for either type of focal melanosis is as follows: Those melanotic macules with a relatively **short history** should be excised to establish a definitive diagnosis and to **rule out** the possibility of malignant melanoma. Lesions present for over five years **without a change in size or color** may be followed unless the patient requests removal.

Which type of **acquired nevi** listed below is the **most common** variety seen in the oral cavity?

- Intradermal Nevus
- Compound nevus
- Junctional nevus
- Blue nevus
- Intramucosal nevus

• **Intramucosal nevus**

Most pigmented skin tumors are composed of nevus cells and are a result of a developmental anomaly of melanocytes; **they are rare in the oral cavity.** The initial, flat, raised lesion can become nodular, with an increase in consistency. Spontaneous involution may occur and malignant transformation is a rare complication. When found intraorally they most frequently occur on the **hard palate (See picture #56 in booklet)** but can also be seen on the gingiva and lips. Intramucosal nevi are the most common variety found in the oral cavity followed by blue nevi. Compound and junctional nevi are **very rare.**

Subtypes of Acquired Nevi

- **Intradermal nevus** → most common lesion of **skin**. Known as the **common mole**. Nevus cells lie exclusively within the dermis.
- **Junctional nevus** → nevus cells are located at the interface between the epithelium and lamina propria. They are flat and not detected by palpation. Some regard as premalignant, may undergo transformation into **malignant melanoma.**
- **Compound nevus** → nevus cells are located at the epithelium/lamina propria interface and also deep in the dermis. They are raised and solid.
- **Blue nevus** → congenital, painless; color based on the deep cutaneous or subcutaneous / submucosal deposits of melanin.
- **Intramucosal nevus** → nevus cells are located in the connective tissue or lamina propria of the oral mucosa. Under palpation these nevi appear solid and are slightly raised over the surface of the mucosa.

Important: If a pigmented lesion shows ulceration, an increase in size, darkening in color, etc., a **biopsy** should be performed → this may indicate transformation into a **malignant melanoma.**

Pseudocyst

Traumatic bone cysts are non-cysts often found in:

- Infants
- Teenagers
- Middle aged adults
- Elderly people

• **Teenagers**

***The traumatic bone cyst has many names, which include the **simple** bone cyst, **hemorrhagic** bone cyst, **unicameral** bone cyst, **extravasation** bone cyst, **idiopathic** bone cyst and **solitary** bone cyst.

Traumatic bone cysts are non-cysts *(referred to as* **pseudocysts***)* often found in the mandible of teenagers that is assumed to be caused by trauma. It manifests as a **well-defined radiolucency,** which frequently extends **between the teeth and has a scalloped appearance.** The usual location for this cyst is in the mandible between the **cuspid and ramus.** Although sometimes asymptomatic, it may produce enlargement of the jaw. **Pain is rarely associated** with the lesion. The regional teeth are **vital. See picture #27 in booklet.**

The treatment of traumatic cysts is relatively easy. It consists of opening the lesion, curettage and closure. The resultant blood clot soon undergoes organization and the bone defect is soon repaired.

Note: This cyst is a closed compartment that has a connective tissue lining of varied thickness. It may contain blood, serosanguineous fluid, debris composed chiefly of a blood clot, or it may be completely devoid of solid material.

The following are also **not true cysts** → they are called "pseudocysts":
• Latent bone cyst
• Lingual mandibular concavity
• Aneurysmal bone cyst

All of the following statements are true concerning the **aneurysmal bone cyst, except:**

- It is an uncommon expansile osteolytic lesion of bone consisting of a proliferation of vascular tissue that forms a lining around blood filled cystic lesions
- Most aneurysmal bone cysts occur in patients under 20 years of age, and it is uncommon after the age of 30
- It commonly involves the jaws
- The lesions are usually tender or painful, particularly upon motion of the bone affected
- Upon entering the lesion surgically, excessive bleeding is encountered

• **It commonly involves the jaws**

***This is false;** it commonly involves the proximal humerus, femur, tibia and pelvis. It is not as common in the jaws, but it does appear here as well.

The aneurysmal bone cyst is a benign lesion of bone that is generally regarded as representative of a reactive rather than a neoplastic or cystic process. Upon entering the lesion during surgery, excessive bleeding is encountered and the tissue is often described as resembling a "blood-soaked" sponge.

Histologically, the aneurysmal bone cyst has no epithelial lining *(thus it is called a* **"pseudocyst"***).* It consists of a fibrous connective tissue stroma containing many cavernous or sinusoidal blood-filled spaces. **Fibroblasts and macrophages** *(histiocytes)* line the sinusoids. **Multinucleated giant cells,** similar to those of a giant cell granuloma, are dispersed throughout.

The radiographic picture of the lesion is often distinctive. The bone is expanded and appears cystic with a **"honeycomb"** or **"soap bubble"** appearance.

Surgical curettage or excision is the treatment of choice, with little chance of recurrence.

Notes:

1. Males and females are **equally** affected.

2. When the jaws are affected, the **mandible** is the most frequent site.

Median rhomboid glossitis frequently affects:

- Toddlers
- Infants
- Middle-aged adults
- Teenagers

• **Middle-aged adults**

This entity was once thought to be a congenital abnormality related to the persistence of the tuberculum impar, **however**, it is now believed that this condition is a permanent end result of a chronic **Candida albicans infection.** Diabetics, immunosuppressed patients and patients on long-term antibiotic therapy are more susceptible to this condition.

Median rhombold glossitis usually presents as a smooth, denuded, beefy, red lesion devoid of filiform papillae. The most common location is the midline of the dorsum of the tongue, just anterior to the circumvallate papillae. It is generally asymptomatic. Generally no treatment is necessary.

See picture #23 in booklet.

"Burning tongue syndrome" typically affects:

- Children
- Teenagers
- Middle-aged females
- Middle-aged males

- **Middle-aged females**

Patients with this syndrome usually exhibit **no clinically detectable lesions,** although symptoms of pain and burning can be intense. This syndrome is a particularly frustrating problem for both the patient and the clinician because there is usually no **clear-cut cause and no uniformly successful treatment.** This condition does affect men, but generally at a later age than females. It is rare in children and teenagers.

The following factors have been implicated as having possible etiologic significance to this syndrome:

- **Anemias**, such as pernicious anemia and iron deficiency anemia
- **Diabetes mellitus**
- **Gastric disturbances,** such as hyperacidity or hypoacidity
- **Psychogenic factors** → emotional conflict, cancerophobia
- **Trigeminal neuralgia**
- **Microorganisms** → especially C. albicans and Streptococci
- **Dry mouth** *(xerostomia)* → associated with Sjögrens syndrome, anxiety, and drugs
- **Local irritation** → tobacco, spices, etc.
- **Vitamin deficiency** → especially B complex

Which of the following is also known as **Rendu-Osler-Weber disease?**

- Hereditary hemorrhagic telangiectasia
- Encephalotrigeminal angiomatosis
- Juvenile nasopharyngeal angiofibroma

• **Hereditary hemorrhagic telangiectasia**

Rendu-Osler-Weber disease is a congenital hereditary form of hemangioma. It is characterized by numerous **spider-like telanglectases** on the face, neck, chest, lips, gingiva, buccal mucosa and tongue. One of the earliest signs of the disease is **epistaxis** *(nosebleeds)*.

Encephalotrigeminal angiomatosis *(Sturge-Weber disease)* is an uncommon congenital syndrome of unknown etiology *(sometimes classified as a variant form of hemangioma)*. It consists of a facial lesion, known as the **port-wine stain,** which is distributed over the trigeminal nerve accompanied by a similar vascular disorder of the underlying meninges and cerebral cortex. It usually occurs **unilaterally**.

Juvenile nasopharyngeal angiofibroma is a rare, benign neoplasm that nearly always affects adolescent males. It characteristically produces a mass in the nasopharynx that leads to obstruction or epistaxis. Treatment is surgery. Recurrences are common.

Which of the following is defined as a **persistent, velvety red patch** that cannot be characterized clinically as any other condition?

- Leukoedema
- Psoriasis
- Erythroplakia
- White sponge nevus

• **Erythroplakia**

The term **"erythroplakia"** like the term "leukoplakia" has **no histologic connotation;** however, the majority of erythroplakias are histologically diagnosed as severe epithelial dysplasia, carcinoma in situ or invasive squamous cell carcinoma.

Erythroplakias may be located anywhere in the mouth, but are **most likely** to be found in the mandibular mucobuccal fold, oropharynx and floor of the mouth. There is no sex predilection and patients over 60 years old are most commonly affected.
See picture #44 in booklet.

Remember: Carcinoma in situ exhibits all of the histologic characteristics of malignancy *(pleomorphism, hyperchromatism, abnormal mitoses, anaplasia, etc.)*, but does **not show** invasiveness or extension into adjacent structures.

The **most common site** for a pyogenic granuloma to occur is:

- The palate
- The mucogingival junction
- The interdental gingiva
- The tongue

• **The interdental gingiva**

See picture #6 in booklet.

Pyogenic granulomas may also occur on the lower lip, tongue and the buccal mucosa. They rarely occur on other areas of the oral mucous membrane. It is generally believed that the pyogenic granuloma arises as a result of some **minor trauma** to the tissues *(cementation of a crown, calculus, etc.)*, which provides a pathway for the invasion of nonspecific types of microorganisms.

Pyogenic granulomas present as **soft pedunculated** broad-based growths that have a smooth red surface. This red appearance is due to the presence of **hyperplastic granulation tissue,** which contains many capillaries. They are often ulcerated, bleed easily and may have a raspberry-like appearance. Treatment consists of excision. They are benign, but may occasionally recur.

Note: Pregnant patients are prone to these lesions *(sometimes called* **"pregnancy tumor"***)*.

The **peripheral giant cell granuloma** always occurs on:

- The buccal mucosa
- The gingiva or the alveolar process
- The lower lip
- The palate

• **The gingiva or the alveolar process**

See picture # 21 in booklet.

Peripheral giant cell granulomas represent an unusual hyperplastic connective tissue response to injury of gingival tissues. These granulomas are relatively uncommon and usually appear on the ginigiva in the area between the **first permanent molars and the incisors. Note:** The mandibular gingiva seems to be more affected.

These granulomas are **pedunculated** broad-based growths, which usually have a smooth surface. They are reddish-blue in color and sometimes lobulated. They usually bleed easily. The vast majority of patients are over 20 years of age. (**Remember:** *The central giant cell granuloma occurs predominantly* **before the age of 20**). Females are affected almost twice as frequently as males. Radiographs are usually negative. Histologic sections are **diagnostic** and are identical to those of a central giant cell granuloma. It consists of a non-encapsulated mass of tissue composed of a delicate reticular and fibrillar connective tissue stroma with **multinucleated giant cells.** Treatment is complete surgical excision.

Note: It may resemble a **fibroma** or a **pyogenic granuloma** clinically.

Which of the following statements are true concerning **hemangiomas?**

- Hemangiomas can range from being small to very large sacs of unsightly red, purple or blue blood vessels
- They can appear at birth or shortly thereafter, or later in life
- They can enlarge sometimes to very alarming sizes
- In certain locations large hemangiomas can interfere with proper development and function of organs
- All of the above statements are true

• **All of the above statements are true**

Hemangioma			
Histogenesis (Etiology)	**Clinical Characteristics**	**Microscopic Characteristics**	**Treatment and Prognosis**
Endothelial cells, Connective tissue origin.	Common tumor characterized by **proliferation of blood vessels.** Females 2:1; soft, smooth, blue, red, purple or purplish-red mass; the tongue, buccal mucosa, lips and palate are common sites; present at birth or arise in early life; enlarge as child grows	**Three types:** 1. Capillary 2. Cavernous 3. Hemangioendothelioma: stratified squamous epithelium covering, loose, fibrous connective tissue that contains many thin-walled, engorged vascular spaces.	Laser therapy or surgery; may regress spontaneously. **Incisional biopsy is contraindicated.**

See picture #45 in booklet.

All of the following are clinical features of **benign** salivary gland tumors, **except:**

- Mucosa is normal
- Painless
- Nodular
- Movable
- Fixed
- Firm
- Slow-growing

- **Fixed**

Benign Salivary Gland Tumors

- **Pleomorphic adenoma** *(mixed tumor)* is the **most common** benign salivary tumor

- **Monomorphic adenoma(s)**
  - Basal cell
  - Canalicular
  - Myoepithelioma
  - Sebaceous
  - Papillary cystadenoma lymphomatosum → Warthin's tumor
  - Oncocytoma → oxyphilic / acidophilic adenoma

Notes:

1. The most common site of intraoral **minor** salivary gland neoplasms is the **palate**

2. The most common site of intraoral **major** salivary gland neoplasms is the **parotid**

The most common site for **necrotizing sialometaplasia** is:

- The soft palate
- The buccal mucosa
- The hard palate
- The tongue

• **The hard palate**

Necrotizing sialometaplasia is a recognized lesion of the minor salivary glands characterized by necrosis of the glandular parenchyma with associated squamous metaplasia and hyperplasia of the ductal epithelium. The etiology is unknown, but it is believed to be related to vascular insufficiency and infarction of the glands.

The lesion shows no racial or sex predilection, and most of the patients are over 40 years of age. The **hard palate** appears to be the most common site. It presents as a tender ulcer, which usually is deep with sharply demarcated margins. Histologically, there is lobular necrosis of the glandular parenchyma, with squamous metaplasia and hyperplasia of the ductal epithelium. Both clinically and histologically, the lesion may simulate a malignancy and in the past, the condition has been **misdiagnosed** as a squamous cell or mucoepidermoid carcinoma.

Following biopsy and the establishment of the diagnosis, further treatment generally is **not recommended** since healing usually occurs within 6-12 weeks.

The most common **viral disease** of salivary glands is:

- Sialolithiasis
- Mumps
- Measles
- Candidiasis

- **Mumps**

Etiology: RNA-paramyxovirus group

Clinical:
- **90%** of the cases occur **before 14 years of age**
- A major sign is **sudden salivary gland swelling** without purulent discharge from the duct.
  ***Parotid** → 90% involvement and bilateral in two-thirds of the cases.
- Mild fever, malaise and anorexia
- Most cases are self-limiting

Complications:
- **Orchitis** *(inflammation of the testis)* and epididymitis can occur in post-pubertal males.
  ***Important:** May cause sterility
- CNS system → meningitis and encephalitis
- Deafness, myocarditis, pancreatitis, oophoritis and pyelonephritis

The **serum amylase** may be elevated during the acute phase. Prevention with a **live attenuated vaccine** is 95% effective for at least five years. However, in non-inoculated individuals, it is still a cause of acute **non-suppurative salivary adenitis.**

All of the following statements concerning **developmental salivary gland defects** are true, **except:**

- They are very common
- They are asymptomatic
- They are next to impossible to palpate manually
- They are discovered only during radiographic examination of the area

• **They are very common**

****This is false;** they are **relatively rare.**

The image of the developmental salivary gland defect is a **round or ovoid radiolucency** that ranges in diameter from 1 to 3 cm. It generally develops below the mandibular canal and above the inferior border of the mandible, just anterior to the angle of the jaw, and below and just behind the third molar.

See picture #12 in booklet.

There is increasing evidence that **Mikulicz's disease** is closely related to:

- Pierre-Robin syndrome
- Sjögren's syndrome
- Goltz-Gorlin syndrome
- Apert syndrome

• **Sjögren's syndrome**

The term **"benign lymphoepithelial lesion"** *(also called* **Mikulicz's disease***)* is manifested essentially as a progressive, asymptomatic enlargement of the parotid and submandibular glands. It is initially unilateral, but over time, it becomes bilateral. The etiology is unknown, however, there is increasing evidence that both Mikulicz's disease and Sjögren's syndrome are both actually autoimmune diseases in which the patient's own salivary gland tissue becomes antigenic.

The benign lymphoepithelial lesion is rare. It occurs most frequently in **middle-aged women.** Histologically, there is replacement of the gland parenchyma by lymphocytic infiltrate within which there are scattered **epimyoepithelial islands.** This is the histologic cornerstone for the diagnosis of the benign lymphoepithelial lesion.

Important: Most of these lesions remain benign, however, **malignant transformation of the epimyoepithelial islands** has been demonstrated.

Mucocele's most frequently occur on the:

- Upper lip
- Palate
- Lower lip
- Gingiva

• **Lower lip**

***They are seldom found on the upper lip

The **mucous retention cyst** *(also called a mucocele)* is generally conceded to be of traumatic origin. It involves the **minor salivary glands and their ducts** *(for example, trauma to the salivary duct by lip biting or pinching).*

A mucocele is a common lesion that may also appear on the palate, cheek, tongue and floor of the mouth. Clinically, the lesion may lie fairly deep in the tissue, or be exceptionally superficial, and depending upon the location, will present a variable clinical appearance. The **superficial lesion** appears as a raised, circumscribed vesicle, several millimeters to a centimeter in diameter, **with a bluish, translucent cast. The deeper lesion** appears as a fluctuant swelling also, but the tissue appears **normal in color.** Treatment is excision.

See picture #35 in booklet.

What is the **most probable diagnosis** for a lesion that presents as a translucent, bluish, well-rounded, smooth-surfaced bulge that protrudes from one side of the floor of the mouth?

- A mucocele
- Squamous cell carcinoma
- A ranula
- A lymphangioma

• **A ranula**

***It is **fluctuant and painless**

The **ranula**, a true retention cyst, characteristically occurs in the floor of the mouth and is unilaterally located. It arises in association with the secretory ducts of the submandibular or sublingual glands, and is usually caused by an obstruction produced by either a salivary stone or soft organic substance.

Of diagnostic significance is a history of **increased size just before or during a meal,** and a decrease in size between meals. The treatment is surgical, either through complete excision or by removing the roof of the cyst. If it persists, excision of the gland may be needed.

See picture #17 In booklet.

Occlusal radiographs are useful in locating:

- The hyoid bone
- The mental foramen
- Sialoliths in Wharton's duct
- Sialoliths in Stensen's duct

• **Sialoliths in Wharton's duct**

The deposition of **calculus** in the salivary ducts and glands becomes more common as we reach middle age. The **most common symptom** of an obstruction is an increase and decrease in swelling of the gland, particularly at mealtime. It may occur in children. Transillumination of the soft tissue is useful in detecting sialolithiasis in the child patient.

Note: The swelling may or may not be painful.

The rate of occurrence in **submandibular gland** and duct is much higher than in the parotid or sublingual areas. This is thought to be due to the tenacity of the submandibular saliva and the long and irregular shape of the duct. **See picture #36 in booklet.**

The treatment of choice is almost invariably **surgical extirpation of the sialolith.** Stones located in the glandular parenchyma usually require removal of the gland as well.

Notes:

1. Wharton's duct is also called the **submandibular duct**

2. A **sialolith** is a salivary calculus and is sometimes referred to as a **"salivary stone"**

The **most common tumor** of major and minor salivary glands is:

- Basal cell adenoma
- Sebaceous adenoma
- Pleomorphic adenoma
- Intraductal papilloma

• **Pleomorphic adenoma**

***It is also called **"benign mixed tumor"**

The term **"mixed tumor"** was used by investigators who believed the neoplasm was of both ectodermal and mesenchymal origin.

Clinical features: The pleomorphic adenoma is the most common salivary gland neoplasm. Women are affected more frequently than men, and the majority of patients are between 40 and 60 years old. Approximately 93% arise in the major salivary glands and these are almost exclusively parotid neoplasms *(84%)*. **They present as painless lumps below and anterior to the ear.** Approximately 7% arise in the oral cavity with the palate by far the most common intraoral site. Here they appear as firm, painless swellings, and in the vast majority of cases, do not cause ulceration of the overlying mucosa. **See pictures #47 and #48 in booklet.**

Histologic features: The epithelial component of a pleomorphic adenoma consists of round, polyhedral, elongated or stellate cells, which are relatively small and stain uniformly. The mesenchymal component varies from areas of myxomatoid tissue to areas of dense, hyalinized connective tissue, pseudocartilage or bone.

Treatment and prognosis: Pleomorphic adenomas are invariably encapsulated or well demarcated, and **surgical excision** with a generous margin of normal tissue is the treatment of choice. Inadequate initial removal of the mixed tumor in major glands may result in recurrence. Approximately 25% of benign mixed tumors will undergo malignant transformation if lesions are untreated for an extended length of time.

All of the following metabolic conditions are associated with chronic salivary gland enlargement, **except:**

- Diabetes mellitus
- Chronic alcoholism
- Malnutrition *(including anorexia and bulimia)*
- Hypothyroidism

- **Hypothyroidism**

Other metabolic conditions that are associated with salivary gland enlargement include obesity, hypertension and hyperlipidema.

Important: The parotid gland is most frequently enlarged. This can be unilateral or bilateral.

Other conditions associated with **parotid gland** enlargement:
- Sjögren's syndrome
- Sarcoidosis
- Warthin's tumor → also called papillary cystadenoma lymphomatosum
- Infections → for example, mumps, actinomycosis, tuberculosis
- Benign lymphoepithelial lesion → also called Mikulicz's disease
- Acute epidemic parotitis
- Malnutrition

Acinic cell carcinoma is a malignant salivary gland tumor that is **most likely** to be associated with the:

- Submandibular gland
- Parotid gland
- Minor glands of the palate

- **Parotid gland**

Acinic cell carcinoma:

- **Site: Parotid** *(96%)*, submandibular *(2-3%)* and minor glands *(1-2%)*.
- **Frequency:** 2-4% of parotid tumors
- **Clinical:** Swelling, **pain** or tenderness is **common**. May have facial weakness or paralysis.

Comment: 90% cure rate

Adenocarcinoma, NOS *(not otherwise specified)*:

- **Site:** Major glands *(50%)* and minor glands *(50%)*
- **Frequency:** 25% of minor, 5 % of submandibular and 3% of parotid tumors
- **Clinical:** Usually **asymptomatic mass**

Comment: 80% survival rate for low-grade carcinoma; 40% survival rate for high-grade carcinoma.

Which malignant salivary gland tumor **most often** occurs in the **minor salivary glands** of the palate?

- Adenoid cystic carcinoma
- Mucoepidermoid carcinoma

• **Adenoid cystic carcinoma**

Adenoid cystic carcinoma:

• **Site:** Minor glands *(70%,* **mostly palate***)*, parotid *(15%),* submandibular *(14%)*
• **Frequency:** 31% of minor gland, 14% of submandibular and 2% of parotid tumors
• **Clinical:** Pain and / or nerve dysfunction in one-quarter to one-third of patients
*****Facial weakness or paralysis is common**

Comment: Often slow but relentless in progression of disease; 20% twenty year survival.

Mucoepidermoid carcinoma:

• **Site:** Parotid *(60%)*, palate *(13%)* and submandibular *(6%)*
• **Frequency:** 10% of minor gland, 6% of parotid and 5% of submandibular tumors
• **Clinical:** Usually **asymptomatic** swelling; peak incidence in third decade
*****May have facial weakness or paralysis**

Comment: Low-grade carcinomas have an 85-100% 5-year cure rate; high-grade carcinomas have a 20-40% 5 year cure rate.

See picture # 46 in booklet.

All of the following are characteristic of Sjögren's syndrome, **except:**

- Rheumatoid arthritis
- Nephrosclerosis
- Xerostomia *(dry mouth)*
- Keratoconjuctiva sicca *(dryness of the eyes)*

• **Nephrosclerosis**

Sjögren's syndrome is a disorder of unknown cause, however, recent evidence suggests that it is autoimmune in nature. It is marked chiefly by chronic inflammation of the salivary glands and lacrimal glands. This usually progresses to fibrosis and atrophy of these glands. **All three symptoms rarely occur in one patient.** A definite diagnosis can be made only when at least two of the symptoms are present. The patients **most commonly affected** are post-menopausal women who present with dry eyes, dry mouth and, in about 50% of the cases, enlargement of the parotid and submandibular glands bilaterally.

Important: The histological features of the salivary gland lesions in both **Sjögren's syndrome** and the **"benign lymphoepithelial lesion"** *(also called Mikulicz's disease)* are identical.

All of the following are clinical features of **malignant** salivary gland tumors, **except:**

- Mucosa is ulcerated
- Painless
- Nodular
- Firm
- Fixed
- Rapid growth

- **Painless**

***Malignant salivary tumors are usually **painful.**

Malignant Salivary Gland Tumors

- Adenocarcinoma, NOS *(not otherwise specified)*
- Adenoid cystic carcinoma
- Mucoepidermoid carcinoma
- Acinic cell carcinoma

Oncocytomas are rare tumors that may occur:

- In the kidney
- In the salivary glands
- In endocrine glands
- All of the above

• **All of the above**

An **oncocytoma** *(also called an oxyphilic adenoma or acidophilic adenoma)* is a small benign, glandular tumor composed of large cells with cytoplasm that is granular and eosinophilic due to the presence of abundant mitochondria. Its development is believed to be related to the aging process.

Clinical features: Oncocytomas are rare tumors seen most frequently in the **parotid glands** of individuals over 50 years of age, and are slightly more common in women. Their growth is slow and they seldom attain any significant size.

Histologic features: The tumor is an encapsulated mass composed of relatively large cells with bright pink cytoplasm and small round nuclei. The cells may be arranged in sheets or cords, or form tubular or acinar structures.

Treatment and prognosis: The treatment of choice is surgical excision and recurrence is rare.

Note: Sialoscintigraphy is a simple and non-invasive procedure that can usually separate benign entities like Warthin's tumor and oncocytoma of the salivary glands from malignant tumors, and significantly affect the course of treatment.

Warthin's tumor is almost exclusively seen in the:

- Submandibular gland
- Sublingual gland
- Parotid gland

• **Parotid gland**

Papillary cystadenoma lymphomatosum *(a fancy name for* **Warthin's tumor***)* is almost exclusively a **parotid** neoplasm. It is believed to arise from heterotopic ductal epithelium within lymph nodes or near the parotid gland.

Clinical features: The vast majority of patients are over 50 years of age, with a 5:1 male predominance. Approximately 5% of the tumors are bilateral. The tumor most often arises in the lower pole of the parotid and presents clinically as a **non-tender**, slowly enlarging, firm to fluctuant nodule over the **angle or ramus** of the mandible.

Histologic features: The tumor is encapsulated and composed of cystic spaces containing an eosinophilic coagulum into which extend papillary projections of the lining epithelium. The epithelium consists of a double row of cells with eosinophilic, granular cytoplasm, a luminal layer of tall columnar cells and a basal layer of round, cuboidal or polygonal cells. Interspersed among the cystic spaces are aggregates of lymphoid tissue, some with germinal centers.

Treatment and prognosis: Surgery is the treatment of choice and recurrence is uncommon. Malignant variants of the tumor have been reported but are rare.

The treatment of choice for **Sjögren's syndrome** is:

- Radiation
- Surgical excision
- Chemotherapy
- None of the above

• **None of the above**

The treatment of Sjögren's syndrome is mainly **symptomatic**. The keratoconjunctivitis is treated with ocular lubricants and the xerostomia is treated by saliva substitutes.

Note: Biopsy of the labial or palatal salivary glands may be helpful in establishing the diagnosis, along with sialograms, salivary flow rate tests and blood work.

It is important to remember that **malignant lymphomas** and **"pseudolymphomas"** *(also called atypical benign lymphoid hyperplasia)* develop in some patients who have been diagnosed with Sjögren's syndrome. **This mandates close follow-up of the patients.**

Important: The decrease in salivation may cause **rampant caries** reminiscent of radiation caries.

The general characteristics below describe which type of neoplasm, **benign** or **malignant?**

- Invasion
- Immovable
- Rapid growth
- Metastasis
- Not well-differentiated *(or anaplastic)*

- **Malignant**

General characteristics of **benign neoplasms:**
- Well-differentiated
- Slow growth
- Encapsulated / Well–circumscribed
- Localized
- Movable

Important:
- **Paresthesia** is suggestive of **metastatic** disease.
- **Metastasis** is the most important characteristic that distinguishes malignancy from benign.

Radiographically, a **benign** neoplasm in bone may be differentiated from a **malignant** neoplasm in the following ways:
- In a **benign** lesion, the cortex tends to **remain intact** but may be thinned and the part involved expanded.
- In a **benign** lesion, the margins are usually **defined** and demarcated from surrounding bone.

Which term is a developmental defect characterized by an overgrowth of tissue **not normal** to the organ in which it arises?

- Teratoma
- Choristoma
- Hamartoma

- **Choristoma**

Terms used in **oncology**:
- **Oncology:** the study of neoplasms
- **Neoplasm:** an uncontrolled new growth of tissue
- **Tumor:** a localized swelling, may or may not be a true neoplasm
- **Hyperplasia:** an increase in the size of a tissue or organ due to an increase in the **number** of component cells
- **Hypertrophy:** an increase in the size of a tissue or organ due to an increase in the **size** of component cells
- **Cancer:** a general term for all **malignant** neoplasms
- **Carcinoma:** a malignant **epithelial** neoplasm
- **Sarcoma:** a malignant **mesenchymal** *(connective tissue)* neoplasm
- **Hamartoma:** a developmental defect characterized by an overgrowth of tissues **normal** to the organ in which it arises
- **Teratoma:** a **neoplasm** composed of multiple tissues **foreign** to the organ in which it arises; may be benign or malignant

ORAL PATHOLOGY

Terms

All of the following are **histological** characteristics of malignancy, **except:**

- Abnormal mitosis
- Metaplasia
- Pleomorphism
- Hyperchromatism
- Anaplasia
- Increased nuclear-cytoplasmic ratio

- **Metaplasia**

Remember: Metaplasia is the process whereby one cell type changes to another cell type **in response to stress** and generally assists the host to adapt to the stress.

Histological grading of **malignant** neoplasms:

- An attempt to estimate the aggressiveness of degree of malignancy of a malignant neoplasm based upon the **degree of differentiation** of the component cells and the number of mitoses.
  - Grade 1. **Well**-differentiated
  - Grade 2. **Moderately well**-differentiated
  - Grade 3. **Poorly** undifferentiated
  - Grade 4. **Undifferentiated**

- Applicable mainly to **squamous cell carcinomas** and of limited clinical usefulness. Most pathologist use **three grades** and prefer to designate squamous cell carcinomas as well-differentiated, moderately well-differentiated or poorly differentiated.

ORAL PATHOLOGY

Terms

Which of the following is a characteristic of **malignancy?**

- Dysplasia
- Anaplasia
- Metaplasia

• **Anaplasia**

***Differentiation is a measure of a tumor's resemblance to normal tissue. **Anaplasia** is the **absence of differentiation.**

Histologic features of malignancy:

• Anaplasia • Hyperchromatism • Pleomorphism • Abnormal mitosis

The host response to a **malignancy** is best reflected by lymphocytic infiltration at the edge of a tumor. The most characteristic feature of a malignancy as opposed to an inflammatory lesion is that a malignancy **will grow after removal of the causative agent.** The most important characteristic of malignant neoplasms, which distinguishes them from benign neoplasms, is their **ability to metastasize.**

Notes:

1. **Dysplasia** is a type of non-malignant cellular growth, but may precede neoplastic changes in the tissue. It is associated with chronic irritation of a tissue by a chemical agent, such as cigarette smoke or by chronic inflammatory irritation, such as chronic cervitis. The tissue appears somewhat structureless and **disorganized** and may consist of **atypical cells** without invasion. Epithelium exhibits **acanthosis** *(which is an abnormal thickening of prickle cell layer).*
2. **Metaplasia** is the process whereby one cell type changes to another cell type in**response to stress** and generally assists the host to adapt to the stress. The most common type of epithelial metaplasia involves replacement of columnar cells by stratified squamous epithelium.

The term **iatrogenic** refers to:

- A disease or condition of unknown cause or origin
- A condition caused by medical personnel or procedures
- A tissue or an organ transferred into a new position within the body of the same individual
- Any of the above

- **A condition caused by medical personnel or procedures**

The term iatrogenic originally applied to disorders induced in the patient by auto-suggestion based on the physician's examination, manner, or discussion. **The term is now applied** to any adverse condition in a patient occurring as the result of treatment by a physician or surgeon, especially to infections acquired by the patient during the course of treatment.

Other terms to remember:

- **Idiopathic** → a disease or condition of unknown cause

- **Autograph** *(autologous graph)* → a tissue or an organ transferred into a new position within the body of the same individual

- **Nosocomial** → pertaining to or originating in the hospital; said of an infection not present prior to admittance to the hospital, but generally occurring 72 hours after admittance. The term is usually used to refer to **patient disease,** but hospital personnel may also acquire a nosocomial infection.

Recurrent **aphthous** ulcers:

- Are caused by the Epstein-Barr virus
- Are caused by the Herpes Simplex virus
- Appear to be associated with stress
- Are caused by a Poxvirus

• **Appear to be associated with stress**

***Recurrent aphthous ulcers are commonly referred to as **"canker sores"** by lay persons. In the literature other terms include aphthous stomatitis, recurrent aphthous stomatitis, recurrent ulcerative stomatitis or ulcerative stomatitis.

The stress factors may include:
• Bacterial infection
• Trauma *(i.e., self-inflicted, oral surgery procedures, routine dental procedures)*
• Endocrine conditions *(i.e., a females menstrual period)*
• Allergic factors *(i.e., certain foods or drugs)*
• Immunologic abnormalities
• Iron, Vitamin B or folic acid deficiencies

***The cause is unknown,** but there is evidence to suggest that the occurrence of these aphthous ulcers is an autoimmune reaction.

Three classifications:
1. Recurrent aphthous minor
2. Recurrent aphthous major
3. Recurrent herpetiform

Remember: Vesicles **do not** precede the formation of the ulcers.

A severe bullous form of **erythema multiforme** is known as:

- Plummer-Vinson syndrome
- Peutz-Jeghers syndrome
- Gardener's syndrome
- Stevens-Johnson syndrome

• **Stevens-Johnson syndrome**

In Stevens-Johnson syndrome, the systemic symptoms are **severe** and the lesions are **extensive**, involving multiple body areas *(especially the mucous membranes)*. This syndrome is characterized by the acute onset of fever along with eruptive, ulcerative lesions of the skin, oral mucosa and eyes. Frequently the genitalia, lungs, and joints are affected. It can have a fatal termination.

The typical **"bull's-eye-shaped "** lesions are present. The classical triad of this syndrome consists of **eye lesions, genital lesions and stomatitis**. The treatment consists of IV fluids, systemic steroids, palliative rinses and antibiotics.

Note: Blindness can occur due to secondary infection.

Important: The lesions of Stevens-Johnson syndrome are **severe** and often **vesicular or bullous,** with hemorrhage after denudation.

The **most common form** of recurrent aphthous stomatitis is:

- Recurrent aphthous minor
- Recurrent aphthous major
- Recurrent herpetiform

• **Recurrent aphthous minor**

There are **three forms** or classifications of recurrent aphthous stomatitis *(which is also called recurrent aphthous ulcers, canker sores, etc.)*

1. **Recurrent aphthous minor** → is the **most common** form of the disease and the one referred to by the lay public as the **"canker sore."** It occurs somewhat more frequently in women than in men. It begins as a single or multiple superficial erosion covered by a gray membrane. The lesion is typically **very painful.** The lesions vary in size from 2-3 mm to over 10 mm in diameter. They generally persist for 7-10 days and heal gradually with **little or no** evidence of scarring.
2. **Recurrent aphthous major** → is characterized by the occurrence of **large, painful ulcers,** usually one to ten in number. These ulcers occur at frequent intervals and many patients with this disease are seldom free from the presence of at least one ulcer. Unlike the minor aphthous ulcer, these lesions persist for up to six weeks and **leave a scar** upon healing.
3. **Recurrent herpetiform** → is characterized by **crops of multiple, small, shallow ulcers,** often up to 100 in number, which may occur at any site in the oral cavity. These lesions are present almost continuously for one to three years, with relatively short remissions.

Remember: Vesicular lesions **do not precede** the formation of ulcers in all of the above. **This is a distinctive diagnostic feature.**

Note: In healing of an ulcer, the epithelium that eventually will cover the defect is derived from intact epithelium at the ulcer margin.

The characteristic lesion of **erythema multiforme** is:

- Melanin pigmentation
- A chancre
- The "bull's-eye-shaped" lesion
- A blister

• **The "bull's-eye-shaped" lesion**

Erythema multiforme is a type of hypersensitivity *(allergic)* reaction that occurs in response to medications, infections, or illness. Medications associated with erythema multiforme include sulfonamides, penicillins, barbiturates, and phenytoin. Associated infections include herpes simplex and mycoplasma infections. The exact cause is unknown. The disorder is believed to involve damage to the blood vessels of the skin with subsequent damage to skin tissues. It occurs primarily in children and young adults. The diagnosis of erythema multiforme is primarily based on the **classic skin lesion** appearance.

Important: The **characteristic lesion** is the target *(or so-called* **"bull's-eye-shaped"***)* lesion which appears as a central lesion surrounded by concentric rings of pallor and redness over the dorsal aspect of the hands and forearms.

A low-grade fever, general malaise and headache usually precede the appearance of the lesions by 4 to 7 days. **Oral lesions** appear as red macules, papules or vesicles that may become eroded and painful. **Note:** These lesions are covered by a yellowish-white membrane after rupturing. **See picture #66 in booklet.**

The **treatment** consists of topical palliative rinses and in some instances, low dose systemic steroids.

Which of the following infections is the one most likely to result in a **chronic suppurative lesion** about the jaws?

- Actinomycosis
- Blastomycosis
- Histoplasmosis

• **Actinomycosis**

Actinomycosis is a subacute-to-chronic bacterial infection with Actinomyces, usually **A. israelii.** It is characterized by contiguous spread, suppurative and granulomatous inflammatory reaction, and the formation of **multiple abscesses** and sinus tracts that **discharge sulfur granules**. The most common clinical forms include: **cervicofacial** *(i.e., lumpy jaw)*, thoracic, and abdominal actinomycosis.

Cervicofacial actinomycosis or *lumpy jaw* is the most common manifestation of actinomycosis. Infection typically occurs in patients with poor dental hygiene or following surgery. This form is characterized in the initial stages by soft tissue swelling of the perimandibular area. Direct extension into the adjacent tissues occurs over time, along with the development of fistulas that discharge purulent material containing yellow *(i.e., sulfur)* granules. **Note:** These granules are actually colonies of infecting organisms. **See picture #58 in booklet.**

Remember: Actinomyces are gram-positive filamentous bacteria that are normal inhabitants of the oral cavity and GI tract.

Histoplasmosis is a disease caused by the fungus Histoplasma capsulatum. Its symptoms vary greatly, but the disease primarily affects the lungs. Occasionally, other organs are affected *(this form of the disease is called* **disseminated histoplasmosis***)*. The infection is usually asymptomatic, but may produce a benign, **mild pulmonary illness** *(which is the primary form of disease)*. **Oral manifestations include** nodular, ulcerative, or vegetative lesions on the buccal mucosa, gingiva, tongue, palate or lips. The lesions are usually covered by a nonspecific gray membrane and are indurated.

All of the following statements concerning **syphilis** are true, **except:**

- It is a sexually transmitted disease caused by a spirochete called Treponema pallidum
- It occurs in three stages: primary, secondary and tertiary
- The first symptom of primary syphilis is an ulcer called a chancre
- Secondary syphilis is often marked by a skin rash that is characterized by brown sores about the size of a penny
- In its late stages, untreated syphilis, although not contagious, can cause serious heart abnormalities, mental disorders, blindness, other neurologic problems, and death
- Syphilis is usually treated with tetracycline, administered by injection

- **Syphilis is usually treated with tetracycline, administered by injection**

***This is false;** syphilis is usually treated with **penicillin**, administered by injection.

Three stages of syphilis:

1. **Primary** → the first symptom is a non-painful **chancre** that generally appears 2-6 weeks after exposure. It usually is found on the part of the body exposed to the partner's ulcer, such as the penis, the vulva, or the vagina. It can also develop on the cervix, tongue, lips, or other parts of the body. The chancre disappears within a few weeks whether or not a person is treated. If not treated during the primary stage, about one-third of people will progress to chronic stages.
2. **Secondary** → is a highly **infectious** stage; it occurs 6 weeks after non-treatment of primary syphilis. Widely disseminated spirochetes cause mucous membranes to exhibit a **reddish-brown maculopapular cutaneous rash** and ulcers that are covered with a mucoid exudate *(called mucous patches).* **Condylomata lata** *(which are elevated broad-based plaques)* are also seen on skin and mucosal surfaces.
3. **Tertiary** → occurs in infected persons many years after non-treatment of secondary syphilis. The **gumma** *(which is a focal nodular mass)* typifies this stage. It most commonly occurs on the palate and tongue. The bacteria damage the heart, eyes, brain, **nervous system**, bones, joints, or almost any other part of the body. **Note:** Headache, stiff neck, and fever are symptoms of **neurosyphilis**.

The term used to describe **permanent incisors** which may be **characteristic of congenital syphilis** is:

- Blandi's incisors
- Turner's incisors
- Franklin's incisors
- Hutchinson's incisors

• **Hutchinson's incisors**

***They are generally peg-shaped, widely spaced, and notched at the end with a centrally placed crescent-shaped deformity.

Congenital syphilis is caused by infection with the spirochete Treponema pallidum during the fetal period. Expectant mothers who have syphilis can transmit the disease through the placenta to the unborn infant. Nearly half of all infants infected with syphilis during gestation die shortly before or after birth.

The severity of congenital syphilis depends upon **1)** The time in which the organisms pass the placental barrier *(protected up to 16th week)* **2)** The mother's stage of syphilis and **3)** The immunologic response of the fetus. If treated by the 4th or 5th month, 95% show no manifestations; if not treated, fetal sepsis may result in stillbirth or visceral and mucocutaneous manifestations.

1. **Symptoms in the newborn**
   - Irritability
   - Bloody discharge from the nose
   - **Early rash:** small blisters or flat or bumpy rash on the face, palms and sole
   - Failure to thrive
   - **Later rash:** copper-colored, vesicles on the palms and soles
   - Saddle nose, frontal bossing, short maxilla and high
2. **Symptoms in older infant and young child**
   - Bone pain
   - Joint swelling
   - Abnormal teeth *(Hutchinson's incisors)*
   - Gray, mucous-like patches on the anus and vulva *(condyloma lata)*
   - Saber shins *(bone abnormality of the lower leg)*
   - Visual loss, CN VIII nerve deafness and interstitial keratitis
   - Scarring of the skin around earlier lesions of the mouth, genitalia, and anus *(called rhagades)*

A person with **mucormycosis** generally is treated with:

- Penicillin
- Erythromycin
- Amphotericin B
- Clindamycin

• **Amphotericin B**

***Given IV or injected directly into the spinal fluid.

Mucormycosis *(also called phycomycosis or zygomycosis)* is an infection caused by a fungus belonging to a large group of organisms called Mucorales *(water mold).* Mucormycosis of the nose and brain *(rhinocerebral mucormycosis)* is a severe and usually fatal infection. This form is seen primarily in patients with chronic debilitating diseases, especially **uncontrolled diabetes mellitus**. The symptoms include pain, fever, and an infection of the eye socket *(orbital cellulitis)* with a bulging of the affected eye *(proptosis).* The nasal septum, the palate, or the sinuses may be destroyed.

A brain infection may cause convulsions, an inability to speak properly, and partial paralysis.

An acute infectious disease caused by a **group A coxsackie virus** is called:

- Herpes zoster
- Chickenpox
- Herpangina
- Herpes labialis

• **Herpangina**

Herpangina refers to a **stomatitis** *(inflammation of the mouth)* caused by a strain of the Coxsackie virus. It is differentiated in clinical practice form Type 1 Herpes infection *(the cold sore virus)* by the fact that the ulcerations of herpangina generally occur in the back of the throat around the tonsils and rear portion of the palate. **Remember:** Herpes Type 1 lesions are found typically more forward in the mouth on the tongue, gingiva, buccal mucosa and appear as vesicles *(small, clear blisters that ulcerate and crust)* around the mouth and on the lips.

The clinical manifestations of herpangina are **comparatively mild and of short duration.** It begins with a sore throat, fever, headache and sometimes vomiting and abdominal pain. Papules or vesicles soon form in the pharynx and evolve into shallow ulcers, which heal spontaneously. The disease usually runs its course in less than a week. The treatment is pallative.

Note: Herpangina affects **young children.** The oral lesions *(vesicles)* can also appear on the tongue. In **hand, foot and mouth disease,** the oral lesions *(vesicles)* appear on the buccal mucosa, tongue, gingiva and lips.

See picture #9 in booklet.

All of the following are oral or para-oral presentations of herpes simplex, **except:**

- Acute herpetic gingivostomatitis
- Herpes digitalis
- Recurrent herpetic stomatitis
- Herpes labialis

- **Herpes digitalis**

Oral and para-oral presentations of **Herpes Simplex Type 1** include:

- **Herpes labialis** *(also called fever blisters or cold sores)* is an extremely common disease caused by the herpes simplex virus Type 1, characterized by an eruption of small and usually painful blisters on the skin of the lips, mouth, gingiva, or the skin around the mouth. **Note:** The reason most patients suffering from recurrent herpes labialis rarely give a history of having had acute herpetic gingivostomatitis is that the primary infection was subclinical.
- **Acute herpetic gingivomatitis** *(also known as* **primary** *herpetic gingivostomatitis)* generally affects children under the age of three and young adults. There are **prodromal symptoms** *(fever, malaise, irritability, headache, dysphagia, vomiting, lymphadenopathy)* 1-2 days prior to local lesions. Then small, yellowish **vesicles** form, which rupture quickly, resulting in shallow, round, discrete ulcers with an erythematous halo. It affects both the **free** and **attached mucosa.** A generalized **marginal gingivitis** may precede the ulcers. Treatment includes fluid intake, good oral hygiene and gentle debridement of the mouth. In healthy individuals the lesions heal spontaneously in 7-14 days without a scar.
- **Recurrent** *(secondary)* **herpetic stomatitis** generally occurs in adult patients and is triggered by trauma, fatigue, URI, stress, allergy or UV exposure, which causes the release *(or reactivation)* of the latent HSV-1 virus. This reactivation causes a recurrent infection *(i.e., cold sores)* on the **lips** *(that is bound to periosteum),* hard palate, attached gingiva and alveolar ridge. **Site-specificity is a characteristic manifestation.**

Primary herpes is most common in:

- Middle age adults
- Elderly people
- Children and young adults
- Infants

• **Children and young adults**

Primary herpes is most common in children and young adults. Patients develop fever, irritability, regional lymphadenopathy and headache. Within days, the **gingiva** becomes intensely inflamed. Any part of the **oral mucosa and lips** may become involved. Vesicles then form and rupture a short time later to leave shallow ulcers covered with a gray membrane and surrounded by a red halo. These ulcers are **very painful**. These ulcers will heal on their own within 7 to 14 days.

After recovery from primary HSV infection, the virus is not cleared from the body, but, rather, it lies dormant in a non-replicating state, in the sensory nervous system *(specifically, the* **trigeminal ganglion***)*. Periodically, latency reactivates, allowing the virus to return to the skin or mucous membranes, where it causes a recurrent infection. **Cold sores** are a manifestation of **recurrent herpes simplex virus** infection around the mouth. The most common site is on the **lips**. Some factors that are often associated with a recurrent outbreak are: sunburn, fatigue, emotional upset, trauma, upper respiratory tract infection or menstruation. Often a day before the formation of vesicles there will be a tingling or itching of the skin or mucosa. Vesicles ulcerate and resolve the same as in the primary infection.

Histologically, the cytopathic effect *(CPE)* take the form of ballooning degeneration of the epithelial cells with loss of cohesion to adjacent cells. The nuclei are often multiple with margination of the chromatin around the intra-nuclear inclusions called **Lipschultz bodies**. These changes can be seen in scrapings taken from an unroofed vesicle *(these scrapings are called a* **Tzanck smear***)*.

All of the following tests can be used to diagnose herpetic lesions, **except:**

- The Tzanck smear
- Fluorescent staining
- An arthrogram
- Isolation in tissue culture
- Antibody titers
- Biopsy

- **An arthrogram**

1. **The Tzanck smear** is a **cytologic** examination of fluid harvested from an unopened vesicle, stained with giemsa, and viewed by the light microscope. The pathologist looks for epithelial cells with intranuclear inclusions *(these inclusions are called* **Lipshultz bodies***)*.
2. **Fluorescent staining** → cells show positive fluorescence when stained with fluorescent labeled HSV immune serum and globulin. This procedure is used to distinguish between herpes zoster and herpes simplex.
3. **Isolation in tissue culture**
4. **Antibody titers** *(anti HSV antibody titers)* → is a test for complement fixing or neutralizing antibody in acute and convalescent sera as well as on tissue sections *(this begins in one week and peaks in three weeks)*.
5. **Biopsied material** will show an intraepithelial cleft covered by an exudates of fibrin and polymorphonuclear leukocyte. The epithelium will exhibit **degenerative cells**, which include bizarre giant cells and cells with displaced chromatin with perinuclear halos and inclusions.

The treatment for herpes is **primarily supportive**:
- Analgesics
- Topical anesthetics prior to eating
- Maintain electrolyte balance
- Antiviral agents

Which virus can cause **herpes zoster** lesions along sensory nerve roots in later life?

- Poxvirus
- Epstein-Barr virus
- Varicella virus

• **Varicella virus**

It is a member of the **herpes virus** group. It causes the disease **chickenpox** *(varicella)* and **shingles** *(herpes zoster)*. The virus is very contagious and may be spread by direct contact or droplets.

Chickenpox is primarily a **disease of childhood,** which peaks at school-age in the winter and spring. It is characterized by the appearance on skin and mucous membranes of successive crops of typical **pruritic vesicular lesions** that are easily broken and become scabbed. It is generally accompanied by mild constitutional symptoms *(fever, malaise)*. It is **most contagious** one day before the onset of the rash and until all the vesicles have crusted. It is relatively benign in children, but adult infection may be complicated by pneumonia and encephalitis. **Note: ZIG** *(Zoster Immune Globulin)* reaches morbidity in high-risk children.

Shingles *(herpes zoster)* is the result of **reactivation of a latent varicella-zoster** virus that may have remained within the body from a childhood case of chickenpox. The virus reaches the sensory ganglia of the spinal and cranial nerves, producing an inflammatory response. It is characterized by **painful vesicles** that occur on the skin or mucosal surfaces **along the distribution of a sensory nerve.**

Note: The histology for both chickenpox and shingles shows the **same cytopathic effect** as seen in herpes simplex.

Herpes simplex is related to viruses that cause all of the following, **except:**

- Mononucleosis
- Chickenpox
- Mumps
- Shingles

• **Mumps**

Herpes simplex is one of the most common viral diseases effecting man. The primary infection, which is known as **primary herpetic gingivostomatitis**, is most common in young children *(under five years old).* It usually occurs in a child who has had no contact with the Type 1 herpes simplex virus, and who therefore has no neutralizing antibodies. It may also affect young adults *(15-25).* Nearly all primary infections are of the subclinical type *(they may only have* **flu-like symptoms***)* and one or two mild sores in the mouth which go unnoticed by the parents.

In other children, the primary infection may be manifested by acute symptoms, which is known as **acute herpetic gingivostomatitis**. These symptoms include fever, irritability, cervical lymphadenopathy, fiery red gingival tissues and small yellowish vesicles that rupture and result in painful ulcers on the free and attached mucosa. The most serious potential problem in a child with this infection is dehydration due to the child not wanting to eat or drink because of the pain. **See picture # 34 in booklet.**

The treatment is **supportive** and aimed toward the relief of the acute symptoms so that fluid and nutritional intake can be maintained. Primary herpetic gingivostomatitis usually runs a course of 12-20 days, and the ulcers heal without scarring.

Note: Corticosteriods are contraindicated in patients with herpes simplex infections.

After the initial primary attack during the early childhood period, the **herpes simplex** virus remains inactive most commonly in the:

- Geniculate ganglion
- Ciliary ganglion
- Trigeminal ganglion
- Pterygopalatine ganglion

• **Trigeminal ganglion**

The inactive herpes simplex virus resides in **sensory nerve ganglia** *(most commonly, the trigeminal ganglion),* but will often reappear later as the familiar **"cold sore"**, usually on the outside of the lips. This disease is referred to as **"recurrent herpes labialis"**. Emotional stress, trauma and excessive exposure to sunlight have been implicated as factors for the appearance of the recurrent herpetic lesions on the lip. **Acyclovir 5%** ointment *(Zovirax)* has been successful in reducing the duration and severity of these sores.

Remember:

1. **Herpes Simplex Type I** *(primary herpetic gingivostomatis, recurrent herpes labialis)* is transmitted by direct contact. It affects the lips, face, skin and oral mucosa.

2. **Herpes Simplex Type II** *(herpes genitalis)* is spread by sexual contact. It affects the mucosa of the genital and anal regions.

Note: Genital herpes may have serious consequences in pregnant women because the virus can be transmitted to the infant during vaginal delivery. The virus can cause damage to the infant's central nervous system and / or eyes.

Remember: The **primary** infection of herpes simplex can range from subclinical *(asymptomatic)* to severe systemic infections.

A chronic skin disease that is characterized by the formation of **vesicles and bullae** is called:

- Lichen planus
- Candidasis
- White sponge nevus
- Pemphigus

• **Pemphigus**

Pemphigus is a rare, bullous skin disease produced by dyhesion *(or* **acantholysis***)* of the epidermal cells, which is brought about by an **autoimmune mechanism** where antibodies attack the intercellular junctions of the epithelium. It seldom occurs before the age of 30 *(usually between 30 and 50)* and occurs more frequently in Jewish people.

There are **four types** of Pemphigus:

1. Pemphigus vulgaris → most common
2. Pemphigus vegetans
3. Pemphigus follaceus
4. Pemphigus erythematosus

Oral lesions are often the first manifestation of the disease. Intact bullae are rarely seen in the oral cavity, instead, large areas of ulceration and erosions are often seen that are covered by a white or blood-tinged exudates. Sometimes, areas of epithelium will slide off simply by rubbing of an apparently unaffected area *(this is termed* **Nikolsky's sign***)*. This disease is often fatal without therapy, which includes high-dose systemic steroids or chemotherapy *(for example,* **methotrexate***)*.

Important histological features: The vesicles and bullae are formed entirely intraepithelially, just above the basal layer of cells *(called* **suprabasilar** *vesicles)*. There is intercellular edema and loss of intercellular bridges with loss of cohesiveness. This is called **acantholysis**. Clumps of cells are often found floating free in the vesicle space *(these cells are called* **Tzanck cells***)*.

All of the following statements concerning **benign mucous membrane pemphigoid** are true, **except:**

- It is a chronic, self-limiting, mucocutaneous disease
- It is usually limited to the oral and ocular mucous membranes
- It usually effects women under the age of 20
- It is more common in the oral cavity than pemphigus, but is associated with much less morbidity and mortality

• **It usually affects women under the age of 20**

***This is false;** It usually effects women over the **age of 50.**

Benign mucous membrane pemphigoid *(BMMP)* is a vesiculobullous disease, probably **autoimmune** in nature, that occurs more often than is realized due to improper diagnosis. The oral lesions most commonly present as a **"desquamative gingivitis"** in which vesicles form, rupture and leave gingival erosions. Systemic steroid therapy has provided adequate management of BMMP. **Note:** Conjunctival involvement may lead to blindness.

Remember: Nikolsky's sign, which is an indication of **pemphigus vulgaris,** may also be found in BMMP. The sign occurs when apparently normal epithelium may be separated at the basal layer and rubbed off when pressed with a sliding motion.

Important: Histologically, the major difference between BMMP and pemphigus vulgaris is that the vesicles in BMMP are **subepidermal** and there is no **evidence of acantholysis** → in pemphigus vulgaris there is **acantholysis and a suprabasilar vesicle.**

The etiology of **verucca vulgaris** is:

- Fungal
- Bacterial
- Viral
- Parasitic

• **Viral**

****Specifically a **papilloma virus**

Verruca vulgaris *(also called squamous papilloma)* has an incubation period from about six weeks to a year. Although it is primarily a lesion of the skin, it may occur in the oral cavity, particularly on the **lips** and **palate**. Clinically, it is a **sessile, soft, cauliflower-like** lesion. Microscopically it is a papillomatous lesion in which the epithelium is thrown into folds. The lesion shows alternating hyperkeratosis, parakeratosis along with long epithelial ridges. If excised, they usually do not recur, but **autoinoculation** is possible. **Note:** Intraorally, that is how most cases develop.

See picture #26 in booklet.

The etiology of **verruciform xanthoma** is:

- Bacterial
- Viral
- Fungal
- Unknown

- **Unknown**

Benign Soft Tissue Tumors				
Tumor	**Histogenesis *(Etiology)***	**Clinical Characteristics**	**Microscopic Characteristics**	**Treatment and Prognosis**
Papilloma	Squamous epithelium *(epithelial origin)*	Sessile or pedunculated, whitish, cauliflower-like mass; tongue, lips, soft palate common sites	Finger-like projections of stratified squamous epithelium supported by thin cores of vascular fibrous connective tissue	Conservative excision; recurrence rare
Fibroma *(Irritation or traumatic fibroma)*	Fibrous connective tissue (connective tissue origin)	Smooth, sessile, soft to firm nodule; buccal mucosa, lips, tongue common sites	Bundles of interlacing collagen fibers	Conservative excision; recurrence rare
Lipoma	Adipose tissue *(connective tissue origin)*	Smooth or lobulated, sessile of pedunculated, soft yellowish mass; vessels visible over surface; floor of mouth, buccal mucosa, tongue common sites	Lobules of mature fat separated by delicate connective tissue septae	Conservative excision; recurrence rare
Verruciform xanthoma *(Histiocytosis "Y")*	Unknown; not associated with any systemic condition	Aduts; alveolar and palatal mucosa common sites; normal or white colored verrucous lesion	Verrucous, hyperparakeratotic surface with parakeratotic plugging; large "foam" cells in connective tissue papillae between elongated rete ridges	Simple excision; no recurrence

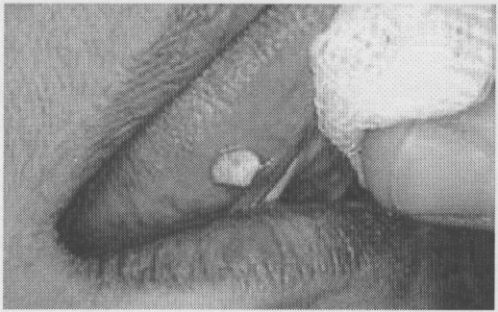
Inflammatory papillary hyperplasia is found on the:

- Soft palate
- Buccal mucosa
- Hard palate
- Tongue

- **Hard palate**

Entity	Etiology	Location	Clinical Features	Treatment
Inflammatory fibrous hyperplasia: **"Epulis fissuratum"**	Ill-fitting prosthesis *(dentures)*	Area of denture borders; maxillary arch is most common	Rolls of tissue in muco-labial fold, pink, elongated, firm ulceration, soft lesion	Surgical excision and reevaluate prosthesis with possible remaking of dentures or relining dentures
Inflammatory papillary hyperplasia: **"Palatal papillomatosis"**	Poor oral hygiene & ill-fitting prosthesis *(dentures)*	Hard palate *(vault)*	Numerous red papillary projections, soft lesion	Surgical excision and correct prosthesis

The picture below shows a **benign epithelial neoplasm** which appears as a pedunculated, whitish cauliflower-like mass on the posterior border of the tongue. The **most likely** diagnosis is:



• **Papilloma**

Note: The common wart, or **verruca vulgaris**, is a frequent tumor of skin analogous to the oral papilloma.

Histogenesis: Squamous epithelium.

Clinical Characteristics: Sessile or pedunculated, **whitish, cauliflower-like mass;** tongue, lips, gingiva, soft palate are common sites. Those within the oral cavity are soft, whereas those on the exposed areas of the lips are usually rough and scaly.

Microscopic Characteristics: Finger-like projections of stratified squamous epithelium supported by thin cores of vascular fibrous connective tissue. Epithelium may show hyperkeratosis or parakeratosis.

Treatment and Prognosis: Conservative excision; recurrence is rare.

Remember: A **fibroma** is a benign neoplasm of **connective tissue** origin.

A condition that **mimics leukoplakia** in that it seems to be a white patch is called:

- Leukanemia
- Leukemoid
- Leukoedema
- Leukochloroma

• **Leukoedema**

See picture #24 in booklet.

The appearance varies from a filmy opalescence of the mucosa in the early stages to a more definite grayish-white cast with a coarsely wrinkled surface in the later stages. The lesions usually occur bilaterally and are most noticeable along the occlusal line in the bicuspid and molar region. Diagnostically, one can stretch the tissue and the white essentially disappears. **Important:** Leukoplakia would **not disappear** when stretched.

Important point: Leukoedema appears to be **simply a variant** of normal mucosa and no treatment is necessary; merely diagnosis.

The **differential diagnosis** should include leukoplakia, white sponge nevus and hereditary benign intraepithelial dyskeratosis.

Histologically, in leukoedema, the epithelium is parakeratotic and acanthotic, with marked intracellular edema of spinous cells. **Note:** The white appearance of leukoedema is caused by water within the spinous cells causing the light to reflect back as whitish.

Actinic cheilitis is caused by:

- A virus
- Stress
- Chronic and excessive exposure to the ultraviolet radiation in sunlight
- Certain medications

• **Chronic and excessive exposure to the ultraviolet radiation in sunlight**

Actinic cheilitis *(also called solar cheilitis)* is the counterpart of actinic keratosis of the skin and can also develop into squamous cell carcinoma. In actinic cheilitis there is thickening whitish discoloration of the lip at the border of the lip and skin. There is also a loss of the usually sharp demarcation between the red of the lip and the normal skin *(vermilion border)*.

Important: This condition is considered **premalignant** and may lead to squamous cell carcinoma. It should be treated accordingly.

See picture #14 in booklet.

An **incisional biopsy** is indicated for which of the following lesions?

- A .5 cm papillary fibroma of the gingiva
- A 2 cm exostosis of the hard palate
- A 2 cm area of Fordyce's disease of the cheek
- A 3 cm hemangioma of the tongue
- A 3 cm area of leukoplakia of the soft palate

• **A 3 cm area of leukoplakia of the soft palate**

Leukoplakia is a **premalignant** lesion. This means that if left untreated, some of the lesions progress to carcinoma. It is because of this chance of malignant transformation that **all leukoplakias should be biopsied.**

It is a slowly developing change in a mucous membrane characterized by thickened, white, firmly attached patches that are slightly raised and sharply circumscribed. It is most often caused by tobacco irritation *(especially pipe).* It is more common in older men. Lesions of the **floor of the mouth** and **base of the tongue** are **most aggressive.** Most display no dysplasia but can develop into malignancy. In all cases, leukoplakia **must be completely excised.**

Explanation of **lesions** on front of card:

• **Papillary fibroma** → is a benign neoplasm of **connective tissue** origin.

• **Exostosis** of the hard palate → tori *(palatal or mandibular):* most common exophytic lesions, slow-growing benign knots of bone.

• **Fordyce's disease** *(or granules)* → ectopic sebaceous glands in the oral mucosa. They are present in over 75% of adults. They usually appear as yellow, sometimes yellow white submucosal clusters that are essentially normal.

• **Hemangioma** → is a benign tumor consisting of a mass of blood vessels.

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A **white patch** on the oral mucosa that cannot be rubbed off and cannot be assigned to any other disease **is most probably**:

- Candidiasis
- Lichen planus
- Leukoplakia
- Erythroplakia

• **Leukoplakia**

The etiology of leukoplakia is thought to be a varied one. Possible etiologic factors include tobacco, alcohol, oral sepsis and chronic irritation. It is most often due to **tobacco use** and **chronic irritation.** Some investigators believe that **pipe smoking** is most harmful. **See picture #57 in booklet.**

Leukoplakia is a clinical white patch or plaque on the oral mucosa which will not rub off and which cannot be characterized as any specific disease. Most reports indicate that leukoplakia is more common in elderly men. Although less common than leukoplakias, **erythroplakias**, have a **much greater potential** for becoming malignant. **Important:** Any white or red lesion that does not resolve itself in two weeks should be reevaluated and considered for biopsy to obtain a definitive diagnosis.

The term **carcinoma in situ** is applied to mucosal lesions which resemble leukoplakia in all respects **except** that dysplasia is very pronounced and involves almost all epithelial layers. It shows **no tendency** to invade or metastasize to other tissues.

The **clinical differential diagnosis** of a white patch should include:

- Leukoplakia
- Lupus erythematosus
- White sponge nevus
- A chemical / thermal burn
- Candidiasis
- Lichen planus
- Migratory glossitis / stomatitis

Which of the following are thought to be predisposing factors to the development of **angular cheilitis** *(Perleche)*:

- Intraoral infection by Candida albicans
- Loss of intermaxillary distance *(decreased vertical dimension)*
- Trauma to the commissure induced by prolonged dental treatment
- Vitamin deficiencies *(particularly riboflavin or thiamine)*
- All of the above

• **All of the above**

Angular cheilitis *(also called* **Perleche***)* is a term used to refer to any inflammatory lesion at the level of the labial commissure *(corners of the mouth);* the cause of which has not been determined. It is generally associated with the loss of vertical dimension. This situation is generally observed in elderly patients. The corners of the mouth become painful, irritated, red, cracked, and scaly. The fungus Candida albicans *(thrush)* may grow in the corners of the mouth, keeping them sore.

Note: Nystatin will invariably eliminate the fungal infection.

See picture #33 in booklet.

All of the following statements concerning **white sponge nevus** are true, **except:**

- It is an autosomal dominant condition
- It is also called familial white folded dysplasia
- It is characterized by soft, white, or opalescent, thickened and corrugated folds of mucous membrane
- The treatment for white sponge nevus is surgical excision and radiation

• **The treatment for white sponge nevus is surgical excision and radiation**

White sponge nevus appears to follow a hereditary pattern as an autosomal dominant trait. It has no sex preference. This mucosal abnormality is congenital in many instances, however, it may occur in childhood or adolescence. The most common location is the **buccal mucosa, bilaterally,** followed by the labial mucosa, alveolar ridge and floor of the mouth. The gingival margin and dorsal tongue are almost never affected. The mucosa appears thickened and folded with a soft or spongy texture and peculiar pearly white hue.

Note: It is often **mistaken for leukoplakia.**

Important: There is no treatment for white sponge nevus, however, since the condition is perfectly **benign**, the prognosis is excellent. There are no serious clinical complications.

See picture #8 in booklet.

Note: Hyperkeratosis is an abnormal increase in the thickness of the keratin layer *(stratum corneum)* of the epithelium. It is one of the **most common** white cheek lesions of the oral mucous membranes *(often in an area of chronic cheek biting)* and presents as being thick and scaly.

Halry tongue is a condition characterized by hypertrophy of the:

- Filiform papillae
- Fungiform papillae
- Circumvallate papillae
- Foliate papillae

• **Filiform papillae**

Hairy tongue is a **benign** condition of the tongue. The dorsum of the tongue appears furry due to the elongated papillae. The color varies from yellowish-white to brown or black. **See picture #40 in booklet.**

The four types of papillae present on the tongue:

1. **Filiform** → **most numerous,** small cones arranged in "V"- shaped rows paralleling the sulcus terminalis. They are characterized by the **absence of taste buds** and **increased keratinization.**

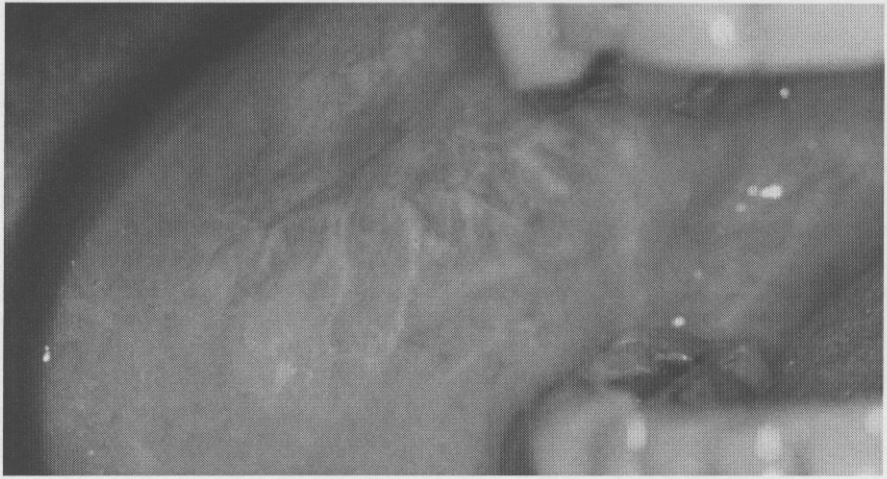
2. **Fungiform** → scattered among the filiform papillae, they are flattened, mushroom-shaped and found mainly at the **tip and lateral margins**.

3. **Circumvallate** → largest, have circular shape. Arranged in an inverted "V"- shaped row toward the back of the tongue. Associated with the ducts of **Von Ebner's glands.** They are the **least numerous** of all papillae.

4. **Foliate** → found on **lateral margins** as 3-4 vertical folds.

Note: Taste buds are **present** on the fungiform, circumvallate and foliate papillae only.

The oral lesion seen below on the buccal mucosa appears as white or grayish-white striae arranged in a **lace-like pattern.** What is the **most likely** diagnosis?



• **Lichen planus**

These lace-like white striae, the so-called **Wickham's striae,** are a classic presentation of lichen planus. They are often bilateral and symmetrical in distribution.

Lichen planus is a fairly common inflammatory disease that usually affects the skin, the mouth, or sometimes both. It affects women slightly more than men, and occurs most often in middle-aged adults. The cause of lichen planus is unknown *(possibly autoimmune).* Lichen planus of the mouth most commonly affects the buccal mucous membrane. It may also been seen on the tongue, lips, hard palate and gingiva. The lace-like striae are usually asymptomatic, but sometimes the patient may complain of a burning sensation. The intraoral lesions respond to topical steroid therapy. In addition to the usual form of lichen planus, there are two other forms, **bullous** and **erosive**. In the **bullous form,** fluid-filled vesicles project from the surface. In the **erosive form,** the lesions are intensely red or raw-appearing. When these lesions of erosive lichen planus involve the gingiva, they resemble desquamative gingivitis.

The microscopic appearance of lichen planus is **characteristic** and **pathognomonic**:
• **Hyperparakeratosis** with thickening of the granular cell layer.
• Development of a **"saw tooth"** appearance of the rete pegs.
• **Degeneration** of the basal layer of cells.
• Infiltration of **inflammatory cells** into the subepithelial layer of connective tissue.

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All of the following statements concerning **oral candidiasis** are true, **except:**

- It is also called "thrush"
- It is a bacterial infection of the mouth
- It is most often caused by Candida albicans
- The most common symptoms of oral candidiasis are discomfort and burning of the mouth and throat and an altered sense of taste
- Antibiotics prescribed for a dental infection may cause oral candidiasis

• **It is a bacterial infection of the mouth**

***This is false;** it is a **fungal** infection of the mouth.

Candidiasis *(also called moniliasis)* is an infection, usually of the oral cavity or vagina, with a Candida species, usually **C. albicans**, which causes an inflammatory, pruritic infection characterized by a thick, white discharge. It appears as diffuse, curly or velvety white mucosal plaques on the cheeks, palate and tongue that can be **wiped off,** leaving a red, raw or bleeding surface.

It is common, especially in patients who are receiving long-term antibiotic therapy or who are receiving chemotherapy and in **immunosuppressed** individuals *(i.e.,* **AIDS patients***)*. This yeast-like fungi is a normal inhabitant of the oral cavity and vaginal tract, however it is normally held in check by the indigenous bacteria of these areas. The treatment for oral candidiasis is topical through the use of lozenges *(also called trouches)* and mouth rinses, the most widely used is **nystatin**.

Notes:

1. **Acute pseudomembranous** candidiasis is the most common form of oral candidiasis and is usually found on the buccal mucosa, tongue and soft palate. **Oral cytology** smears are useful for a diagnosis *(it will reveal budding organisms with branching pseudohyphae)*.
2. **Angular cheilitis** *(Perleche)* has also been linked to C. albicans.
3. Factors that may **stimulate** Candida growth include: the extended use of antibiotics, steroids, diabetes, pregnancy, or a deficiency in iron, folate, Vitamin B_{12} or zinc.

ORAL PATHOLOGY

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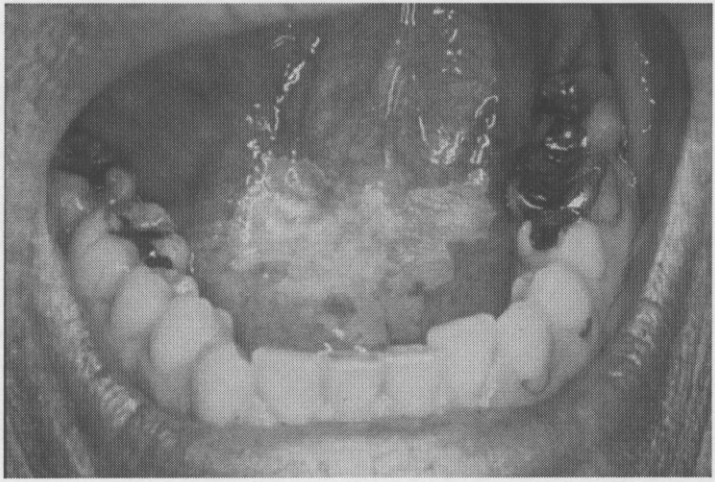
The **"white patch"** seen below has been present for nine months on a patient who is a **heavy pipe smoker**. What is the **treatment** of choice?



• **Biopsy**

Clinically, this is **leukoplakia** and should always be biopsed due to the possibility of it being a premalignant lesion. **Remember: Pipe smoking** is thought to be one of the most important predisposing etiologic factors for the development of a leukoplakia.

Important: The floor of the mouth, tongue and lower lip are the regions at greatest risk for carcinoma occurring in leukoplakia.

Stomatitis nicotina *(or "pipe-smoker's palate")* is related to pipe smoking and occurs exclusively on the palate. It affects males predominantly. The palate first appears red and inflamed. Soon it develops a diffuse, grayish-white, thickened, multinodular popular appearance with a small red **"spot"** in the center of each tiny nodule. This "spot" corresponds to the orifices of palatal salivary gland ducts. The treatment for this condition is the cessation of smoking. It is usually not considered to be a premalignant lesion.

See picture #22 in booklet.

Benign migratory glossitis is also known as:

- Fissured tongue
- Macroglossia
- Geographic tongue
- Hairy tongue

• **Geographic tongue**

***It is also called **erythema migrans.**

Geographic tongue is a harmless and very common condition in which there is desquamation of the **filiform papillae.** It is characterized by having one or more irregularly shaped patches on the tongue. The center area is redder than the rest of the tongue and the edges of the patch are whitish in color. These patches appear and remain for a short time, heal, then reappear at another site. The patches usually do not respond to treatment but disappear spontaneously. **Note:** The patient may complain of a slight burning of the tongue. **See picture #41 in booklet.**

Fissured tongue *(also called scrotal tongue)* is characterized by a deep median fissure with laterally radiating grooves. The lateral grooves vary in number but are usually symmetrical in arrangement. It is rare in children and increases in incidence with age. The fissuring occurs across the dorsum of the tongue and is usually asymptomatic, but may become painful if infected with Candida Albicans. **See picture #42 in booklet.**

Note: Fissured tongue is found in **Melkersson-Rosenthal Syndrome** *(along with cheilitis granulomatosum and facial nerve paralysis).*

Remember: Geographic tongue often occurs in **association** with fissured tongue.