Oral and Maxillofacial
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Oral and Maxillofacial Pathology

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Fourth Edition

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This book is dedicated to three of our mentors:

Charles A. Waldron
William G. Shafer
Robert J. Gorlin

*in appreciation for all that they taught us and in recognition of their contributions
to the field of oral and maxillofacial pathology.*

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About every seven years now, we receive a (dreaded?) phone call from our kindly editor suggesting that the time has arrived for us to write an updated edition of our textbook. This generally is followed by a lot of wailing and gnashing of teeth on the part of the authors—probably akin to the sound of children who have been asked to clean up their bedrooms. Just like children who may not see the necessity of changing their bed sheets, we often do not immediately appreciate the importance of revising our textbook. However, once the task is undertaken, the need becomes readily obvious.

It has become somewhat of a cliché to comment on how rapidly the world of dentistry and medicine is changing, but nothing could be closer to the truth. Admittedly, some areas seem to change very little (Fordyce granules will always be Fordyce granules). However, the expansion of our knowledge in many areas has been dramatic and, sometimes, even transformative (e.g., the relationship between high-risk strains of human papillomavirus and oropharyngeal carcinoma).

In addition to a complete update of information on previous topics, we also introduce a variety of new entities to this fourth edition, such as globodontia, lobodontia, localized juvenile spongiotic gingival hyperplasia, oral leishmaniasis, oral lesions associated with cosmetic fillers, IgG4-related disease, and mammary analogue secretory carcinoma of salivary gland origin. A total of 154 new images have been added, and we are greatly indebted to our many colleagues who have shared their excellent teaching photographs with us. We have attempted to be as thorough as possible in listing credit for these images. However, if someone's name has been inadvertently omitted, please accept our apologies.

A significant change in authorship has occurred in this fourth edition with the retirement of Dr. Jerry Bouquot, whose valuable efforts will be greatly missed. However, we are delighted and fortunate to add Dr. Angela Chi as full-fledged author/editor for this fourth edition, following up on the valuable contributions that she made to the third edition. Our great appreciation goes again to Dr. Edward Herschaft, who has updated his excellent chapter on Forensic Dentistry. We also thank Dr. Theresa Gonzales for her help with the revision of the chapter on Facial Pain and Neuromuscular Diseases. In addition, we must acknowledge the excellent guidance provided by the staff at Elsevier for their hard work and support in making this book a success. Special praise goes to Courtney Sprehe, Marquita Parker, and Kathy Falk for all of their efforts in the editorial process.

Finally, our greatest appreciation goes to our families, who again have provided us with their unconditional love and support during the long hours spent working on this latest edition. We never could have accomplished it without you.

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1

Developmental Defects of the Oral and Maxillofacial Region

◆ OROFACIAL CLEFTS

The formation of the face and oral cavity is complex in nature and involves the development of multiple tissue processes that must merge and fuse in a highly orchestrated fashion. Disturbances in the growth of these tissue processes or their fusion may result in the formation of **orofacial clefts.**

Development of the central face begins around the end of the fourth week of human development with the appearance of the nasal (olfactory) placodes on either side of the inferior aspect of the frontonasal process. Proliferation of ectomesenchyme on both sides of each placode results in the formation of the medial and lateral nasal processes. Between each pair of processes is a depression, or nasal pit, that represents the primitive nostril.

During the sixth and seventh weeks of development, the upper lip forms when the medial nasal processes merge with each other and with the maxillary processes of the first branchial arches. Thus the midportion of the upper lip is derived from the medial nasal processes, and the lateral portions are derived from the maxillary processes. The lateral nasal processes are not involved in the formation of the upper lip, but they give rise to the alae of the nose.

The **primary palate** also is formed by the merger of the medial nasal processes to form the intermaxillary segment. This segment gives rise to the premaxilla, a triangular-shaped piece of bone that will include the four incisor teeth. The **secondary palate,** which makes up 90% of the hard and soft palates, is formed from the maxillary processes of the first branchial arches.

During the sixth week, bilateral projections emerge from the medial aspects of the maxillary processes to form the palatal shelves. Initially, these shelves are oriented in a vertical position on each side of the developing tongue. As the mandible grows, the tongue drops down, allowing the palatal shelves to rotate to a horizontal position and grow toward one another. By the eighth week, sufficient growth has occurred to allow the anterior aspects of these shelves to begin fusion with one another. The palatal shelves also fuse with the primary palate and the nasal septum. The fusion of the palatal shelves begins in the anterior palate and progresses posteriorly; it is completed by the twelfth week.

Defective fusion of the medial nasal process with the maxillary process leads to **cleft lip (CL).** Likewise, failure of the palatal shelves to fuse results in **cleft palate (CP).** Frequently, CL and CP occur together. Approximately 45% of cases are CL + CP with 30% being CP only (CPO) and 25% being isolated CL. Both isolated CL and CL associated with CP are thought to be etiologically related conditions and can be considered as a group: CL, with or without CP (i.e., CL ± CP). Isolated CPO appears to represent a separate entity from CL ± CP.

The cause of CL ± CP and CPO is still being debated. First of all, distinguishing isolated clefts from cases associated with specific syndromes is important. Although many facial clefts are isolated anomalies, more than 400 developmental syndromes have been identified that may be associated with CL ± CP or CPO. Studies have suggested that up to 30% of patients with CL ± CP and 50% of those with CPO have associated anomalies. Some of these cases are single-gene syndromes that may follow autosomal dominant, autosomal recessive, or X-linked inheritance patterns. Other syndromes are the result of chromosome anomalies or are idiopathic.

The cause of nonsyndromic clefts does not follow any simple Mendelian pattern of inheritance but appears to be heterogeneous. Thus the propensity for cleft development may be related to a number of major genes, minor genes, and environmental factors that can combine to surpass a developmental threshold. Numerous candidate clefting genes and loci have been identified on different chromosome regions. Maternal alcohol consumption has been associated with an increased risk for both syndromic and nonsyndromic clefts. Maternal cigarette smoking at least doubles the frequency of cleft development compared with nonsmoking mothers. An increased frequency also has been related to anticonvulsant therapy, especially phenytoin, which causes a nearly tenfold greater risk of cleft formation. Although evidence has been mixed, a number of studies have suggested that folic acid supplementation may play a role in prevention of orofacial clefts.

CL ± CP and CPO represent the vast majority of orofacial clefts. However, other rare clefts also may occur.

The **lateral facial cleft** is caused by lack of fusion of the maxillary and mandibular processes and represents 0.3% of all facial clefts. This cleft may be unilateral or bilateral, extending from the commissure toward the ear, resulting in macrostomia. The lateral facial cleft may occur as an isolated defect, but more often it is associated with other disorders, such as the following:

- Mandibulofacial dysostosis (see page 41)
- Oculo-auriculo-vertebral spectrum (hemifacial microsomia)
- Nager acrofacial dysostosis
- Amniotic rupture sequence

The **oblique facial cleft** extends from the upper lip to the eye. It is nearly always associated with CP, and severe forms often are incompatible with life. The oblique facial cleft may involve the nostril, as in CL, or it may bypass the nose laterally as it extends to the eye. This cleft is rare, representing only 1 in 1300 facial clefts. Some of these clefts may represent failure of fusion of the lateral nasal process with the maxillary process; amniotic bands may cause others.

Median cleft of the upper lip is an extremely rare anomaly that results from failure of fusion of the medial nasal processes. It may be associated with a number of syndromes, including the oral-facial-digital syndromes and Ellis-van Creveld syndrome. Most apparent median clefts of the upper lip actually represent agenesis of the primary palate associated with holoprosencephaly.

Clinical and Radiographic Features

Clefting is one of the most common major congenital defects in humans. Considerable racial variation in prevalence is seen. In whites, CL ± CP occurs in 1 of every 700 to 1000 births. The frequency of CL ± CP in Asian populations is about 1.5 times higher than in whites. In contrast, the prevalence of CL ± CP in blacks is much lower, occurring in 0.4 of 1000 births. Native Americans appear to have the highest frequency, around 3.6 of 1000 births. CPO is less common than CL ± CP, with a frequency of 0.4 of 1000 births in whites and blacks.

CL ± CP is more common in males than in females. The more severe the defect, the greater the male predilection; the male-to-female ratio for isolated CL is 1.5 : 1; the ratio for CL + CP is 2 : 1. In contrast, CPO is more common in females. Likewise, the more severe the cleft, the greater the female predilection. Clefts of both the hard and soft palates are twice as common in females, but the ratio is nearly equal for clefts of the soft palate only.

Approximately 80% of cases of CL will be unilateral with 20% bilateral (Fig. 1-1). Approximately 70% of unilateral CLs occur on the left side. In addition, about 70% of unilateral CLs will be associated with CP, whereas the frequency of concomitant CP increases to 85% for patients with bilateral CL. A complete CL extends upward into the

● **Fig. 1-1 Cleft Lip (CL).** Infant with bilateral cleft of the upper lip. (Courtesy of Dr. William Bruce.)
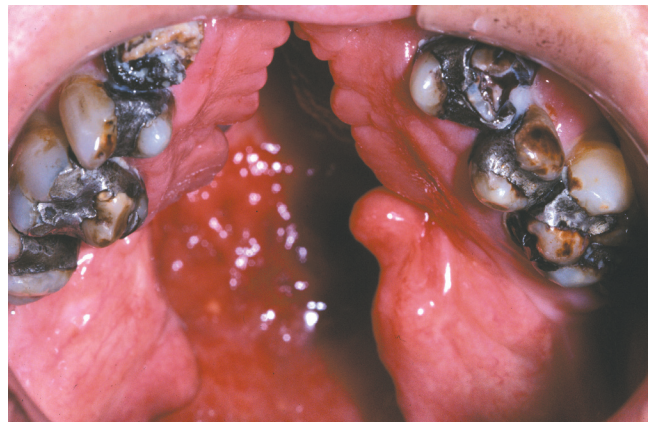
● **Fig. 1-2 Cleft Palate (CP).** Palatal defect resulting in communication with the nasal cavity.

nostril, but an incomplete CL does not involve the nose. Complete clefts involving the alveolus usually occur between the lateral incisor and cuspid. It is not unusual for teeth, especially the lateral incisor, to be missing in the cleft area. Conversely, supernumerary teeth may be discovered. The bony defect can be observed on radiographs.

A CP shows considerable range in severity (Fig. 1-2). The defect may involve the hard and soft palates or the soft palate alone. The minimal manifestation of CP is a **cleft** or **bifid uvula** (Fig. 1-3). The prevalence of cleft uvula is much higher than that of CP, with a frequency of 1 in every 80 white individuals. The frequency in Asian and Native American populations is as high as 1 in 10. Cleft uvula is less common in blacks, occurring in 1 out of every 250 persons.

In some instances a **submucous palatal cleft** develops. The surface mucosa is intact, but a defect exists in the underlying musculature of the soft palate (Fig. 1-4). Frequently a notch in the bone is present along the posterior margin of the hard palate. This incomplete cleft occasionally appears as a bluish midline discoloration but is best identified by palpation with a blunt instrument. An associated cleft uvula also usually is seen.

• **Fig. 1-3 Bifid Uvula.**

• **Fig. 1-4 Submucous Palatal Cleft.** A cleft of the midline palatal bone exists, but the overlying mucosa is intact. A bifid uvula also is present.
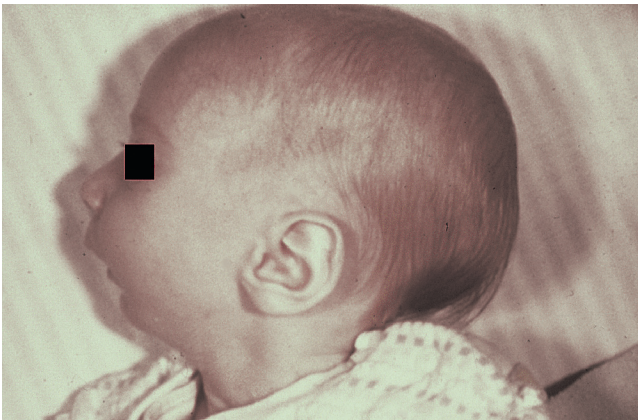
• **Fig. 1-5 Pierre Robin Sequence.** Micrognathic mandible in an infant with cleft palate (CP). (Courtesy of Dr. Robert Gorlin.)

The **Pierre Robin sequence** (Pierre Robin anomalad) (Fig. 1-5) is a well-recognized presentation characterized by CP, mandibular micrognathia, and glossoptosis (airway obstruction caused by lower, posterior displacement of the tongue). The Pierre Robin sequence may occur as an isolated phenomenon, or it may be associated with a wide

variety of syndromes or other anomalies. Stickler syndrome and velocardiofacial syndrome are the two most frequently associated genetic disorders. Researchers have theorized that constraint of mandibular growth *in utero* results in failure of the tongue to descend, thus preventing fusion of the palatal shelves. The retruded mandible results in the following:
• Posterior displacement of the tongue
• Lack of support of the tongue musculature
• Airway obstruction

Respiratory difficulty, especially when the child is in a supine position, usually is noted from birth and can cause asphyxiation. The palatal cleft often is U-shaped and wider than isolated CP.

The patient with a cleft is burdened with a variety of problems, some obvious and some less so. The most obvious problem is the clinical appearance, which may lead to psychosocial difficulties. Feeding and speech difficulties are inherent, especially with CP. Malocclusion is caused by collapse of the maxillary arch, possibly along with missing teeth, supernumerary teeth, or both.

Treatment and Prognosis

The management of the patient with an orofacial cleft is challenging. Ideally, treatment should involve a multidisciplinary approach, including (but not limited to) a pediatrician, oral and maxillofacial surgeon, otolaryngologist, plastic surgeon, pediatric dentist, orthodontist, prosthodontist, speech pathologist, and geneticist.

Surgical repair often involves multiple primary and secondary procedures throughout childhood. The specific types of surgical procedures and their timing will vary, depending on the severity of the defect and the philosophy of the treatment team. A detailed discussion of these procedures is beyond the scope of this text. However, primary lip closure usually is accomplished during the first few months of life, followed later by repair of the palate. Prosthetic and orthopedic appliances often are used to mold or expand the maxillary segments before closure of the palatal defect. Later in childhood, autogenous bone grafts can be placed in the area of the alveolar bone defect. Secondary soft tissue and orthognathic procedures may be used to improve function and cosmetic appearance. Distraction osteogenesis of the maxilla can prove useful in patients in whom palatal scarring limits the amount of advancement possible at the time of osteotomy.

Breathing difficulties in infants with Pierre Robin sequence are managed best with conservative measures, such as side and prone positioning. However, in children with significant airway obstruction, placement of a nasopharyngeal airway may be warranted. In more severe cases, mandibular distraction osteogenesis may be a preferable treatment alternative to tracheostomy.

Genetic counseling is important for the patient and family. In nonsyndromic cases of orofacial clefting, the risk for cleft development in a sibling or offspring of an affected

person is 3% to 5% if no other first-degree relatives also are affected. The risk increases to 10% to 20% if other first-degree relatives are affected. The risk may be even higher for those with clefts that are associated with syndromes, depending on the possible inheritance pattern.

◆ COMMISSURAL LIP PITS

Commissural lip pits are small mucosal invaginations that occur at the corners of the mouth on the vermilion border. Their location suggests that they may represent a failure of normal fusion of the embryonal maxillary and mandibular processes.

Commissural lip pits appear to be common in adults, where they have been reported in 12% to 20% of the population. Their prevalence in children is considerably lower, ranging from 0.2% to 0.7% of those examined.

Although commissural lip pits are generally considered to be congenital lesions, these figures suggest that these invaginations often develop later in life. Commissural pits are seen more often in males than in females. A family history suggestive of autosomal dominant transmission has been noted in some cases.

Clinical Features

Commissural lip pits usually are discovered on routine examination, and the patient often is unaware of their presence. These pits may be unilateral or bilateral. They manifest as blind fistulas that may extend to a depth of 1 to 4 mm (Fig. 1-6). In some cases, a small amount of fluid may be expressed when the pit is squeezed, presumably representing saliva from minor salivary glands that drain into the depth of the invagination.

Unlike **paramedian lip pits** (described in the following section), commissural lip pits are not associated with facial or palatal clefts. However, there does appear to be a significantly higher prevalence of preauricular pits (aural sinuses) in these patients.

Histopathologic Features

Although biopsy rarely is performed for patients with commissural lip pits, microscopic examination reveals a narrow invagination lined by stratified squamous epithelium. Ducts from minor salivary glands may drain into this invagination.

Treatment and Prognosis

Because commissural lip pits are virtually always asymptomatic and innocuous, no treatment is usually necessary. In extremely rare instances, salivary secretions may be excessive or secondary infection may occur, necessitating surgical excision of the pit.

◆ PARAMEDIAN LIP PITS (CONGENITAL FISTULAS OF THE LOWER LIP; CONGENITAL LIP PITS)

Paramedian lip pits are rare congenital invaginations of the lower lip. They are believed to arise from persistent lateral sulci on the embryonic mandibular arch. These sulci normally disappear by 6 weeks of embryonic age.

Clinical Features

Paramedian lip pits typically appear as bilateral and symmetric fistulas on either side of the midline of the vermilion of the lower lip (Fig. 1-7). Their appearance can range from subtle depressions to prominent humps. These blind sinuses can extend down to a depth of 1.5 cm and may express salivary secretions. Occasionally, only a single pit is present that may be centrally located or lateral to the midline.

The greatest significance of paramedian lip pits is that they usually are inherited as an autosomal dominant trait in combination with cleft lip (CL) and/or cleft palate (CP)

• **Fig. 1-6 Commissural Lip Pit.** Depression at the labial commissure.

• **Fig. 1-7 Paramedian Lip Pits.** Bilateral pits on the lower lip in a patient with van der Woude syndrome. (Courtesy of Dr. Nadarajah Vigneswaran.)

• **Fig. 1-8 Van der Woude Syndrome.** Same patient as depicted in Figure 1-7 with a cleft of the soft palate. (Courtesy of Dr. Nadarajah Vigneswaran.)

• **Fig. 1-9 Double Lip.** When the patient smiles, a redundant fold of tissue partially covers the right anterior maxillary teeth. (Courtesy of Dr. Logan Barnes.)

(**van der Woude syndrome**) (Fig. 1-8). Van der Woude syndrome is the most common form of syndromic clefting and accounts for 2% of all cases of CL and CP. Associated hypodontia also may be observed. Genetic studies have shown that this condition is caused by mutations in the gene that encodes interferon regulatory factor 6 (IRF6), which has been mapped to chromosome locus 1q32-q41. Some people who carry the trait may not demonstrate clefts or may have a submucous CP; however, they may pass the full syndrome to their offspring.

Paramedian lip pits also may be a feature of the **popliteal pterygium syndrome** and **Kabuki syndrome.** Popliteal webbing (**pterygia**), CL and/or CP, genital abnormalities, and congenital bands connecting the upper and lower jaws (**syngnathia**) characterize popliteal pterygium syndrome, which is closely related to van der Woude syndrome. Kabuki syndrome received its name because affected patients exhibit eversion of the lower lateral eyelids, which is reminiscent of the makeup used by actors in Kabuki, the traditional form of Japanese theater. Other common findings include intellectual disability, large ears, CL and/or CP, hypodontia, joint laxity, and various skeletal abnormalities.

Histopathologic Features

Microscopic examination of a paramedian lip pit shows a tract that is lined by stratified squamous epithelium. Minor salivary glands may communicate with the sinus. A chronic inflammatory cell infiltrate often is noted in the surrounding connective tissue.

Treatment and Prognosis

If necessary, the labial pits may be excised for cosmetic reasons. The most significant problems are related to associated congenital anomalies, such as CL and/or CP, and the potential for transmission of the trait to subsequent generations.

◆ DOUBLE LIP

Double lip is a rare oral anomaly characterized by a redundant fold of tissue on the mucosal side of the lip. Most often it is congenital in nature, but it may be acquired later in life. Congenital cases are believed to arise during the second to third month of gestation as a result of the persistence of the sulcus between the pars glabrosa and pars villosa of the lip. Acquired double lip may be a component of **Ascher syndrome,** or it may result from trauma or oral habits, such as sucking on the lip.

Clinical Features

In a patient with double lip, the upper lip is affected much more often than the lower lip; occasionally, both lips are involved. With the lips at rest, the condition is usually unnoticeable, but when the patient smiles or when the lips are tensed, the excess fold of tissue is visible (Fig. 1-9).

Ascher syndrome is characterized by a triad of features:
• Double lip
• Blepharochalasis
• Nontoxic thyroid enlargement

In a person with blepharochalasis, recurring edema of the upper eyelid leads to sagging of the lid at the outer canthus of the eye (Fig. 1-10). This drooping may be severe enough to interfere with vision. Both the double lip and blepharochalasis usually occur abruptly and simultaneously, but in some cases they develop more gradually.

The nontoxic thyroid enlargement occurs in as many as 50% of patients with Ascher syndrome and may be mild in degree. The cause of Ascher syndrome is not certain; autosomal dominant inheritance has been suggested in some cases.

Histopathologic Features

On microscopic examination, double lip shows essentially normal structures. Often there is an abundance of minor

• **Fig. 1-10 Ascher Syndrome.** Edema of the upper eyelids (blepharochalasis).

• **Fig. 1-11 Fordyce Granules.** Yellow papules on the vermilion of the upper lip.

salivary glands. The blepharochalasis of Ascher syndrome usually shows hyperplasia of the lacrimal glands or prolapse of orbital fat.

Treatment and Prognosis

In mild cases of double lip, no treatment may be required. In more severe cases, simple surgical excision of the excess tissue can be performed for aesthetic purposes.
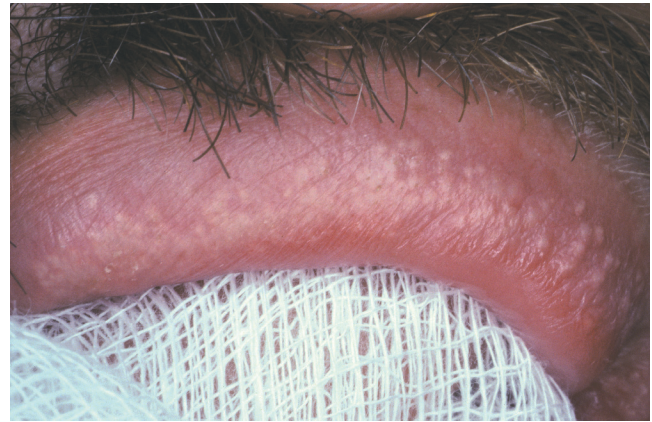
◆ FORDYCE GRANULES

Fordyce granules are sebaceous glands that occur on the oral mucosa. Similar lesions also have been reported on the genital mucosa. Because sebaceous glands typically are considered to be dermal adnexal structures, those found in the oral cavity often have been considered to be "ectopic." However, because Fordyce granules have been reported in more than 80% of the population, their presence must be considered a normal anatomic variation.

Clinical Features

Fordyce granules appear as multiple yellow or yellow-white papules that are most common on the buccal mucosa and the lateral portion of the vermilion of the upper lip (Figs. 1-11 and 1-12). Occasionally, these glands also may appear in the retromolar area and anterior tonsillar pillar. They are more common in adults than in children, probably as a result of hormonal factors; puberty appears to stimulate their development. The lesions are typically asymptomatic, although patients may be able to feel a slight roughness to the mucosa. Considerable clinical variation may exist; some patients may have only a few lesions, whereas others may have literally hundreds of these "granules."

Histopathologic Features

Except for the absence of associated hair follicles, Fordyce granules closely resemble normal sebaceous glands found in

• **Fig. 1-12 Fordyce Granules.** Lesions on the buccal mucosa.

• **Fig. 1-13 Fordyce Granules.** Multiple sebaceous glands below the surface epithelium.

the skin. Acinar lobules can be seen immediately beneath the epithelial surface, often communicating with the surface through a central duct (Fig. 1-13). The sebaceous cells in these lobules are polygonal in shape, containing centrally located nuclei and abundant foamy cytoplasm.

Treatment and Prognosis

Because Fordyce granules represent a normal anatomic variation and are asymptomatic, no treatment is indicated. Usually, the clinical appearance is characteristic and biopsy is not necessary for diagnosis.

On occasion, Fordyce granules may become hyperplastic or may form keratin-filled pseudocysts. Tumors arising from these glands are exceedingly rare.

◆ LEUKOEDEMA

Leukoedema is a common oral mucosal condition of unknown cause. It occurs more commonly in blacks than in whites, supporting the likelihood of an ethnic predisposition to its development. Leukoedema has been reported in 70% to 90% of black adults and in 50% of black children. The prevalence in whites is considerably less, although published reports have ranged from less than 10% to more than 90%. This variation may reflect differing population groups, examination conditions, and stringency of criteria used to make the diagnosis. At any rate, leukoedema shows a much milder presentation in whites and often is hardly noticeable. The difference in racial predilection may be explained by the presence of background mucosal pigmentation in blacks that makes the edematous changes more noticeable.

Because leukoedema is so common, it can reasonably be argued that it represents a *variation of normal* rather than a disease. The finding of similar edematous mucosa in the vagina and larynx further supports this argument. Although leukoedema appears to be developmental in nature, some studies have indicated that it is more common and more severe in smokers and becomes less pronounced with cessation of smoking.

Clinical Features

Leukoedema is characterized by a diffuse, gray-white, milky, opalescent appearance of the mucosa (Fig. 1-14). The surface frequently appears folded, resulting in wrinkles or whitish streaks. The lesions do not rub off. Leukoedema typically occurs bilaterally on the buccal mucosa and may extend forward onto the labial mucosa. On rare occasions, it also can involve the floor of the mouth and palatopharyngeal tissues. Leukoedema can be easily diagnosed clinically because the white appearance greatly diminishes or disappears when the cheek is everted and stretched (Fig. 1-15).

Histopathologic Features

Biopsy specimens of leukoedema demonstrate an increase in thickness of the epithelium with striking intracellular edema of the spinous layer (Fig. 1-16). These vacuolated cells appear large and have pyknotic nuclei. The epithelial surface is frequently parakeratinized, and the rete ridges are broad and elongated.

Treatment and Prognosis

Leukoedema is a benign condition, and no treatment is required. The characteristic milky-white, opalescent lesions of the buccal mucosa that disappear when stretched help distinguish it from other common white lesions, such as leukoplakia, candidiasis, and lichen planus. The affected mucosa always should be stretched during clinical

A

B
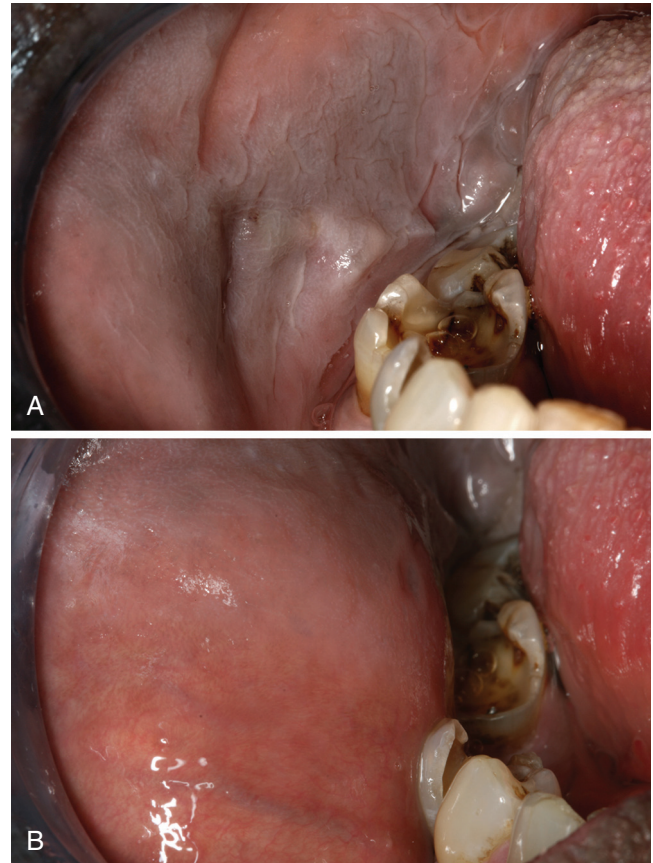
• **Fig. 1-15 Leukoedema. A,** Diffuse white appearance of the buccal mucosa. **B,** Whiteness disappears when the cheek is stretched.

• **Fig. 1-14 Leukoedema.** White, wrinkled appearance of the buccal mucosa.
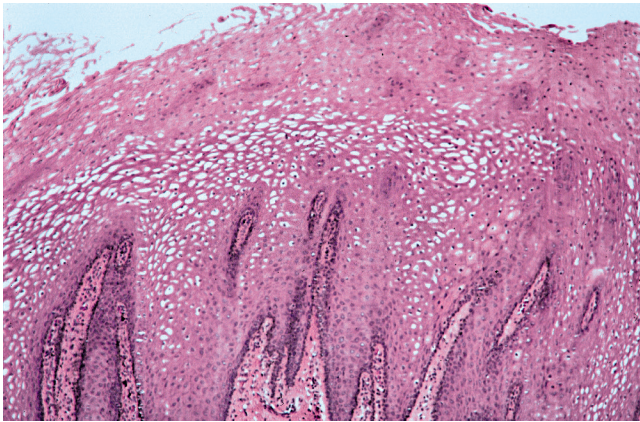
• **Fig. 1-16 Leukoedema.** Parakeratosis and intracellular edema of the spinous layer.

examination to rule out any underlying lesions that may be hidden by the edematous change.

◆ MICROGLOSSIA (HYPOGLOSSIA)

Clinical Features

Microglossia is an uncommon developmental condition of unknown cause that is characterized by an abnormally small tongue. In rare instances, virtually the entire tongue may be missing **(aglossia).** Isolated microglossia is known to occur, and mild degrees of microglossia may be difficult to detect and may go unnoticed. However, most reported cases have been associated with one of a group of overlapping conditions known as **oromandibular-limb hypogenesis syndromes.** These syndromes feature associated limb anomalies, such as **hypodactylia** (i.e., absence of digits) and **hypomelia** (i.e., hypoplasia of part or all of a limb). Other patients have had coexisting anomalies, such as cleft palate, intraoral bands, and *situs inversus.* Microglossia frequently is associated with hypoplasia of the mandible, and the lower incisors may be missing (Fig. 1-17).

Treatment and Prognosis

Treatment of the patient with microglossia depends on the nature and severity of the condition. Surgery and orthodontics may improve oral function. Surprisingly, speech development often is quite good but depends on tongue size.

◆ MACROGLOSSIA

Macroglossia is an uncommon condition characterized by enlargement of the tongue. The enlargement may be caused by a wide variety of conditions, including congenital malformations and acquired diseases. The most frequent causes are vascular malformations and muscular hypertrophy. Box 1-1 lists the most common and important causes of macroglossia. Many of these diseases are discussed in greater detail in subsequent chapters of this book.

• **Fig. 1-17 Microglossia. A,** Abnormally small tongue associated with constricted mandibular arch. **B,** Same patient with associated constriction of the maxillary arch.

• BOX 1-1   Causes of Macroglossia

Congenital and Hereditary
- Vascular malformations
- Lymphangioma
- Hemangioma
- Hemihyperplasia
- Cretinism
- Beckwith-Wiedemann syndrome
- Down syndrome
- Duchenne muscular dystrophy
- Mucopolysaccharidoses
- Neurofibromatosis type I
- Multiple endocrine neoplasia, type 2B

Acquired
- Edentulous patients
- Amyloidosis
- Myxedema
- Acromegaly
- Angioedema
- Myasthenia gravis
- Amyotrophic lateral sclerosis
- Carcinoma and other tumors

Clinical Features

Macroglossia most commonly occurs in children and can range from mild to severe (Fig. 1-18). In infants,

• **Fig. 1-18 Macroglossia.** Large tongue in a patient with Down syndrome. (Courtesy of Dr. Sanford Fenton.)

• **Fig. 1-19 Macroglossia.** The tongue enlargement has resulted in a crenated border that corresponds to the embrasures between the teeth.

macroglossia may be manifested first by noisy breathing, drooling, and difficulty in eating. The tongue enlargement may result in a lisping speech. The pressure of the tongue against the mandible and teeth can produce a crenated lateral border to the tongue (Fig. 1-19), open bite, and mandibular prognathism. If the tongue constantly protrudes from the mouth, it may ulcerate and become secondarily infected or may even undergo necrosis. Severe macroglossia can produce airway obstruction.

Macroglossia is a characteristic feature of **Beckwith-Wiedemann syndrome,** a rare hereditary condition that includes many other possible defects, such as the following:
- Omphalocele (i.e., protrusion of part of the intestine through a defect in the abdominal wall at the umbilicus)
- Visceromegaly
- Gigantism
- Neonatal hypoglycemia

Individuals with Beckwith-Wiedemann syndrome have an increased risk for several childhood visceral tumors, including Wilms tumor, adrenal carcinoma, hepatoblastoma, rhabdomyosarcoma, and neuroblastoma. Facial features may include nevus flammeus of the forehead and eyelids, linear indentations of the earlobes, and maxillary

hypoplasia (resulting in relative mandibular prognathism). Most examples of Beckwith-Wiedemann syndrome are sporadic, but 10% to 15% of cases show autosomal dominant inheritance with preferential maternal transmission. The genetic basis is complex, involving a variety of alterations within two domains of imprinted growth-regulatory genes on chromosome 11p15.

In patients with **hypothyroidism** (see page 777), Beckwith-Wiedemann syndrome, or neuromuscular disorders, the tongue usually shows a diffuse, smooth, generalized enlargement. In those with other forms of macroglossia, the tongue usually has a multinodular appearance. Examples of this nodular type include **amyloidosis** (see page 766) and neoplastic conditions, such as **neurofibromatosis** (see page 495) and **multiple endocrine neoplasia, type 2B** (see page 497).

In patients with **lymphangiomas** (see page 510), the tongue surface is characteristically pebbly and exhibits multiple vesicle-like blebs that represent superficial dilated lymphatic channels. The enlarged tongue in those with **Down syndrome** typically demonstrates a papillary, fissured surface.

In patients with **hemifacial hyperplasia** (see page 35), the enlargement will be unilateral. Some patients with neurofibromatosis also can have unilateral lingual enlargement.

In edentulous patients, the tongue often appears elevated and tends to spread out laterally because of loss of the surrounding teeth; as a result, wearing a denture may become difficult.

Histopathologic Features

The microscopic appearance of macroglossia depends on the specific cause. In some cases, such as the tongue enlargement seen with Down syndrome or in edentulous patients, no histologic abnormality can be detected. When macroglossia is due to tumor, a neoplastic proliferation of a particular tissue can be found (e.g., lymphatic vessels, blood vessels, neural tissue). Muscular enlargement occurs in those with hemihyperplasia and Beckwith-Wiedemann syndrome. In neuromuscular disorders, such as myasthenia gravis or amyotrophic lateral sclerosis, tongue enlargement may result from muscular atrophy with prominent fatty replacement. In the patient with amyloidosis, an abnormal protein material is deposited in the tongue.

Treatment and Prognosis

The treatment and prognosis of macroglossia depend on the cause and severity of the condition. In mild cases, surgical treatment may not be necessary, although speech therapy may be helpful if speech is affected. In symptomatic patients, reduction glossectomy may be needed.

◆ ANKYLOGLOSSIA (TONGUE-TIE)

Ankyloglossia is a developmental anomaly of the tongue characterized by a short, thick lingual frenum resulting in

• **Fig. 1-20 Ankyloglossia.** Abnormal attachment of the lingual frenum, limiting tongue mobility.

limitation of tongue movement. It has been reported to occur in 1.7% to 10.7% of neonates, being more common in boys than in girls. In adults, mild forms are not unusual, but severe ankyloglossia is a relatively uncommon condition that has been estimated to occur in about two to three of every 10,000 people. Most examples of ankyloglossia appear to be sporadic, although evidence suggests that there could be a genetic influence in some cases.

Clinical Features

Ankyloglossia can range in severity from mild cases with little clinical significance to rare examples of complete ankyloglossia in which the tongue is actually fused to the floor of the mouth (Fig. 1-20). Sometimes the frenum extends forward and attaches to the tip of the tongue, and slight clefting of the tip may be seen.

Some investigators have speculated that ankyloglossia may lead to the development of an anterior open bite because the inability to raise the tongue to the roof of the mouth prevents development of the normal adult swallowing pattern. However, others have questioned this theory. It also is possible that a high mucogingival attachment of the lingual frenum may contribute to gingival recession, although a clear relationship has not been established.

It has been suggested that tongue-tie may result in speech defects. Usually, however, the shortened frenum results in only minor difficulties because most people can compensate for the limitation in tongue movement. Yet there are rare examples of patients who have experienced an immediate noticeable improvement in speech after surgical correction of ankyloglossia. With the increase in popularity of breast-feeding over the past several decades, clinicians have related tongue-tie with feeding problems, such as nipple pain or difficulty in the baby attaching to the breast.

Treatment and Prognosis

Because most cases of ankyloglossia result in few or no clinical problems, treatment is often unnecessary. For infants

with specific breast-feeding problems, a frenotomy ("clipping" or simple release of the frenulum) can be performed, which has been shown to improve nipple-pain and breast-feeding scores. In children or adults with associated functional or periodontal difficulties, a frenuloplasty (release with plastic repair) may allow greater freedom of tongue movement. In young children, it often is recommended that surgery be postponed until age 4 or 5. Because the tongue is always short at birth, assessing the degree of tongue limitation caused by ankyloglossia is difficult in the infant's early life. As the infant grows, the tongue becomes longer and thinner at the tip, often decreasing the severity of the tongue-tie. The condition probably is self-correcting in many cases because it is less common in adults.
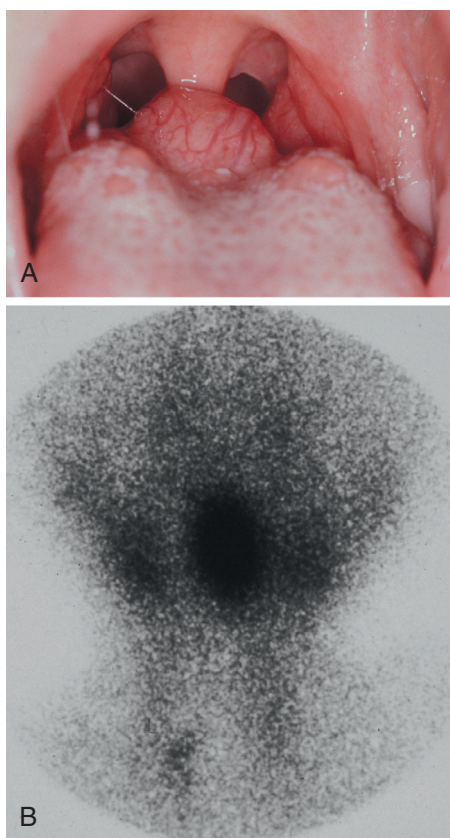
◆ LINGUAL THYROID

During the third to fourth week of fetal life, the thyroid gland begins as an epithelial proliferation in the floor of the pharyngeal gut. By the seventh embryonic week, this thyroid bud normally descends into the neck to its final resting position anterior to the trachea and larynx. The site where this descending bud invaginates later becomes the foramen cecum, located at the junction of the anterior two-thirds and posterior third of the tongue in the midline. If the primitive gland does not descend normally, ectopic thyroid tissue may be found between the foramen cecum and the epiglottis. Of all ectopic thyroids, 90% are found in this region.

Clinical Features

Based on autopsy studies, small asymptomatic remnants of thyroid tissue can be discovered on the posterior dorsal tongue in about 10% of both men and women. However, clinically evident or symptomatic **lingual thyroids** are much less common and are four to seven times more frequent in females, presumably because of hormonal influences. Symptoms most often develop during puberty, adolescence, pregnancy, or menopause. In 70% of cases, this ectopic gland is the patient's only thyroid tissue.

Lingual thyroids may range from small, asymptomatic, nodular lesions to large masses that can block the airway (Fig. 1-21). The most common clinical symptoms are dysphagia, dysphonia, and dyspnea. The mass often is vascular, but the physical appearance is variable, and there are no reliable features to distinguish it from other masses that might develop in this area. Hypothyroidism has been reported in up to 33% of patients. Many authors say that lingual thyroid enlargement is a secondary phenomenon, compensating for thyroid hypofunction. Interestingly, as many as 75% of patients with infantile hypothyroidism have some ectopic thyroid tissue.

Diagnosis is best established by thyroid scan using iodine isotopes or technetium-99m (^{99m}Tc). Computed tomography (CT), magnetic resonance imaging (MRI), and ultrasonography can be helpful in delineating the size and extent

Fig. 1-21 Lingual Thyroid. A, Nodular mass of the posterior dorsal midline of the tongue in a 4-year-old girl. **B,** Thyroid scan of the same patient. The scan shows localization (central dark zone) of iodine isotope in the tongue mass and minimal uptake in the neck.

Fig. 1-22 Fissured Tongue. Extensive fissuring involving the entire dorsal tongue surface. (Courtesy of Chris Neville.)

of the lesion. Biopsy is often avoided because of the risk of hemorrhage and because the mass may represent the patient's only functioning thyroid tissue. In some cases, incisional biopsy may be needed to confirm the diagnosis or to rule out malignant changes.

Treatment and Prognosis

No treatment except periodic follow-up is required for patients with asymptomatic lingual thyroids. In symptomatic patients, suppressive therapy with supplemental thyroid hormone often can reduce the size of the lesion. Some authors advise that this treatment also should be tried in asymptomatic patients to prevent possible subsequent enlargement. If hormone therapy does not eliminate symptoms, surgical removal or ablation with radioactive iodine-131 can be performed. If the mass is excised, autotransplantation to another body site can be attempted to maintain functional thyroid tissue and to prevent hypothyroidism.

Rare examples of carcinomas arising in lingual thyroids have been reported; malignancy develops in about 1% of identified cases. Although lingual thyroids are decidedly more common in females, this predilection for females is less pronounced for lingual thyroid carcinomas. Because a

disproportionate number of these malignancies have been documented in males, some authors have advocated prophylactic excision of lingual thyroids in men older than 30 years of age.

◆ FISSURED TONGUE (SCROTAL TONGUE)

Fissured tongue is a relatively common condition that is characterized by the presence of numerous grooves, or fissures, on the dorsal tongue surface. The cause is uncertain, but heredity appears to play a significant role. Evidence indicates that the condition may be either a polygenic trait or an autosomal dominant trait with incomplete penetrance. Aging or local environmental factors also may contribute to its development.

Clinical Features

Patients with fissured tongue exhibit multiple grooves, or furrows, on the surface of the tongue, ranging from 2 to 6 mm in depth (Fig. 1-22). Considerable variation can be seen. In the most severe cases, numerous fissures cover the entire dorsal surface and divide the tongue papillae into multiple separate "islands." Some patients have fissures that are located mostly on the dorsolateral areas of the tongue. Other patients exhibit a large central fissure with smaller fissures branching outward at right angles. The condition usually is asymptomatic, although some patients may complain of mild burning or soreness.

Most studies have shown that the prevalence of fissured tongue ranges from 2% to 5% of the overall population. The condition may be seen in children or adults, but the prevalence and severity appear to increase with age, with some studies noting the presence of fissured tongue in as many as 30% of older adults. In some investigations, a male predilection has been noted.

A strong association has been found between fissured tongue and **geographic tongue** (see page 726) with many patients having both conditions. A hereditary basis also has been suggested for geographic tongue, and the same gene

or genes may possibly be linked to both conditions. In fact, it even has been suggested that geographic tongue may *cause* fissured tongue. Fissured tongue also may be a component of **Melkersson-Rosenthal syndrome** (see page 313).

Histopathologic Features

Microscopic examination of fissured tongue reveals hyperplasia of the rete ridges and loss of the keratin "hairs" on the surface of the filiform papillae. The papillae vary in size and often are separated by deep grooves. Polymorphonuclear leukocytes can be seen migrating into the epithelium, often forming microabscesses in the upper epithelial layers. A mixed inflammatory cell infiltrate is present in the lamina propria.

Treatment and Prognosis

Fissured tongue is a benign condition, and no specific treatment is indicated. The patient should be encouraged to brush the tongue, because food or debris entrapped in the grooves may act as a source of irritation.

◆ HAIRY TONGUE (BLACK HAIRY TONGUE; COATED TONGUE)

Hairy tongue is characterized by marked accumulation of keratin on the filiform papillae of the dorsal tongue, resulting in a hairlike appearance. The condition apparently represents an increase in keratin production or a decrease in normal keratin desquamation. Hairy tongue is found in about 0.5% of adults. Although the cause is uncertain, many affected people are heavy smokers. Other possible associated factors include general debilitation, poor oral hygiene, drugs that induce xerostomia, and a history of radiation therapy to the head and neck.

Clinical Features

Hairy tongue most commonly affects the midline just anterior to the circumvallate papillae, sparing the lateral and anterior borders (Fig. 1-23). The elongated papillae are usually brown, yellow, or black as a result of growth of pigment-producing bacteria or staining from tobacco and food. Sometimes most of the dorsal tongue may be involved, resulting in a thick, matted appearance (Fig. 1-24). Multiple individual elongated filiform papillae may be elevated by using gauze or a dental instrument. The condition is typically asymptomatic, although occasionally patients complain of a gagging sensation or a bad taste in the mouth. Because the diagnosis usually can be made from the clinical appearance, biopsy is unnecessary in most instances.

In some individuals, numerous bacteria and desquamated epithelial cells accumulate on the dorsal tongue surface, but without the hairlike filiform projections (Fig. 1-25). Such cases, which often are designated as a **coated**

• **Fig. 1-23 Hairy Tongue.** Elongated, yellowish white filiform papillae on the dorsal surface of the tongue.
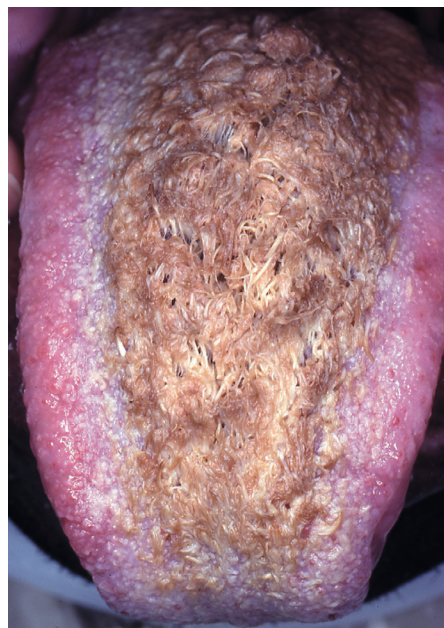
• **Fig. 1-24 Hairy Tongue.** Marked elongation and brown staining of the filiform papillae, resulting in a hairlike appearance.

tongue, also may be a source of oral malodor. Coated tongue often is misdiagnosed as candidiasis and treated unnecessarily with antifungal medications.

Transitory black staining of the dorsal tongue without elongation of the filiform papillae sometimes can occur in

• Fig. 1-25 Coated Tongue. The dorsal tongue appears white and mildly thickened from the accumulation of keratin and bacteria on the surface.

• Fig. 1-26 Bismuth Staining. Transitory staining of the posterior dorsal tongue after using bismuth subsalicylate for an upset stomach.

patients who use bismuth subsalicylate to control upset stomach. The bismuth in such preparations can react with trace amounts of sulfur in the saliva to form bismuth sulfide, which accumulates on the tongue surface (Fig. 1-26). However, this discoloration rapidly resolves after discontinuation of the medication.
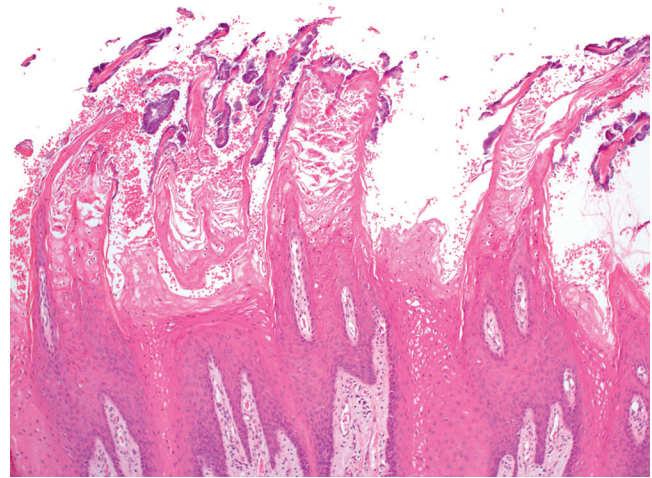
• Fig. 1-27 Hairy Tongue. Elongation and marked hyperkeratosis of the filiform papillae with bacterial accumulation on the surface.

Histopathologic Features

On histopathologic examination, hairy tongue is characterized by marked elongation and hyperparakeratosis of the filiform papillae (Fig. 1-27). Usually, numerous bacteria can be seen growing on the epithelial surface.

Treatment and Prognosis

Hairy or coated tongue is a benign condition with no serious sequelae. The major concern is often the aesthetic appearance of the tongue along with possible associated bad breath. Any predisposing factors, such as tobacco, should be eliminated, and excellent oral hygiene should be encouraged. Periodic scraping or brushing with a toothbrush or tongue scraper can promote desquamation of the hyperkeratotic papillae and surface debris. Keratolytic agents, such as podophyllin, also have been tried with success, but for safety reasons their use probably should not be encouraged.

Because of the similarity in names, care should be taken to avoid confusing hairy tongue with **hairy leukoplakia** (see page 242), which typically occurs on the lateral border of the tongue. Hairy leukoplakia is caused by the Epstein-Barr virus and usually is associated with human immunodeficiency virus (HIV) infection or other immunosuppressive conditions.

◆ VARICOSITIES (VARICES)

Varicosities, or **varices,** are abnormally dilated and tortuous veins. Age appears to be an important etiologic factor because varices are rare in children but common in older adults. This suggests that their development may be an age-related degeneration, in which a loss of connective tissue tone supporting the vessels occurs. One study found that people with varicose veins of the legs are more likely to have varicosities of the tongue. Although some studies have reported no connection between oral varices and

cardiopulmonary diseases, one recent investigation did show a significantly increased prevalence of sublingual varices in patients with a history of smoking or cardiovascular disease.

Clinical Features

The most common type of oral varicosity is the **sublingual varix,** which occurs in two-thirds of people older than 60 years of age. Sublingual varicosities classically present as multiple blue-purple, elevated or papular blebs on the ventral and lateral border of the tongue (Fig. 1-28). The lesions usually are asymptomatic, except in rare instances when secondary thrombosis occurs.

Less frequently, solitary varices occur in other areas of the mouth, especially the lips and buccal mucosa. These isolated varicosities often are first noticed after they have become thrombosed (Fig. 1-29). Clinically, a thrombosed varix presents as a firm, nontender, blue-purple nodule that may feel like a BB beneath the mucosal surface.

Histopathologic Features

Microscopic examination of a varix reveals a dilated vein, the wall of which shows little smooth muscle and poorly developed elastic tissue. If secondary thrombosis has occurred, then the lumen may contain concentrically layered zones of platelets and erythrocytes (lines of Zahn). The clot can undergo organization via granulation tissue with subsequent recanalization. Older thrombi may exhibit dystrophic calcification, resulting in formation of a **phlebolith** (*phlebo* = vein; *lith* = stone).
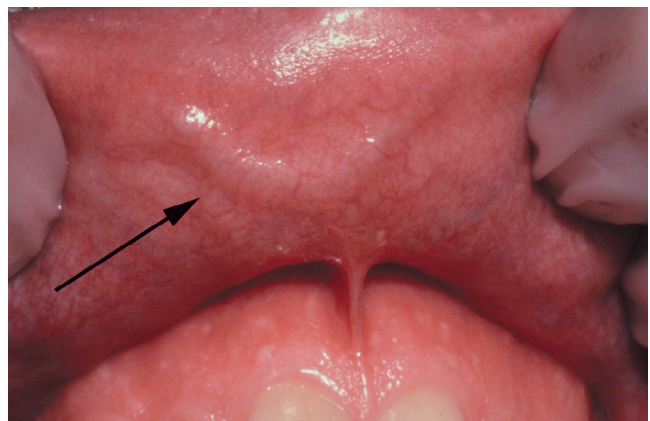
Treatment and Prognosis

Sublingual varicosities typically are asymptomatic, and no treatment is indicated. Solitary varicosities of the lips and buccal mucosa may need to be surgically removed to confirm the diagnosis or for aesthetic purposes.

◆ CALIBER-PERSISTENT ARTERY

A **caliber-persistent artery** is a common vascular anomaly in which a main arterial branch extends up into the superficial submucosal tissues without a reduction in its diameter. Similar to oral varices, caliber-persistent arteries are seen more frequently in older adults. This suggests that their development may be an age-related degenerative phenomenon in which there is a loss of tone in the surrounding supporting connective tissue.

Clinical Features

The caliber-persistent artery occurs almost exclusively on the lip mucosa. Either lip may be affected, and some patients have bilateral lesions or lesions on both lips. The average patient age is 58 years, and the gender ratio is nearly equal. The lesion presents as a linear, arcuate, or papular elevation that ranges from pale to normal to bluish in color (Fig. 1-30). Stretching the lip usually causes the artery to become inconspicuous. The unique feature is pulsation—not only vertically but also in a lateral direction. However, usually it is not possible to feel a pulse in a caliber-persistent artery with gloved fingers.

• **Fig. 1-28 Varicosities.** Multiple purple dilated veins on the ventral and lateral surface of the tongue.

• **Fig. 1-29 Varicosity.** Firm, thrombosed varix on the lower lip.

• **Fig. 1-30 Caliber-Persistent Artery.** Linear, arcuate lesion on the upper labial mucosa *(arrow).* (Courtesy of Dr. John Lovas.)

The lesion is usually asymptomatic, being discovered as an incidental finding during an oral examination; rarely a patient may notice a pulsatile lip nodule. A few cases have been associated with ulceration of the overlying mucosa. In addition, a couple of examples have been found adjacent to labial squamous cell carcinomas, although this is probably coincidental.

Histopathologic Features

Microscopic examination shows a thick-walled artery situated close to the mucosal surface (Fig. 1-31).

Treatment and Prognosis

If the true nature of the caliber-persistent artery can be recognized clinically, no treatment is necessary. Oftentimes a biopsy is performed when the lesion is mistaken for a mucocele or another vascular lesion, such as a varix or hemangioma. Brisk bleeding typically is encountered if the lesion is removed.
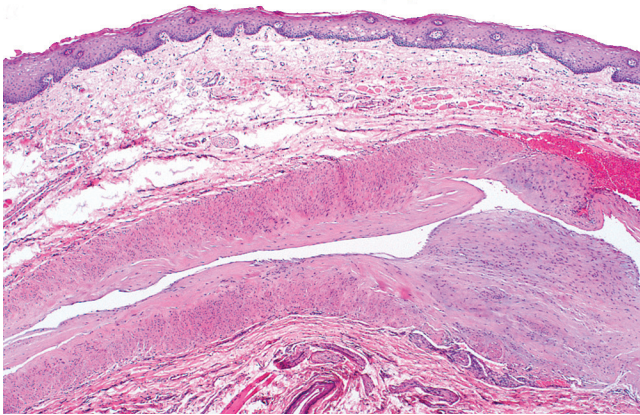
• **Fig. 1-31 Caliber-Persistent Artery.** Thick-walled artery located just beneath the mucosal surface.

◆ LATERAL SOFT PALATE FISTULAS

Lateral soft palate fistulas are rare anomalies of uncertain pathogenesis. Many cases appear to be congenital, possibly related to a defect in the development of the second pharyngeal pouch. Some fistulas may be the result of infection or surgery of the tonsillar region.

Clinical Features

Lateral soft palate fistulas usually are bilateral, but they may occur only on one side. They are more common on the anterior tonsillar pillar (Fig. 1-32), but they also may involve the posterior pillar. The perforations typically are asymptomatic, ranging from a few millimeters to more than 1 cm. A few cases have been associated with other anomalies, such as absence or hypoplasia of the palatine tonsils, hearing loss, and preauricular fistulas.

Treatment and Prognosis

The lesions are innocuous, and no treatment is necessary.

◆ CORONOID HYPERPLASIA

Hyperplasia of the coronoid process of the mandible is a rare developmental anomaly that may result in limitation of mandibular movement. The cause of **coronoid hyperplasia** is unknown, but the condition is three to five times more common in males than in females. Because most cases have been seen in pubertal males, an endocrine influence has been suggested. Heredity also may play a role, because cases have been noted in siblings.

Coronoid hyperplasia may be unilateral or bilateral, although bilateral cases are over four times more common than unilateral examples. Unilateral enlargement of the coronoid process also can result from a true tumor, such as an osteoma or osteochondroma, and such cases should be distinguished from pure coronoid hyperplasia. However,
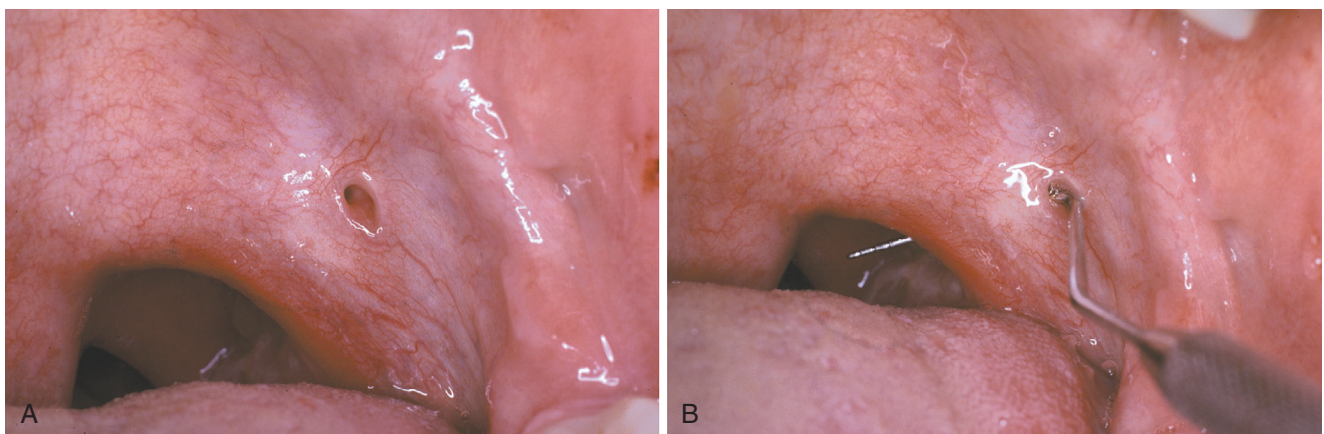
• **Fig. 1-32 Lateral Palatal Fistula. A,** Asymptomatic "hole" in the anterior tonsillar pillar. **B,** Periodontal probe has been used to demonstrate the communication of the lesion with the tonsillar fossa.

some cases reported as tumors of the coronoid process actually may have been hyperplastic processes rather than true neoplasms.

Clinical and Radiographic Features

In a person with unilateral coronoid hyperplasia, the enlarged coronoid process impinges on the posterior surface of the zygoma, restricting mandibular opening. In addition, the mandible may deviate toward the affected side. Usually, there is no pain or associated abnormality in occlusion. Radiographs may reveal an irregular, nodular growth of the tip of the coronoid process.

In bilateral coronoid hyperplasia, the limitation of mandibular opening may progressively worsen over several years during childhood, reaching maximum severity during the late teens. The radiographic appearance is characterized by regular elongation of both processes. Because the coronoid process often is superimposed on the zygoma on conventional radiographs, CT scans often demonstrate the hyperplasia more effectively.

Treatment and Prognosis

Treatment of coronoid hyperplasia consists of surgical removal of the elongated coronoid process or processes to allow freedom of mandibular motion. Coronoidectomy or coronoidotomy usually is accomplished via an intraoral approach. Although initial improvement in oral opening can be effected, the long-term results in some patients can be disappointing because of surgically induced fibrosis and the tendency for coronoid regrowth. Postoperative physiotherapy is important for reestablishing normal function.

◆ CONDYLAR HYPERPLASIA

Condylar hyperplasia is an uncommon malformation of the mandible created by excessive growth of one of the condyles. The cause of this hyperplasia is unknown, but local circulatory problems, endocrine disturbances, and trauma have been suggested as possible etiologic factors.

Condylar hyperplasia can be difficult to distinguish from **hemifacial hyperplasia** (see page 35); however, in the latter condition the associated soft tissues and teeth also may be enlarged.

Clinical and Radiographic Features

Condylar hyperplasia may manifest itself in a variety of ways, including facial asymmetry, prognathism, crossbite, and open bite (Fig. 1-33). Sometimes compensatory maxillary growth and tilting of the occlusal plane occurs. The condition most commonly is discovered in adolescents and young adults. Several studies have shown a significant female predilection, with an overall female-to-male ratio of approximately 3:1.

● **Fig. 1-33 Condylar Hyperplasia.** Enlargement of the patient's right condyle has resulted in facial asymmetry.

The radiographic features are quite variable. Some patients have an enlargement of the condylar head, and others show elongation of the condylar neck (Fig. 1-34). Many cases also demonstrate hyperplasia of the entire ramus, suggesting that the condition sometimes affects more than just the condyle. Single-photon emission computed tomography (SPECT) and scintigraphy using ^{99m}Tc-methylenediphosphonate (MDP) have been advocated as useful methods for assessing the degree of bone activity in condylar hyperplasia.

Histopathologic Features

During active growth, proliferation of the condylar cartilage is noted. Once condylar growth has ceased, the condyle has a normal histologic appearance.

Treatment and Prognosis

Condylar hyperplasia is a self-limiting condition, and treatment is determined by the degree of functional difficulty and aesthetic change. Some patients can be treated with unilateral condylectomy, whereas others require unilateral or bilateral mandibular osteotomies. In patients with compensatory maxillary growth, a maxillary osteotomy also may be needed. Concomitant orthodontic therapy frequently is necessary.

◆ CONDYLAR HYPOPLASIA

Condylar hypoplasia, or underdevelopment of the mandibular condyle, can be either congenital or acquired. **Congenital condylar hypoplasia** often is associated with head and neck syndromes, including **mandibulofacial dysostosis** (see page 41), **oculoauriculovertebral syndrome (Goldenhar syndrome),** and **hemifacial microsomia.** In the most severe cases, complete agenesis of the condyle or ramus **(condylar aplasia)** is seen.

Acquired condylar hypoplasia results from disturbances of the growth center of the developing condyle. The

• **Fig. 1-34 Condylar Hyperplasia.** Panoramic radiograph of patient seen in Fig. 1-33, which shows prominent enlargement of the right mandibular condyle.

most frequent cause is trauma to the condylar region during infancy or childhood. Other causes include infections, radiation therapy, and rheumatoid or degenerative arthritis.

Clinical and Radiographic Features

Condylar hypoplasia can be unilateral or bilateral, producing a small mandible with a Class II malocclusion. Unilateral hypoplasia results in distortion and depression of the face on the affected side. The mandibular midline shifts to the involved side when the mouth is opened, accentuating the deformity. Ankylosis of the temporomandibular joint (TMJ) can develop in cases caused by trauma.

The deformity is observed easily on panoramic films and can range in severity. In severe cases the condyle or ramus may be totally absent. Milder types demonstrate a short condylar process, shallow sigmoid notch, and poorly formed condylar head. A prominent antegonial notch may be present. CT scans may be helpful in evaluating the condyles.

Treatment and Prognosis

Treatment of the patient with condylar hypoplasia depends on the cause and severity of the defect, but surgery often is required. If the condyle is missing, then a costochondral rib graft can be placed to help establish an active growth center. In addition, osteotomies sometimes provide a cosmetically acceptable result. In certain instances, distraction osteogenesis can be used to stimulate new bone formation.

◆ BIFID CONDYLE

A **bifid condyle** is a rare developmental anomaly characterized by a double-headed mandibular condyle. Most bifid condyles have a medial and lateral head divided by an anteroposterior groove. Some condyles may be divided into an anterior and posterior head.

The cause of bifid condyle is uncertain. Anteroposterior bifid condyles may be of traumatic origin, such as a childhood fracture. Mediolaterally divided condyles may result from trauma, abnormal muscle attachment, teratogenic agents, or persistence of a fibrous septum within the condylar cartilage.

Clinical and Radiographic Features

A bifid condyle usually is unilateral, but occasionally both sides may be affected. The malformation is often asymptomatic and may be discovered on routine radiographs, although some patients may have a "pop" or "click" of the TMJ when opening their mouths. Panoramic radiographs and CT scans demonstrate a bilobed appearance of the condylar head (Fig. 1-35). Extremely rare examples of trifid and tetrafid condyles also have been reported.

Treatment and Prognosis

Because a bifid condyle is usually asymptomatic, no treatment is necessary in most instances. If the patient has joint
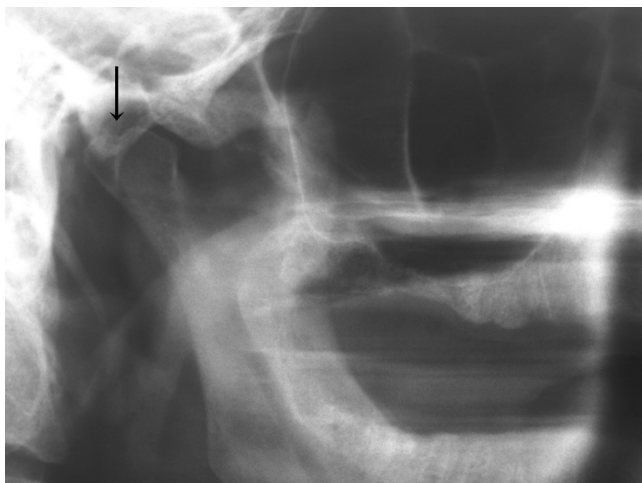
• **Fig. 1-35 Bifid Condyle.** Radiograph of the mandibular condyle showing a double head *(arrow).*

• **Fig. 1-36 Exostoses.** Multiple buccal exostoses of the maxillary and mandibular alveolar ridges.

complaints, the appropriate temporomandibular therapy may be required.

◆ EXOSTOSES

Exostoses are localized bony protuberances that arise from the cortical plate. These benign growths frequently affect the jaws and may be related to stresses placed on the bone from the function of teeth. The best-known oral exostoses, the **torus palatinus** and the **torus mandibularis,** are described later in this chapter. Other types of exostoses also may affect the jaws and are considered here.

Clinical and Radiographic Features

Exostoses are discovered most often in adults. **Buccal exostoses** occur as a bilateral row of bony hard nodules along the facial aspect of the maxillary and/or mandibular alveolar ridge (Fig. 1-36). They usually are asymptomatic, unless the thin overlying mucosa becomes ulcerated from trauma. One
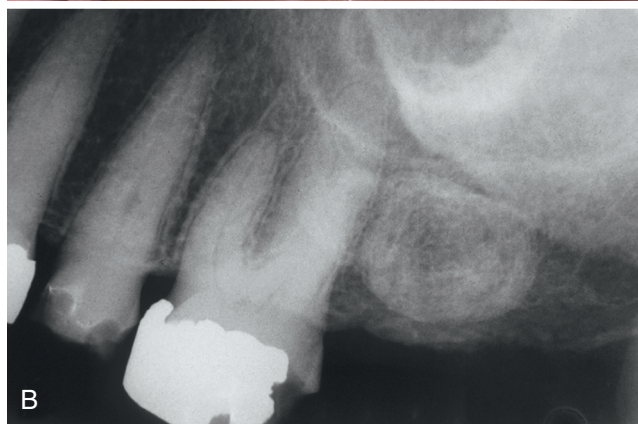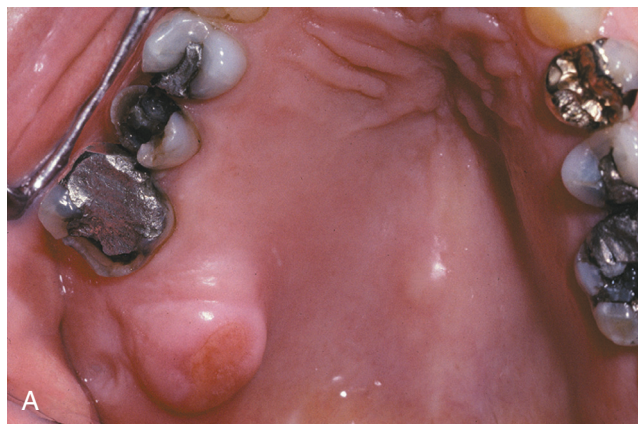
• **Fig. 1-37 Exostosis. A,** Secondarily ulcerated palatal exostosis. **B,** Radiograph shows an ovoid radiopacity distal to the molar.

study reported that buccal exostoses were found in nearly 1 of every 1000 adults (0.09%); however, a more recent survey found a much higher prevalence of nearly 19%. This variation may be due to the different populations being studied or to the clinical criteria used to make the diagnosis.

Palatal exostoses (palatal tubercles) are similar bony protuberances that develop from the lingual aspect of the maxillary tuberosities. These lesions usually are bilateral but may affect only one side (Fig. 1-37). They are more common in males and have been reported in 8% to 69% of various populations. Many patients with buccal or palatal exostoses also will have palatal or mandibular tori (Fig. 1-38).

Less commonly, **solitary exostoses** may occur, possibly in response to local irritation. Such lesions may develop from the alveolar bone beneath free gingival grafts and skin grafts. Presumably placement of the graft acts as a stimulant to the periosteum to form new bone.

Another uncommon, interesting variant is the **reactive subpontine exostosis (subpontic osseous proliferation, subpontic osseous hyperplasia),** which may develop from the alveolar crestal bone beneath the pontic of a posterior bridge (Fig. 1-39).

If enough excess bone is present, exostoses may exhibit a relative radiopacity on dental radiographs (see Fig. 1-37, B). In rare instances an exostosis may become so large that distinguishing it from a tumor, such as an osteoma, is difficult (see page 605).
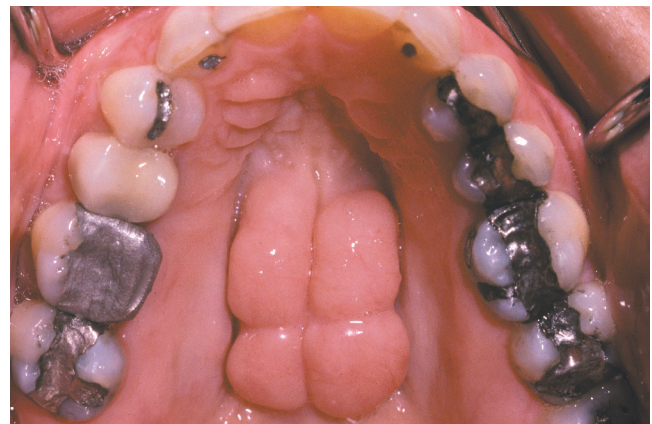
• **Fig. 1-38 Palatal Exostoses and Torus Palatinus.** Massive bilateral palatal exostoses in a patient with a large palatal torus.
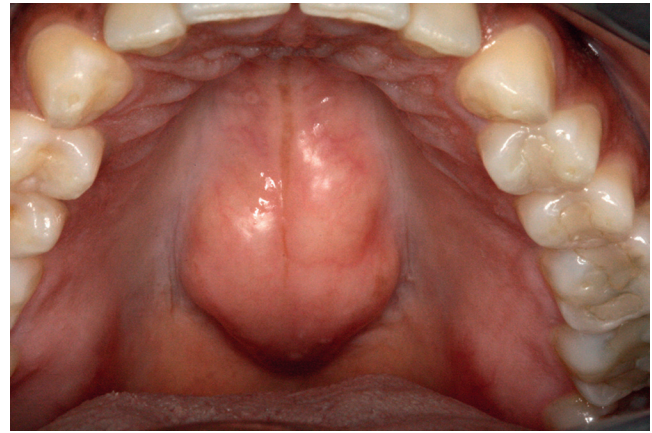
• **Fig. 1-40 Torus Palatinus.** Midline bony nodule of the palatal vault.
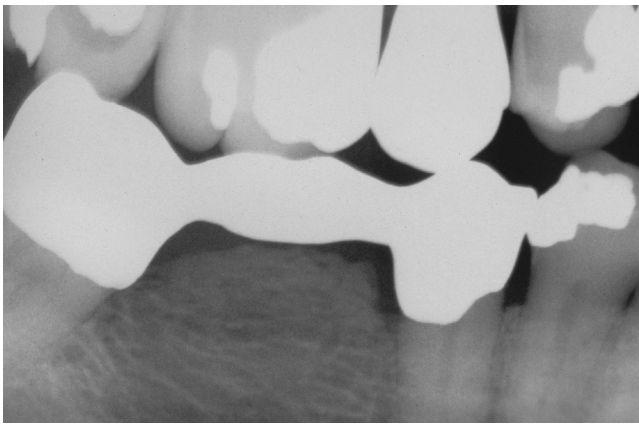
• **Fig. 1-39 Reactive Subpontine Exostosis.** Nodular growth of bone beneath the pontic of a posterior mandibular bridge.

• **Fig. 1-41 Torus Palatinus.** Large, lobulated palatal mass.

Histopathologic Features

Microscopic examination reveals a mass of dense, lamellar, cortical bone with a small amount of fibrofatty marrow. In some cases an inner zone of trabecular bone also is present.

Treatment and Prognosis

Most exostoses are distinctive enough clinically to make biopsy unnecessary. If the diagnosis is uncertain, biopsy should be performed to rule out other bony pathosis. Sometimes the exostosis must be removed if it repeatedly has been exposed to trauma or has become ulcerated and painful. In addition, surgical removal may be required to accommodate a dental prosthesis or to allow for proper flap adaptation during periodontal surgery. Reactive subpontine exostoses may need to be removed if they interfere with oral hygiene or are associated with adjacent periodontal disease. Exostoses that develop secondary to adjacent tooth function may recur after removal if the teeth creating the stresses remain in place.

◆ TORUS PALATINUS

The **torus palatinus** is a common exostosis that occurs in the midline of the vault of the hard palate. The pathogenesis of these tori has long been debated with arguments centering on genetic versus environmental factors, such as masticatory stress. Some authorities have suggested that the torus palatinus is inherited as an autosomal dominant trait. However, others believe that the development of this lesion is multifactorial, including both genetic and environmental influences. In this model, patients are affected by a variety of hereditary and local environmental factors. If enough of these factors are present, then a "threshold" is surpassed and the trait (torus palatinus) is expressed.

Clinical and Radiographic Features

The torus palatinus presents as a bony hard mass that arises along the midline suture of the hard palate (Figs. 1-40 and 1-41). Tori sometimes are classified according to their morphologic appearance:

• The **flat torus** has a broad base and a slightly convex, smooth surface. It extends symmetrically onto both sides of the midline raphe.

- The **spindle torus** has a midline ridge along the palatal raphe. A median groove is sometimes present.
- The **nodular torus** arises as multiple protuberances, each with an individual base. These protuberances may coalesce, forming grooves between them.
- The **lobular torus** is also a multilobulated mass, but it rises from a single base. Lobular tori can be either sessile or pedunculated.

Most palatal tori are small, measuring less than 2 cm in diameter; however, they can slowly increase in size throughout life—sometimes to the extent that they fill the entire palatal vault. Most tori cause no symptoms, but in some cases the thin overlying mucosa may become ulcerated secondary to trauma.

The torus palatinus usually does not appear on routine dental radiographs. Rarely, it may be seen as a radiopacity on periapical films if the film is placed behind the torus when the radiograph is taken.

The prevalence of palatal tori has varied widely in a number of population studies, ranging from 9% to 60%. Some of this variation may be due to the criteria used to make the diagnosis and also may be based on whether the study was conducted on live patients or skulls. There appear to be significant racial differences, however, with a higher prevalence in Asian and Inuit populations. In the United States, most studies have shown a prevalence of 20% to 35%, although these figures likely include a significant number of relatively small lesions. Almost all studies from around the world have shown a pronounced female-to-male ratio of 2 : 1. The prevalence peaks during early adult life, tapering off in later years. This finding supports the theory that tori are dynamic lesions that are related, in part, to environmental factors; in later life, some may undergo resorption remodeling in response to decreased functional stresses.

Histopathologic Features

Microscopic examination of the torus shows a mass of dense, lamellar, cortical bone. An inner zone of trabecular bone sometimes is seen.

Treatment and Prognosis

Most palatal tori can be diagnosed clinically based on their characteristic appearance; therefore biopsy rarely is necessary. In edentulous patients, the torus may need to be removed surgically to accommodate a denture base. Surgical removal also may be indicated for palatal tori that repeatedly become ulcerated or that interfere with oral function. It also should be noted that palatal tori are prone to medication-related osteonecrosis (see page 271).

◆ TORUS MANDIBULARIS

The **torus mandibularis** is a common exostosis that develops along the lingual aspect of the mandible. As with torus palatinus, the cause of mandibular tori probably is
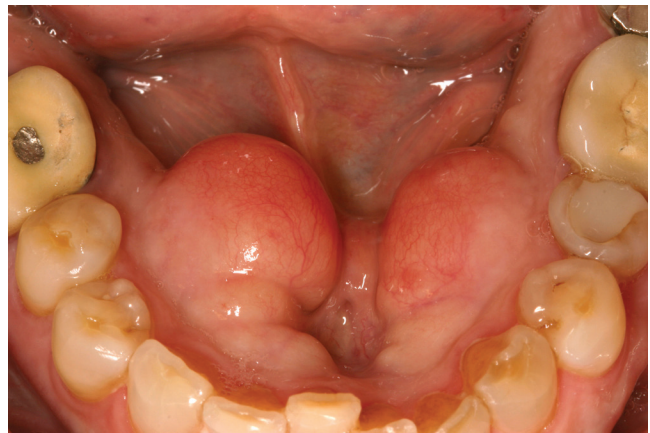
Fig. 1-42 Torus Mandibularis. Bilateral lobulated bony protuberances of the mandibular lingual alveolar ridge.

Fig. 1-43 Torus Mandibularis. Massive "kissing" tori meet in the midline.

multifactorial, including both genetic and environmental influences.

Clinical and Radiographic Features

The mandibular torus presents as a bony protuberance along the lingual aspect of the mandible above the mylohyoid line in the region of the premolars (Fig. 1-42). Bilateral involvement occurs in more than 90% of cases. Most mandibular tori occur as single nodules, although multiple lobules paralleling the teeth are not unusual. Patients often are unaware of their presence unless the overlying mucosa becomes ulcerated secondary to trauma. In rare instances, bilateral tori may become so large that they almost meet in the midline (Fig. 1-43). A large mandibular torus may appear on periapical radiographs as a radiopacity superimposed on the roots of the teeth (Fig. 1-44), especially on anterior films. Mandibular tori are easily visualized on occlusal radiographs (Fig. 1-45).

Most studies indicate that the torus mandibularis is not as common as the torus palatinus; the prevalence ranges from 5% to 40%. Like the torus palatinus, the mandibular torus appears to be more common in Asians and the Inuit. The prevalence in the United States ranges from 7% to 10%

• **Fig. 1-44 Torus Mandibularis.** Torus is causing a radiopacity that is superimposed over the roots of the mandibular teeth.

• **Fig. 1-45 Torus Mandibularis.** Occlusal radiograph showing bilateral mandibular tori.
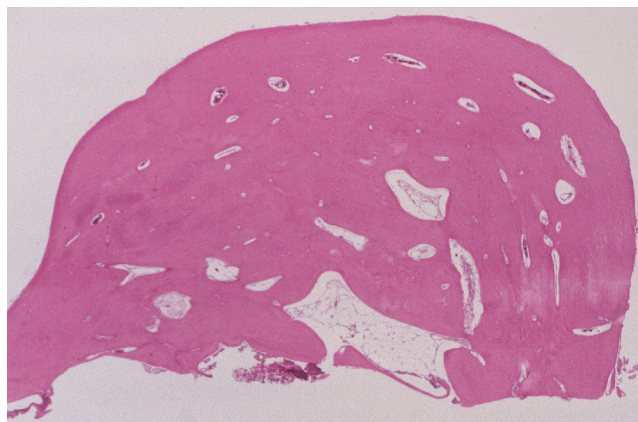
• **Fig. 1-46 Torus Mandibularis.** Nodular mass of dense, cortical bone. Some fatty marrow is visible at the base of the specimen.

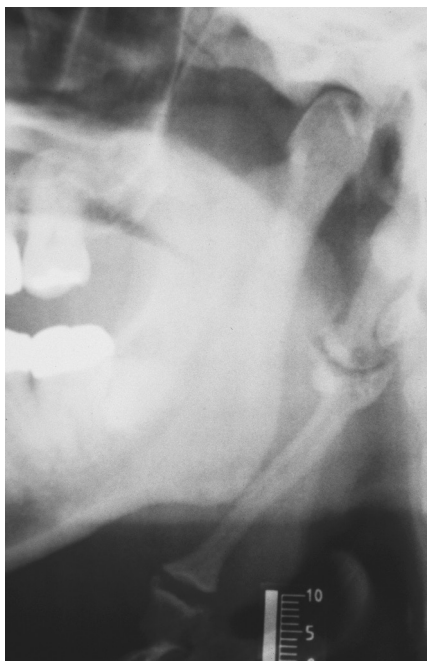## ◆ EAGLE SYNDROME (STYLOHYOID SYNDROME; CAROTID ARTERY SYNDROME; STYLALGIA)

The styloid process is a slender bony projection that originates from the inferior aspect of the temporal bone, anterior and medial to the stylomastoid foramen. It is connected to the lesser cornu of the hyoid bone by the stylohyoid ligament. The external and internal carotid arteries lie on either side. Elongation of the styloid process or mineralization of the stylohyoid ligament complex is not unusual, having been reported in 18% to 84% of the population with an increasing incidence with advancing age. Such mineralization is usually bilateral, but it may affect only one side. Most cases are asymptomatic; however, a small number of such patients experience symptoms of **Eagle syndrome,** caused by impingement or compression of adjacent nerves or blood vessels.

with little difference between blacks and whites. A slight male predilection has been noted.

The prevalence of mandibular torus peaks in early adult life, tapering slightly in later years. In addition, the prevalence has been correlated with both bruxism and the number of teeth remaining present. These findings support the theory that the torus mandibularis is multifactorial in development and responds to functional stresses.

Histopathologic Features

The histopathologic appearance of the torus mandibularis is similar to that of other exostoses, consisting primarily of a nodular mass of dense, cortical lamellar bone (Fig. 1-46). An inner zone of trabecular bone with associated fatty marrow sometimes is visible.

Treatment and Prognosis

Most mandibular tori are easily diagnosed clinically, and no treatment is necessary. However, surgical removal may be required to accommodate a lower full or partial denture. Occasionally, tori may recur if teeth are still present in the area.

Clinical and Radiographic Features

Eagle syndrome most commonly affects adults, occurring more often in women than men. The patient experiences vague facial pain, especially while swallowing, turning the head, or opening the mouth. Other symptoms may include dysphagia, dysphonia, otalgia, headache, dizziness, syncope, and transient ischemic attacks.

Elongation of the styloid process or mineralization of the stylohyoid ligament complex can be seen on panoramic or lateral-jaw radiographs (Fig. 1-47). The mineralized stylohyoid complex may be palpated in the tonsillar fossa area, and pain often is elicited.

Classic Eagle syndrome occurs after a tonsillectomy. Development of scar tissue in the area of a mineralized stylohyoid complex then results in cervicopharyngeal pain in the region of cranial nerves V, VII, IX, and X, especially during swallowing. Some authors reserve the term *Eagle syndrome* only for those cases in which the ossification of the stylohyoid chain occurs as a result of the tonsillectomy or other neck trauma.

• **Fig. 1-47 Eagle Syndrome.** Mineralization of the stylohyoid ligament is visible posterior to the mandibular ramus.

A second form of this condition unrelated to tonsillectomy is sometimes known as **carotid artery syndrome** or **stylohyoid syndrome.** The elongated, mineralized complex is thought to impinge on the internal or external carotid arteries and associated sympathetic nerve fibers. The patient may complain of pain in the neck when turning the head, and this pain may radiate to other sites in the head or neck.

Traumatic Eagle syndrome also has been reported, in which symptoms develop after fracture of a mineralized stylohyoid ligament.

Treatment and Prognosis

Treatment of Eagle syndrome depends on the severity of the symptoms. For mild cases, no treatment may be necessary (except reassurance of the patient). Local injection of corticosteroids sometimes provides relief. In more severe cases, partial surgical excision of the elongated styloid process or mineralized stylohyoid ligament is required. Usually, this is accomplished via an intraoral approach, although an extraoral approach also can be used. The prognosis is good.

◆ STAFNE DEFECT (STAFNE BONE CYST; LINGUAL MANDIBULAR SALIVARY GLAND DEPRESSION; LATENT BONE CYST; STATIC BONE CYST; STATIC BONE DEFECT; LINGUAL CORTICAL MANDIBULAR DEFECT)

In 1942, Stafne described a series of asymptomatic radiolucent lesions located near the angle of the mandible.

• **Fig. 1-48 Stafne Defect.** Radiolucency of the posterior mandible below the mandibular canal.

Subsequent reports of similar lesions have shown that this condition represents a focal concavity of the cortical bone on the lingual surface of the mandible. In most cases, biopsy has revealed histologically normal salivary gland tissue, suggesting that these lesions represent developmental defects containing a portion of the submandibular gland. However, a few of these defects have been reported to be devoid of contents or to contain muscle, fibrous connective tissue, blood vessels, fat, or lymphoid tissue.

Similar lingual cortical defects also have been noted more anteriorly in the mandible, in the area of the incisor, canine, or premolar teeth. These rare defects have been related to the sublingual gland or to aberrant salivary gland tissue. In addition, one report has implicated the parotid gland as the cause of an apparent cortical defect in the upper mandibular ramus. Therefore, all of the major salivary glands appear to be capable of causing such cortical concavities.

In rare examples, the radiolucent defect has been reported to be totally surrounded by intact bone. Such cases might be explained by entrapment of embryonic salivary gland tissue within the jawbone.

Clinical and Radiographic Features

The classic **Stafne defect** presents as an asymptomatic radiolucency below the mandibular canal in the posterior mandible, between the molar teeth and the angle of the mandible (Fig. 1-48). The lesion is typically well circumscribed and has a sclerotic border. Sometimes the defect may interrupt the continuity of the inferior border of the mandible, with a palpable notch observed clinically in this area. Most Stafne defects are unilateral, although bilateral cases may be seen. Anterior lingual salivary defects associated with the sublingual gland present as well-defined radiolucencies that may appear superimposed over the apices of the anterior teeth (Fig. 1-49).

Posterior Stafne defects are not rare, having been reported in 0.08% to 0.48% of panoramic radiographs. A striking male predilection is observed, with 80% to 90% of all cases seen in men.

Although the defect is believed to be developmental in nature, it does not appear to be present from birth. Most cases have been reported in middle-aged and older adults, with children rarely affected; this implies that the lesion usually "develops" at a later age. Stafne defects typically

• **Fig. 1-49 Stafne Defect.** Anterior radiolucent lesion of the body of the mandible associated with the sublingual gland.

remain stable in size; hence the name **static bone cyst.** In a few cases, however, the lesion has increased in size over time (Fig. 1-50). This also indicates that these lesions are not congenital.

The diagnosis usually can be made on a clinical basis by the typical radiographic location and lack of symptoms. If the clinical diagnosis is in doubt, then it can be confirmed by conventional CT scans, cone beam CT, MRI, or sialography. CT scans and MRIs show a well-defined concavity on the lingual surface of the mandible. Sialograms may be able to demonstrate the presence of salivary gland tissue in the area of the defect.

Histopathologic Features

Because of the typical radiographic appearance, biopsy usually is not necessary to establish the diagnosis of Stafne defects of the posterior mandible. If biopsy is performed, normal submandibular gland tissue usually is seen. However, some defects are devoid of tissue or contain muscle, blood vessels, fat, connective tissue, or lymphoid tissue. In cases reported to be devoid of contents, it is possible that the gland was simply displaced at the time of biopsy.

Treatment and Prognosis

No treatment is necessary for patients with Stafne defects, and the prognosis is excellent. If the diagnosis is in question on plain films, appropriate CT imaging studies can confirm the presence of a well-defined, cupped-out lingual cortical defect, allowing a presumptive diagnosis to be made on a radiographic basis. Because anterior lingual salivary defects
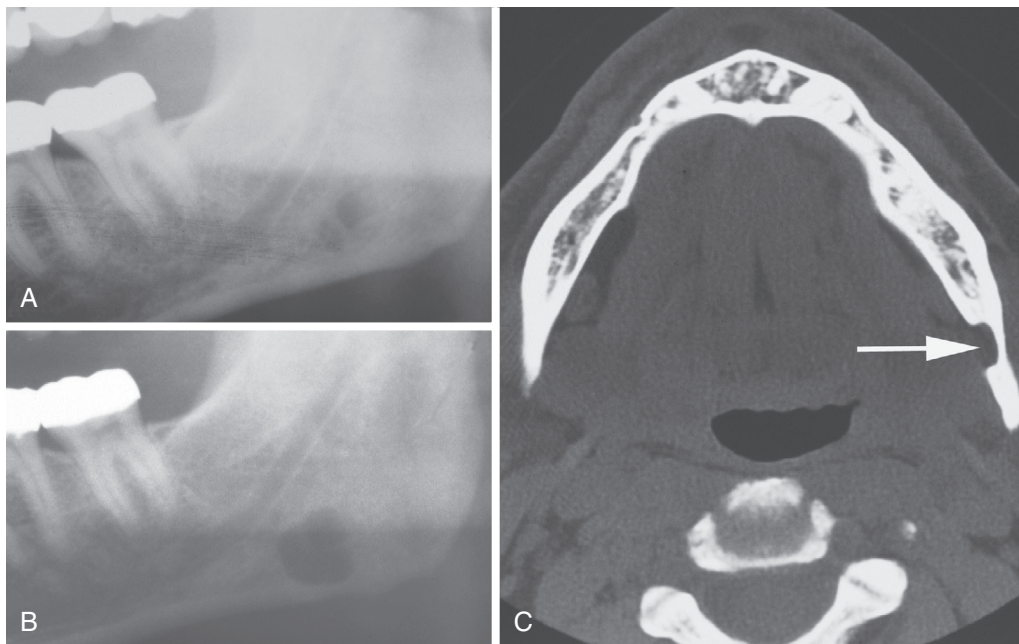
• **Fig. 1-50 Stafne Defect. A,** Ill-defined radiolucency near the angle of the mandible. **B,** Appearance of the same defect several years later showing enlargement of the lesion. **C,** Computed tomography (CT) image of the same lesion showing a left lingual cortical defect *(arrow).* (Courtesy of Dr. Carroll Gallagher.)

are more difficult to recognize, the diagnosis often is not suspected, and biopsy may be done to rule out other pathologic lesions.

DEVELOPMENTAL CYSTS

By definition, a **cyst** is a pathologic cavity (often fluid-filled) that is lined by epithelium. A number of different developmental cysts of the head and neck have been described. Some of these have been considered historically as "fissural" cysts because they were thought to arise from epithelium entrapped along embryonal lines of fusion. However, the concept of a fissural origin for many of these cysts has been questioned in more recent years. In many instances the exact pathogenesis of these lesions is still uncertain. Regardless of their origin, once cysts develop in the oral and maxillofacial region, they tend to slowly increase in size, possibly in response to a slightly elevated hydrostatic luminal pressure.

◆ PALATAL CYSTS OF THE NEWBORN (EPSTEIN'S PEARLS; BOHN'S NODULES)

Small developmental cysts are a common finding on the palate of newborn infants. Researchers have theorized that these "inclusion" cysts may arise in one of two ways. First, as the palatal shelves meet and fuse in the midline during embryonic life to form the secondary palate, small islands of epithelium may become entrapped below the surface along the median palatal raphe and form cysts. Second, these cysts may arise from epithelial remnants derived from the development of the minor salivary glands of the palate.

As originally described, **Epstein's pearls** occur along the median palatal raphe and presumably arise from epithelium entrapped along the line of fusion. **Bohn's nodules** are scattered over the hard palate, often near the soft palate junction and are believed to be derived from the minor salivary glands. However, these two terms have been used almost interchangeably in the literature and also have often been used to describe gingival cysts of the newborn (see page 644), similar-appearing lesions of dental lamina origin. Therefore, the term **palatal cysts of the newborn** may be preferable to help distinguish them from gingival cysts of the newborn. In addition, because these cysts are most common near the midline at the junction of the hard and soft palates, it is usually difficult to ascertain clinically whether they are arising from epithelium entrapped by fusion of the palate or from the developing minor salivary glands.

Clinical Features

Palatal cysts of the newborn are quite common and have been reported in as many as 55% to 85% of neonates. The cysts are small, 1- to 3-mm, white or yellow-white papules that appear most often along the midline near the junction
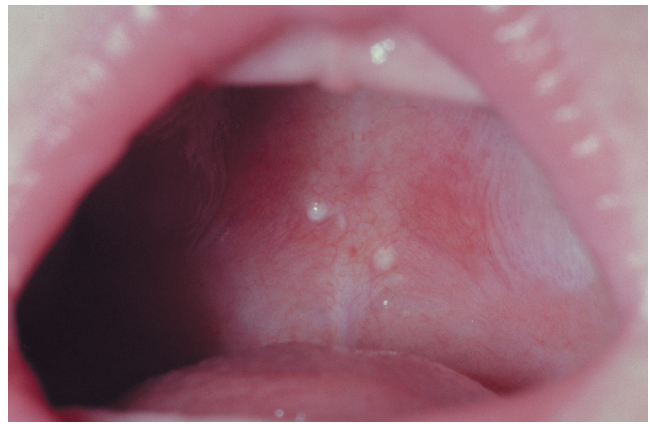
● **Fig. 1-51 Epstein's Pearls.** Small keratin-filled cysts at the junction of the hard and soft palates. (Courtesy of Tristan Neville.)

of the hard and soft palates (Fig. 1-51). Occasionally, they may occur in a more anterior location along the raphe or on the posterior palate lateral to the midline. Frequently a cluster of two to six cysts is observed, although the lesions also can occur singly.

Histopathologic Features

Microscopic examination reveals keratin-filled cysts that are lined by stratified squamous epithelium. Sometimes these cysts demonstrate a communication with the mucosal surface.

Treatment and Prognosis

Palatal cysts of the newborn are innocuous lesions, and no treatment is required. They are self-healing and rarely observable several weeks after birth. Presumably the epithelium degenerates, or the cysts rupture onto the mucosal surface and eliminate their keratin contents.

◆ NASOLABIAL CYST (NASOALVEOLAR CYST; KLESTADT CYST)

The **nasolabial cyst** is a rare developmental cyst that occurs in the upper lip lateral to the midline. The pathogenesis is uncertain, although there are two major theories. One theory considers the nasolabial cyst to be a "fissural" cyst arising from epithelial remnants entrapped along the line of fusion of the maxillary, medial nasal, and lateral nasal processes. A second theory suggests that these cysts develop from misplaced epithelium of the nasolacrimal duct because of their similar location and histologic appearance.

Clinical and Radiographic Features

The nasolabial cyst usually appears as a swelling of the upper lip lateral to the midline, resulting in elevation of the ala of
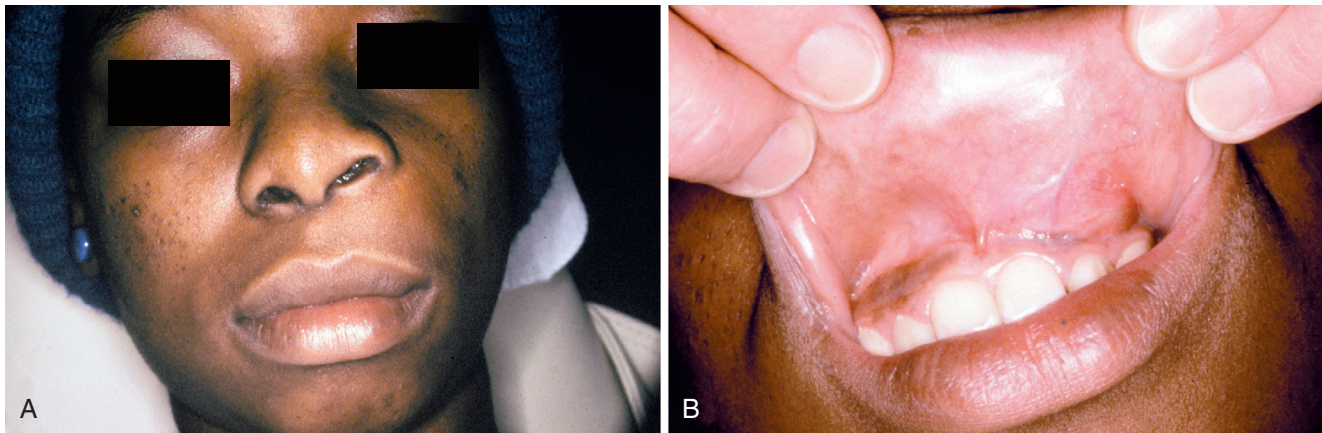
• **Fig. 1-52 Nasolabial Cyst. A,** Enlargement of the left upper lip with elevation of the ala of the nose.
B, Intraoral swelling fills the maxillary labial fold. (Courtesy of Dr. Jim Weir.)

the nose. The enlargement often elevates the mucosa of the nasal vestibule and obliterates the maxillary mucolabial fold (Fig. 1-52). On occasion, this expansion may result in nasal obstruction or may interfere with the wearing of a denture. Pain is uncommon unless the lesion is secondarily infected. The cyst may rupture spontaneously and may drain into the oral cavity or nose.

Nasolabial cysts are most commonly seen in adults, with peak prevalence in the fourth and fifth decades of life. A significant predilection exists for women, with a female-to-male ratio of 3 : 1. Approximately 10% of the reported cases have been bilateral.

Because the nasolabial cyst arises in soft tissues, in most cases no radiographic changes are seen. Occasionally, pressure resorption of the underlying bone may occur.

Histopathologic Features

The nasolabial cyst is characteristically lined by pseudostratified columnar epithelium, often demonstrating goblet cells and cilia (Fig. 1-53). Areas of cuboidal epithelium and squamous metaplasia are not unusual. Apocrine changes also have been reported. The cyst wall is composed of fibrous connective tissue with adjacent skeletal muscle. Inflammation may be seen if the lesion is secondarily infected.

Treatment and Prognosis

Complete surgical excision of the cyst via an intraoral approach has been the traditional treatment of choice. Because the lesion is often close to the floor of the nose, it is sometimes necessary to sacrifice a portion of the nasal mucosa to ensure total removal. However, an alternative transnasal approach has been developed that allows endoscopic marsupialization of the lesion, converting the cyst into an air-containing sinus with its opening on the nasal floor. Recurrence is rare.
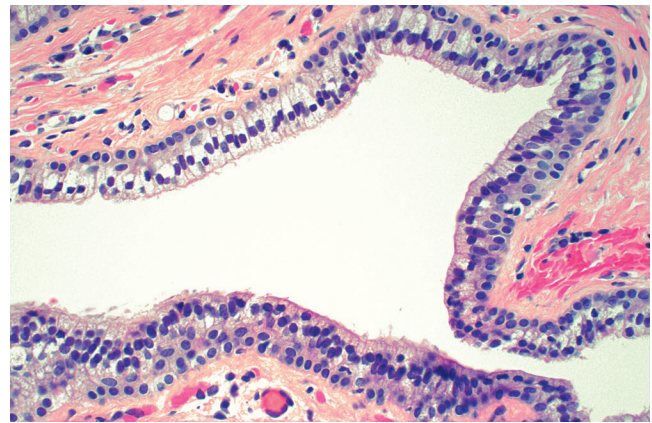
• **Fig. 1-53 Nasolabial Cyst.** Pseudostratified columnar epithelial lining.

◆ "GLOBULOMAXILLARY CYST"

As originally described, the "**globulomaxillary cyst**" was purported to be a fissural cyst that arose from epithelium entrapped during fusion of the globular portion of the medial nasal process with the maxillary process. This concept has been questioned, however, because the globular portion of the medial nasal process is primarily united with the maxillary process and a fusion does not occur. Therefore, epithelial entrapment should not occur during embryologic development of this area.

Virtually all cysts in the globulomaxillary region (between the lateral incisor and canine teeth) can be explained on an odontogenic basis. Many are lined by inflamed stratified squamous epithelium and are consistent with **periapical cysts** (see page 119). Some exhibit specific histopathologic features of an **odontogenic keratocyst** (see page 636) or developmental **lateral periodontal cyst** (see page 645). On rare occasions, cysts in the globulomaxillary area may be lined by pseudostratified, ciliated, columnar epithelium. Such cases may lend credence to the fissural theory of

origin. However, this epithelium may be explained by the close proximity of the sinus lining. In addition, respiratory epithelium also has been reported in periapical cysts, dentigerous cysts, and glandular odontogenic cysts found in other locations.

Because a fissural cyst in this region probably does not exist, the term *globulomaxillary cyst* should no longer be used. When a radiolucency between the maxillary lateral incisor and canine is encountered, the clinician should first consider an odontogenic origin for the lesion.

◆ NASOPALATINE DUCT CYST (INCISIVE CANAL CYST)

The **nasopalatine duct cyst** is the most common nonodontogenic cyst of the oral cavity, occurring in about 1% of the population. The cyst is believed to arise from remnants of the **nasopalatine duct,** an embryologic structure connecting the oral and nasal cavities in the area of the incisive canal.

In the 7-week-old fetus, the developing palate consists of the **primary palate,** which is formed by the fusion of the medial nasal processes. Behind the primary palate, downgrowth of the nasal septum produces two communications between the oral and nasal cavities, the primitive nasal choanae. Formation of the **secondary palate** begins around the eighth intrauterine week, with downward growth of the medial parts of the maxillary processes (palatine processes) to a location on either side of the tongue.

As the mandible develops and the tongue drops down, these palatine processes grow horizontally, fusing with the nasal septum in the midline and with the primary palate along their anterior aspect. Two passageways persist in the midline between the primary and secondary palates (the **incisive canals**). Also formed by this fusion and found within the incisive canals are epithelial structures—the **nasopalatine ducts.** These ducts normally degenerate in humans but may leave epithelial remnants behind in the incisive canals.

The incisive canals begin on the floor of the nasal cavity on either side of the nasal septum, coursing downward and forward to exit the palatal bone via a common foramen in the area of the incisive papilla. In addition to the nasopalatine ducts, these canals contain the nasopalatine nerve plus anastomosing branches of the descending palatine and sphenopalatine arteries. Occasionally, two smaller foramina carrying the nasopalatine nerves—the **canals of Scarpa**—are found within the incisive foramen.

In some mammals the nasopalatine ducts remain patent and provide communication between the oral and nasal cavities. On rare occasions, patent or partially patent nasopalatine ducts may be encountered in humans. In mammals the nasopalatine ducts may communicate with the vomeronasal **organ of Jacobson,** acting as an accessory olfactory organ. However, in humans, Jacobson's organ usually recedes in uterine life to become a vestigial structure.
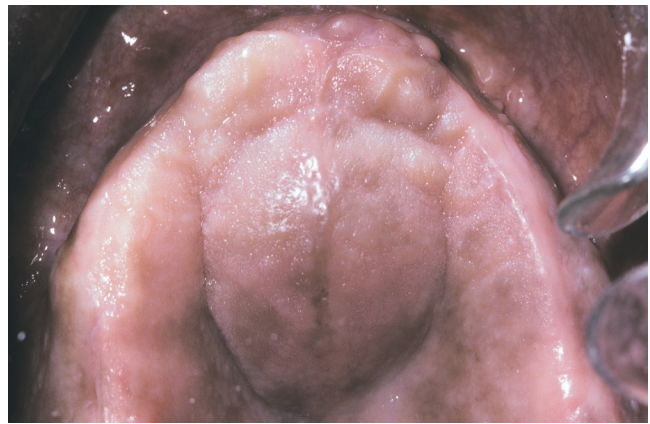
• Fig. 1-54 Nasopalatine Duct Cyst. Fluctuant swelling of the anterior hard palate.

Researchers have suggested that the nasopalatine duct cyst may arise from the epithelium of Jacobson's organ, but this appears highly unlikely. Trauma or infection of the duct and mucous retention of adjacent minor salivary glands also have been mentioned as possible etiologic factors, but the role of each has been questioned. Although the pathogenesis of this lesion is still uncertain, the lesion most likely represents a spontaneous cystic degeneration of remnants of the nasopalatine duct.

Clinical and Radiographic Features

The nasopalatine duct cyst may develop at almost any age but is most common in the fourth to sixth decades of life. In spite of its being a "developmental" cyst, the nasopalatine duct cyst is rarely seen during the first decade. Most studies have shown a male predilection.

The most common presenting symptoms include swelling of the anterior palate, drainage, and pain (Fig. 1-54). Patients sometimes relate a long history of these symptoms, probably because of their intermittent nature. However, many lesions are asymptomatic and are discovered on routine radiographs. Rarely a large cyst may produce a "through-and-through" fluctuant expansion involving the anterior palate and labial alveolar mucosa.

Radiographs usually demonstrate a well-circumscribed radiolucency in or near the midline of the anterior maxilla, between and apical to the central incisor teeth (Figs. 1-55 and 1-56). Root resorption rarely is noted. The lesion most often is round or oval with a sclerotic border. Some cysts may have an inverted pear shape, presumably because of resistance of adjacent tooth roots. Other examples may show a classic heart shape as a result of superimposition of the nasal spine or because they are notched by the nasal septum.

The radiographic diameter of nasopalatine duct cysts can range from small lesions, less than 6 mm, to destructive lesions as large as 6 cm. However, most cysts are in the range of 1.0 to 2.5 cm, with an average diameter of 1.5 to 1.7 cm.
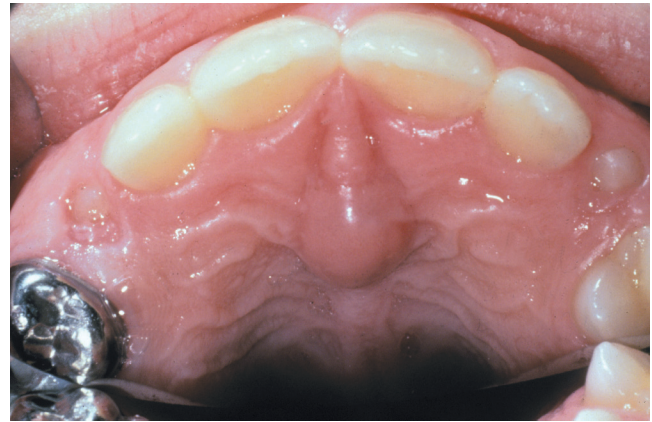
• **Fig. 1-55 Nasopalatine Duct Cyst.** Well-circumscribed radiolucency between and apical to the roots of the maxillary central incisors.
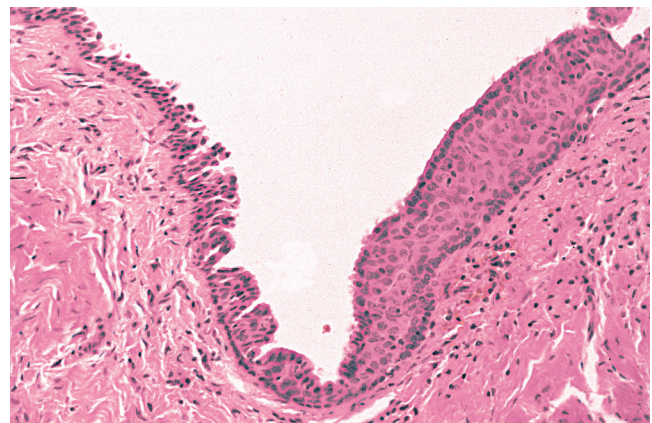

• **Fig. 1-56 Nasopalatine Duct Cyst.** Large destructive cyst of the palate.

• **Fig. 1-57 Cyst of the Incisive Papilla.** Swelling of the incisive papilla.

• **Fig. 1-58 Nasopalatine Duct Cyst.** Cystic lining showing transition from pseudostratified columnar to stratified squamous epithelium.

It may be difficult to distinguish a small nasopalatine duct cyst from a large incisive foramen. It is generally accepted that a diameter of 6 mm is the upper limit of normal size for the incisive foramen. Therefore, a radiolucency that is 6 mm or smaller in this area usually is considered a normal foramen unless other clinical signs or symptoms are present.

In rare instances, a nasopalatine duct cyst may develop in the soft tissues of the incisive papilla area without any bony involvement. Such lesions often are called **cysts of the incisive papilla.** These cysts frequently demonstrate bluish discoloration as a result of the fluid contents in the cyst lumen (Fig. 1-57).

Histopathologic Features

The epithelial lining of nasopalatine duct cysts is highly variable (Figs. 1-58 and 1-59). It may be composed of the following:
- Stratified squamous epithelium
- Pseudostratified columnar epithelium
- Simple columnar epithelium
- Simple cuboidal epithelium

Frequently, more than one epithelial type is found in the same cyst. Stratified squamous epithelium is most common, present in at least three fourths of all cysts. Pseudostratified columnar epithelium has been reported in from one-third to three-fourths of all cases. Simple cuboidal or columnar epithelium is discovered less frequently.

Cilia and goblet cells may be found in association with columnar linings. The type of epithelium may be related to the vertical position of the cyst within the incisive canal.
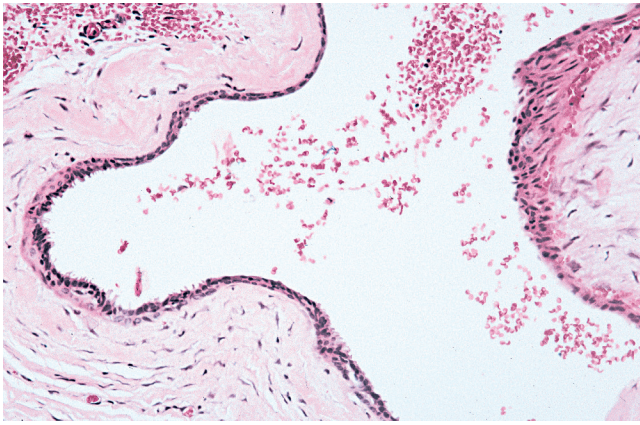
• **Fig. 1-59 Nasopalatine Duct Cyst.** Flattened cuboidal epithelial lining.
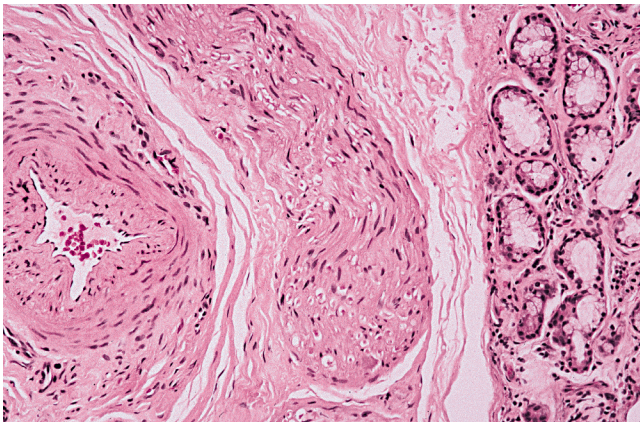
• **Fig. 1-60 Nasopalatine Duct Cyst.** Cyst wall showing blood vessels, nerve bundles, and minor salivary glands.

Cysts developing within the superior aspect of the canal near the nasal cavity are more likely to demonstrate respiratory epithelium; those in an inferior position near the oral cavity are more likely to exhibit squamous epithelium.

The contents of the cyst wall can be a helpful diagnostic aid. Because the nasopalatine duct cyst arises within the incisive canal, moderate-sized nerves and small muscular arteries and veins usually are found in the wall of the cyst (Fig. 1-60). Small mucous glands have been reported in as many as one-third of cases. Occasionally, the clinician may see small islands of hyaline cartilage. Frequently, an inflammatory response is noted in the cyst wall and may range from mild to heavy. This inflammation usually is chronic in nature and is composed of lymphocytes, plasma cells, and histiocytes. Associated acute inflammatory cells (neutrophils) sometimes may be seen.

Treatment and Prognosis

Nasopalatine duct cysts are treated by surgical enucleation. Biopsy is recommended because the lesion is not diagnostic radiographically; other benign and malignant lesions have been known to mimic the nasopalatine duct cyst. The lesion is best approached with a palatal flap that is reflected after an incision is made along the lingual gingival margin of the anterior maxillary teeth. Recurrence is rare. Malignant transformation has been reported in a couple of cases, but this is an extremely rare complication.

◆ MEDIAN PALATAL (PALATINE) CYST

The median palatal cyst is a rare fissural cyst that theoretically develops from epithelium entrapped along the embryonic line of fusion of the lateral palatal shelves of the maxilla. This cyst may be difficult to distinguish from a nasopalatine duct cyst. In fact, most "median palatal cysts" may represent posteriorly positioned nasopalatine duct cysts. Because the nasopalatine ducts course posteriorly and superiorly as they extend from the incisive canal to the nasal cavity, a nasopalatine duct cyst that arises from posterior remnants of this duct near the nasal cavity might be mistaken for a median palatal cyst. On the other hand, if a true median palatal cyst were to develop toward the anterior portion of the hard palate, then it could easily be mistaken for a nasopalatine duct cyst.

Clinical and Radiographic Features

The median palatal cyst presents as a firm or fluctuant swelling of the midline of the hard palate posterior to the palatine papilla (Fig. 1-61). The lesion appears most frequently in young adults. Often it is asymptomatic, but some patients complain of pain or expansion. The average size of this cyst is 2 × 2 cm, but sometimes it can become quite large. Occlusal radiographs demonstrate a well-circumscribed radiolucency in the midline of the hard palate (Fig. 1-62). Occasional reported cases have been associated with divergence of the central incisors, although it may be difficult to rule out a nasopalatine duct cyst in these instances.

To differentiate the median palatal cyst from other cystic lesions of the maxilla, Gingell and associates suggested the following diagnostic criteria:
- Grossly appears symmetrical along the midline of the hard palate
- Located posterior to the palatine papilla
- Appears ovoid or circular radiographically
- Not intimately associated with a nonvital tooth
- Does not communicate with the incisive canal
- Shows no microscopic evidence of large neurovascular bundles, hyaline cartilage, or minor salivary glands in the cyst wall

It must be stressed that a true median palatal cyst should exhibit clinical enlargement of the palate. A midline radiolucency without clinical evidence of expansion is probably a nasopalatine duct cyst.

Histopathologic Features

Microscopic examination shows a cyst that usually is lined by stratified squamous epithelium. Areas of ciliated
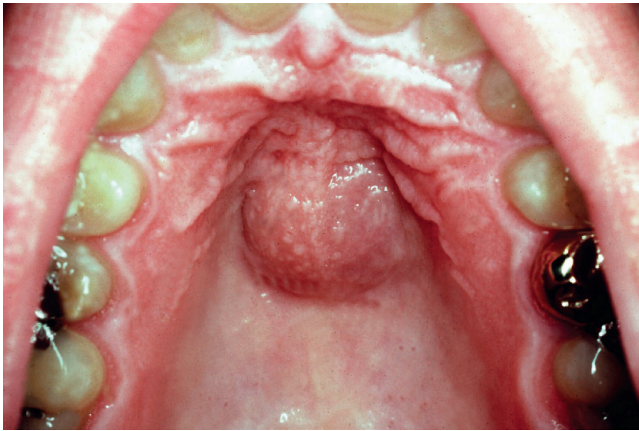
● **Fig. 1-61 Median Palatal Cyst.** Compressible mass in the midline of the hard palate posterior to the incisive papilla. (Courtesy of Dr. Craig Fowler.)

● **Fig. 1-62 Median Palatal Cyst.** Occlusal radiograph of same patient depicted in Figure 1-61. A well-circumscribed midline radiolucent defect can be seen, which is separate from the incisive canal. (Courtesy of Dr. Craig Fowler.)

pseudostratified columnar epithelium have been reported in some cases. Chronic inflammation may be present in the cyst wall.

Treatment and Prognosis

The median palatal cyst is treated by surgical removal. Recurrence should not be expected.

◆ "MEDIAN MANDIBULAR CYST"

The **"median mandibular cyst"** is a controversial lesion of questionable existence. Theoretically, it represents a fissural cyst in the anterior midline of the mandible that develops from epithelium entrapped during fusion of the halves of the mandible during embryonic life. However, the mandible actually develops as a single bilobed proliferation of mesenchyme with a central isthmus in the midline. As the mandible grows, this isthmus is eliminated. Therefore, because no fusion of epithelium-lined processes occurs, entrapment of epithelium should not be possible.

Because respiratory prosoplasia is not uncommon in odontogenic cysts, it appears likely that most (if not all) of these midline cysts are of odontogenic origin. Many purported cases would be classified today as examples of the *glandular odontogenic cyst* (see page 649), which has a propensity for occurrence in the midline mandibular region. Others could be classified as *periapical cysts, odontogenic keratocysts,* or *lateral periodontal cysts.* Because a fissural cyst in this region probably does not exist, the term *median mandibular cyst* should no longer be used.

◆ FOLLICULAR CYSTS OF THE SKIN

Follicular cysts of the skin are common keratin-filled lesions that arise from one or more portions of the hair follicle. The most common type, which is derived from the follicular infundibulum, is known as an **epidermoid** or **infundibular cyst.** These cysts often arise after localized inflammation of the hair follicle and probably represent a nonneoplastic proliferation of the infundibular epithelium resulting from the healing process. The term **sebaceous cyst** sometimes is used mistakenly as a synonym for both the epidermoid cyst and another cyst of the scalp known as a **pilar, trichilemmal,** or **isthmus-catagen cyst.** However, because both the epidermoid cyst and pilar cyst are derived from the hair follicle rather than the sebaceous gland, the term *sebaceous cyst* should be avoided.

Keratin-filled cysts of the skin may occasionally arise after traumatic implantation of epithelium, although such lesions may be difficult to distinguish from an infundibular cyst. Rarely, such **epidermal inclusion (implantation) cysts** also can develop in the mouth. These small inclusion cysts should be distinguished from oral epidermoid cysts that occur in the midline floor of mouth region and represent the minimal manifestation of the teratoma-dermoid cyst-epidermoid cyst spectrum (see page 31).

Clinical Features

Epidermoid (infundibular) cysts account for approximately 80% of follicular cysts of the skin and are most common in the acne-prone areas of the head, neck, and back. They are unusual before puberty unless they are associated with **Gardner syndrome** (see page 606). Young adults are more likely to have cysts on the face, whereas older adults are
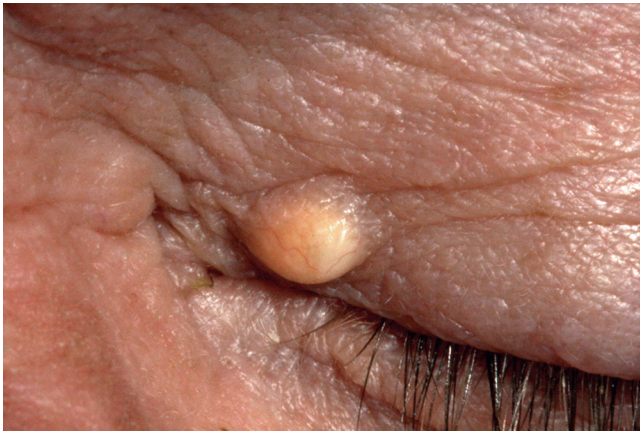
• **Fig. 1-63 Epidermoid Cyst.** Yellow nodule at the medial aspect of the eyelid.

• **Fig. 1-64 Pilar Cyst.** Nodular mass on the scalp.

more likely to have cysts on the back. Males are affected more frequently than females.

Epidermoid cysts present as nodular, fluctuant subcutaneous lesions that may or may not be associated with inflammation (Fig. 1-63). If a noninflamed lesion presents in an area of thin skin, such as the earlobe, then it may be white or yellow.

Pilar (trichilemmal) cysts comprise approximately 10% to 15% of skin cysts, occurring most frequently on the scalp (Fig. 1-64). They are twice as common in women as in men. The lesion is usually movable and shells out easily. **Milia** (singular: **milium**) are tiny keratin-filled cysts that resemble miniature epidermoid cysts (Fig. 1-65). A variety of such lesions have been described, including primary congenital milia, genodermatosis-associated milia, and milia that develop secondary to bullous disorders, trauma, or certain medications. Primary milia are thought to arise from the sebaceous collar of vellus hairs, whereas secondary milia may develop from eccrine ducts, hair follicles, or the overlying epidermis.

Histopathologic Features

Microscopic examination of an epidermoid cyst reveals a cavity that is lined by stratified squamous epithelium
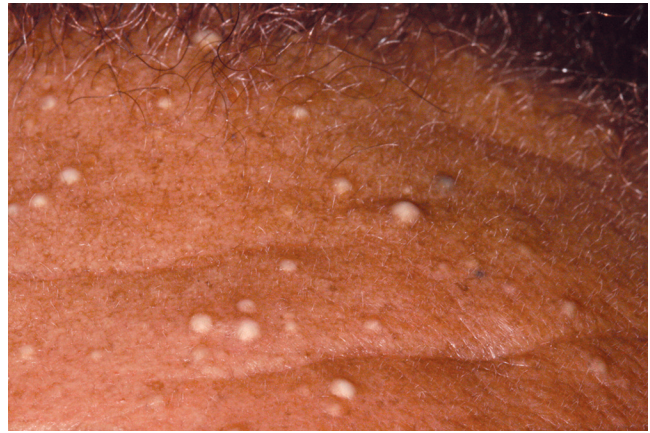
• **Fig. 1-65 Milia.** Multiple tiny keratin-filled cysts on the forehead.

resembling epidermis (Fig. 1-66). A well-developed granular cell layer is seen, and the lumen is filled with degenerating orthokeratin. Not infrequently, the epithelial lining will be disrupted. When this occurs, a prominent granulomatous inflammatory reaction, including multinucleated giant cells, can be present in the cyst wall because the exposed keratin is recognized as a foreign material.

The pilar cyst also is lined by stratified squamous epithelium, although a granular cell layer usually is absent or greatly diminished (Fig. 1-67). The keratinocytes remain large in the upper epithelial layers with an abrupt transition to dense, compact keratin that fills the cyst lumen.

Treatment and Prognosis

Epidermoid and pilar cysts usually are treated by conservative surgical excision, and recurrence is uncommon. Malignant transformation has been reported but is exceedingly rare.

An individual milium can be removed by evacuation, in which the lesion is nicked by a scalpel blade, followed by application of pressure with a comedone extractor or curette. Multiple milia can be managed via electrocautery or application of topical retinoids.

◆ DERMOID CYST (DYSONTOGENIC CYST)

The **dermoid cyst** is an uncommon developmental cystic malformation. The cyst is lined by epidermis-like epithelium and contains dermal adnexal structures in the cyst wall. It is generally classified as a benign cystic form of **teratoma.**

By definition, a teratoma is a developmental tumor composed of tissue from more than one germ layer and sometimes all three: 1) ectoderm, 2) mesoderm, and 3) endoderm. Such tumors are believed to arise from germ cells or entrapped totipotent blastomeres, which can produce derivatives of all three germ layers.

Teratomatous malformations have a spectrum of complexity. In their most complex form, these lesions produce multiple types of tissue that are arranged in a disorganized
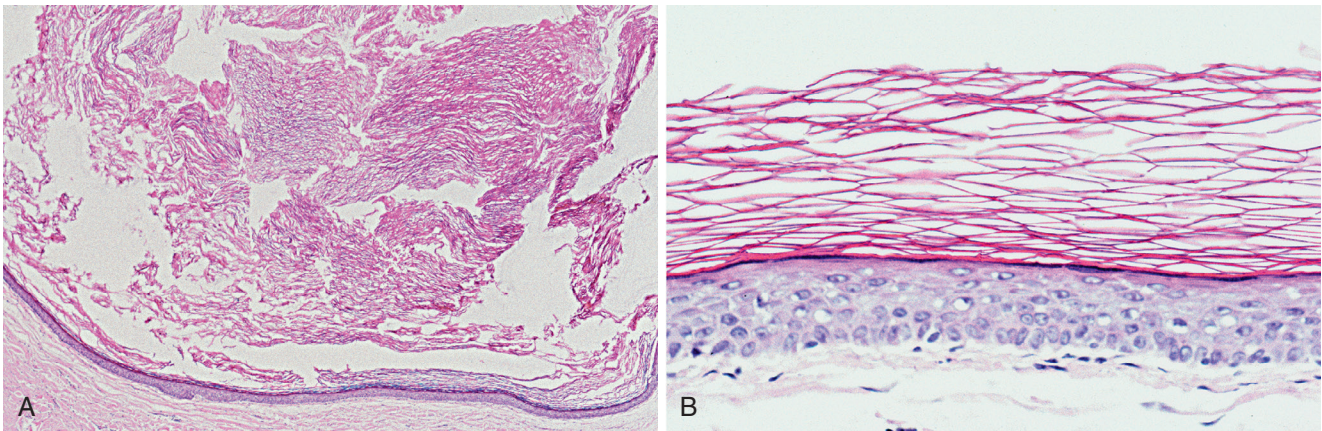
• **Fig. 1-66 Epidermoid Cyst. A,** Low-power view showing a keratin-filled cystic cavity. **B,** High-power view showing stratified squamous epithelial lining with orthokeratin production.
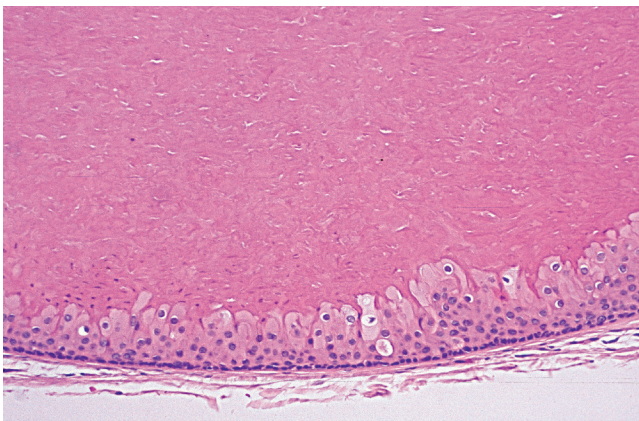
• **Fig. 1-67 Pilar Cyst.** Medium-power view showing an abrupt transition between the stratified squamous epithelial lining and compact keratin without the presence of a transitional granular cell layer.

• **Fig. 1-68 Dermoid Cyst.** Fluctuant midline swelling in the floor of the mouth. (From Budnick SD: *Handbook of pediatric oral pathology,* Chicago, 1981, Year Book Medical.)

fashion. These "complex" teratomas are most common in the ovaries or testes and can be benign or malignant. Occasionally, ovarian teratomas (or "dermoids") produce well-formed teeth, or even partially complete jaws. Complex teratomas of the oral cavity are rare and usually are congenital in nature. When they occur, they usually extend through a cleft palate from the pituitary area via Rathke's pouch. Cervical teratomas also have been reported.

The term **teratoid cyst** has been used to describe a cystic form of teratoma that contains a variety of germ layer derivatives:
1. Skin appendages, including hair follicles, sebaceous glands, and sweat glands
2. Connective tissue elements, such as muscle, blood vessels, and bone
3. Endodermal structures, such as gastrointestinal lining

Rarely, oral cysts may be lined entirely by gastrointestinal epithelium. These **heterotopic oral gastrointestinal cysts (enterocystomas, enteric duplication cysts)** usually are considered to be choristomas, or histologically normal tissue found in an abnormal location. However, these lesions probably can be included under the broad umbrella of

teratomatous lesions, especially because they occasionally are found in combination with dermoid cysts.

Dermoid cysts are simpler in structure than complex teratomas or teratoid cysts. Although they do not contain tissue from all three germ layers, they probably represent a *forme fruste* of a teratoma. Similar cysts of the oral cavity can be seen that are lined by epidermis-like epithelium, but they contain no dermal appendages in the cyst wall. These lesions have been called **epidermoid cysts** and represent the simplest expression of the teratoma spectrum. These intraoral epidermoid cysts should not be confused with the more common **epidermoid cyst of the skin** (see page 29), a non-teratomatous lesion that arises from the hair follicle. Because the teratoid cyst/dermoid cyst/epidermoid cyst spectrum represents defective embryologic development, these cysts sometimes are known collectively as **dysontogenic cysts**.

Clinical and Radiographic Features

Dermoid cysts most commonly occur in the midline of the floor of the mouth (Fig. 1-68), although occasionally they are displaced laterally or develop in other locations. If the
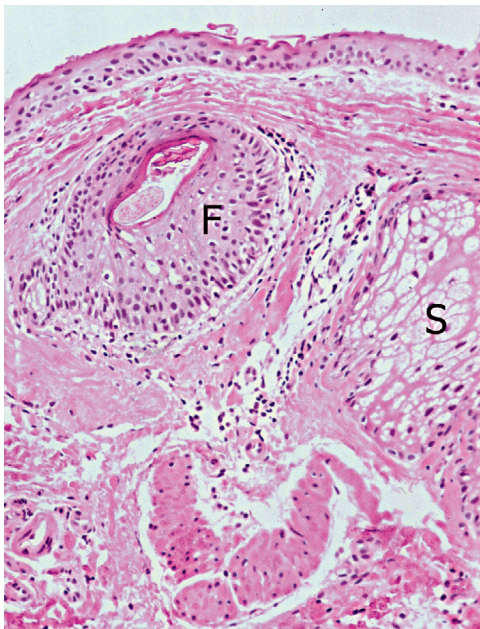
• **Fig. 1-69 Dermoid Cyst.** Squamous epithelial lining *(top),* with hair follicle *(F),* sebaceous glands *(S)* in the cyst wall.

cyst develops above the geniohyoid muscle, then a sublingual swelling may displace the tongue toward the roof of the mouth and create difficulty in eating, speaking, or even breathing. Cysts that occur below the geniohyoid muscle often produce a submental swelling, with a "double-chin" appearance.

Oral dermoid cysts can vary in size from a few millimeters to 12 cm in diameter. They are most common in children and young adults; 15% of reported cases have been congenital. The lesion is usually slow growing and painless, presenting as a doughy or rubbery mass that frequently retains pitting after application of pressure. Secondary infection can occur, and the lesion may drain intraorally or onto the skin. MRIs, CT scans, or contrast medium radiographs may be helpful in delineating the extent of the lesion.

Histopathologic Features

Dermoid cysts are lined by orthokeratinized stratified squamous epithelium with a prominent granular cell layer. Abundant keratin often is found within the cyst lumen. On rare occasions, areas of respiratory epithelium can be seen. The cyst wall is composed of fibrous connective tissue that contains one or more skin appendages, such as sebaceous glands, hair follicles, or sweat glands (Fig. 1-69).

Treatment and Prognosis

Dermoid cysts are treated by surgical removal. Those located above the geniohyoid muscle can be removed by an intraoral incision, and those below the geniohyoid muscle may require an extraoral approach. Recurrence is uncommon.

Malignant transformation into squamous cell carcinoma has been reported only rarely.

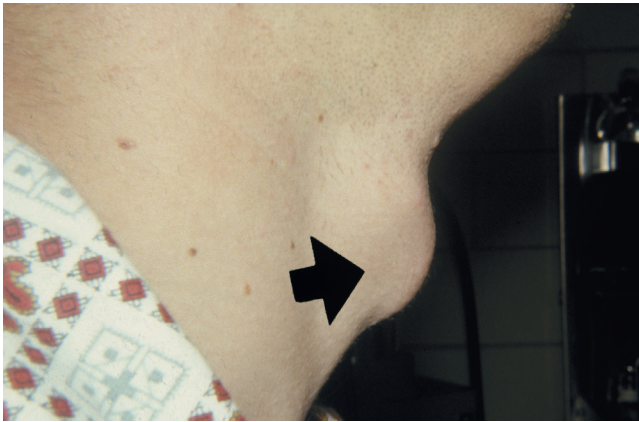## ◆ THYROGLOSSAL DUCT CYST (THYROGLOSSAL TRACT CYST)

The thyroid gland begins its development during the third to fourth week of embryonic life as a proliferation of endodermal cells from the ventral floor of the pharynx, between the tuberculum impar and copula of the developing tongue—a point that later becomes the foramen cecum. This thyroid anlage descends into the neck as a bilobed diverticulum anterior to the developing hyoid bone and reaches its definitive level below the thyroid cartilage by the seventh embryonic week. Along this path of descent, an epithelial tract or duct is formed, maintaining an attachment to the base of the tongue. This thyroglossal duct becomes intimately associated with the developing hyoid bone. As the hyoid matures and rotates to its adult position, the thyroglossal duct passes in front and beneath the hyoid, looping upward and behind it before curving downward again into the lower neck. The caudal segment of this duct often persists, forming the pyramidal lobe of the thyroid gland.

The thyroglossal duct epithelium normally undergoes atrophy and is obliterated, although autopsy studies have shown that as many as 7% of the population will have *thyroglossal tract remnants.* These epithelial remnants usually are asymptomatic, although some can give rise to cysts along this tract known as **thyroglossal duct cysts.** The impetus for cystic degeneration is uncertain. Inflammation is the most frequently suggested stimulus, especially from adjacent lymphoid tissue that may react to draining infections of the head and neck. Retention of secretions within the duct is another possible factor. In addition, there are several reports of familial occurrence of such cysts.

Clinical Features

Thyroglossal duct cysts classically develop in the midline and may occur anywhere from the foramen cecum area of the tongue to the suprasternal notch. In 60% to 80% of cases, the cyst develops adjacent to the hyoid bone. Suprahyoid cysts may be submental in location. Cysts that develop in the area of the thyroid cartilage often are deflected lateral to the midline because of the sharp anterior margin of the thyroid cartilage. Intralingual cysts are rare.

Thyroglossal duct cysts may develop at any age, but they are most commonly diagnosed in the first two decades of life; about 50% of cases occur before the age of 20. There is no sex predilection. The cyst usually presents as a painless, fluctuant, movable swelling unless it is complicated by secondary infection (Fig. 1-70). Lesions that develop at the base of the tongue may cause laryngeal obstruction. Most thyroglossal duct cysts are smaller than 3 cm in diameter, but occasional cysts may reach 10 cm in size. If the cyst

• **Fig. 1-70 Thyroglossal Duct Cyst.** Swelling *(arrow)* of the anterior midline of the neck. (Courtesy of Dr. Philip Sprinkle.)
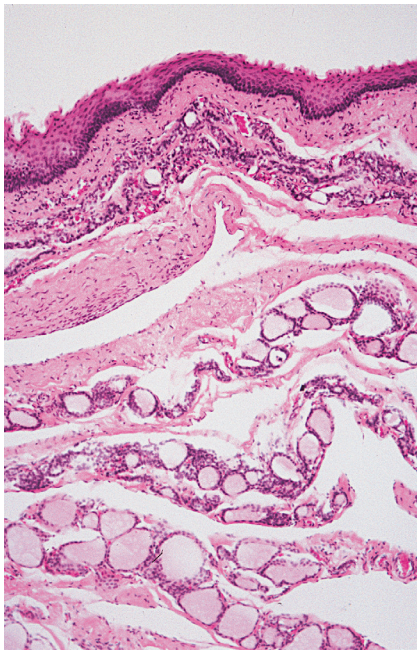
• **Fig. 1-71 Thyroglossal Duct Cyst.** Cyst *(top)* lined by stratified squamous epithelium. Thyroid follicles can be seen in the cyst wall *(bottom).*

maintains an attachment to the hyoid bone or tongue, it will move vertically during swallowing or protrusion of the tongue. Sinus tracts to the skin or mucosa develop in as many as one-third of cases, usually from rupture of an infected cyst or as a sequela of surgery.

Histopathologic Features

Thyroglossal duct cysts usually are lined by columnar or stratified squamous epithelium, although occasionally, cuboidal or even small intestine epithelium may be documented (Fig. 1-71). Sometimes a mixture of epithelial types is present, or an intact epithelial lining cannot be found due to secondary inflammation. Thyroid tissue may occur in the cyst wall, but this is not a constant finding.

Treatment and Prognosis

Thyroglossal duct cysts are best treated by a Sistrunk procedure. In this operation the cyst is removed in addition to the midline segment of the hyoid bone and a generous portion of muscular tissue along the entire thyroglossal tract. The recurrence rate associated with this procedure is less than 10%. A much higher recurrence rate can be expected with less aggressive surgery.

Carcinoma arising in a thyroglossal duct cyst is a rare complication that occurs in approximately 1% to 2% of cases. Most of these have been papillary thyroid adenocarcinomas. Fortunately, metastases from thyroglossal carcinoma are rare, and the prognosis for people with these tumors is good.

◆ BRANCHIAL CLEFT CYST (CERVICAL LYMPHOEPITHELIAL CYST)

The **branchial cleft cyst** is a developmental cyst that is derived from remnants of the branchial arches. During the fourth week of gestation, the head and neck region of the embryo develops paired branchial arches, which are covered by ectoderm on the external surface and endoderm on the internal surface. The outer arch surfaces are separated by clefts and the inner surfaces are divided by pouches. In fish and amphibians, the branchial arches are destined to become the gill apparatus; in humans, the clefts and pouches gradually are eliminated during embryonic life by ingrowth of mesenchyme. However, incomplete obliteration of these pharyngeal clefts and pouches may give rise to branchial cleft anomalies, such as cysts, fistulae, or sinus tracts. About 95% of these anomalies are believed to arise from the second branchial arch, with the remaining 5% originating from the first, third, and fourth branchial arches.

Clinical Features

Branchial cleft cysts from the second arch occur in the upper lateral neck anterior or deep to the sternocleidomastoid muscle (Figs. 1-72 and 1-73). They most frequently develop in children and young adults between the ages of 10 and 40. Clinically, the cyst appears as a soft, fluctuant mass that can range from 1 to 10 cm in diameter. Associated tenderness or pain sometimes may occur with secondary infection. Occasionally, the lesion becomes evident after an upper respiratory tract infection or trauma. Some branchial cleft anomalies appear as sinuses or fistulae that may produce a mucoid discharge onto the skin. In rare instances, bilateral cysts may develop.

Anomalies from the first branchial arch comprise approximately 1% of branchial cleft malformations and usually are found in close proximity to the parotid gland. Third-cleft and fourth-cleft anomalies are rare and may develop in the lower neck or mediastinum.