INTRODUCTION

Sensory development is complex, with both morphologic and neural components. The senses begin to develop well before birth based on in-utero stimuli. They all mature rapidly in the first year of life. This article focuses on the cranial senses of vision, hearing, smell, and taste. Tactile development and pain perception are not addressed. Sensory function, embryogenesis, external and genetic effects, and common malformations that may affect development are discussed, along with the corresponding sensory organ examination and evaluation.

VISION

Eye Development

The eye is derived from an outgrowth of neuroectoderm of the forebrain. By the 32nd embryonic day, a distinct optic cup with a ventral groove is detectable. The optic cup further invaginates to form the globe with anterior and posterior chambers.
Surface ectoderm is pulled in to form the lens, iris, and other associated structures to separate the 2 chambers. The cornea is formed from surface ectoderm and a fine layer of mesoderm between the neuroectoderm and surface ectoderm. The eyelids and lacrimal glands are formed from surface ectoderm. The retina forms from the internal walls of the optic cup. A thick neuroepithelium differentiates into rods and cones. Myelination is incomplete before birth at term but, after light exposure for approximately 10 weeks, myelination is complete. This process is markedly delayed in babies born prematurely and may be disrupted significantly in retinopathy of prematurity.\(^3\)

**Examination of the Eye**

The eyelids meet and adhere by the tenth week of gestation.\(^2\),\(^4\) They remain adherent until approximately 26 weeks’ gestation. Although uncommon, babies born vaginally with a face presentation may have everted eyelids, which readily reduce with few complications and normal eyes otherwise (Fig. 1). An eyelid coloboma (notched lid) is a rare defect limited to the upper eyelid that requires surgery to protect the cornea and conjunctiva.

Conjunctival hemorrhage, often associated with a difficult delivery, is absorbed within several weeks. The sclera may be discolored yellow with significant jaundice and may appear bluish in inherited collagen vascular diseases because of scleral thinning and visualization of the underlying retina. Newborn eye prophylaxis to prevent bacterial infection often produces a transient chemical conjunctivitis. Conjunctival discharge may be caused by an infection, with gonorrhea and chlamydia being the most serious infections (Fig. 2). Obstruction of the nasolacrimal duct results in excessive tearing. Cloudy or protruding cornea indicates glaucoma (Fig. 3). The increased pressure of the aqueous humor in the anterior chamber is an emergency requiring immediate consultation and intervention by a pediatric ophthalmologist.

The iris color at birth is bluish in most infants. Pigmentation often progresses to a darker color, with the final iris color achieved by 4 months. Lack of pigmentation with a pink iris is a primary feature of albinism. Aniridia, complete lack of irises, is caused by an arrest of development of the rim of the optic cup at the eighth week. A failure of the ventral groove to fuse in early development leads to an iris coloboma, seen as a keyhole defect of the iris, which may extend into the ventral retina. The ciliary body is similarly affected, resulting in the inability to constrict the pupil and subsequent photophobia.

**Fig. 1.** Everted eyelids.
The classic newborn eye test is the red reflex, elicited by shining a light into the eye, and the reflecting light off the highly vascular retina appears red. Any color but red may indicate anterior chamber disease (glaucoma), cataract, or retinal disorder such as detached retina or retinoblastoma (Fig. 4). Premature babies with immature retinas at birth may develop retinal scarring and detachment, a condition termed retinopathy of prematurity, which can also cause abnormal red reflex.

Examination of the extraocular muscles is difficult at birth. It is common for newborns to have discordant muscle movement because their ability to focus and the resultant conjugate gaze take several months to mature. In addition, unusually long eyelashes can be an indication of a genetic syndrome, the foremost being Cornelia de Lange syndrome.

**Early Vision**

Neonatal vision is limited, such that term infants can only focus approximately 25 cm (10 inches) shortly after birth. At less than 34 weeks’ gestation, neonates do not have sufficient cone development to see color and can discriminate between dark and light only at a limited distance. The initial color humans see is red, presumably because of low light exposure from transillumination of the red color of maternal oxygenated hemoglobin into the uterus. With continued exposure to various

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**Fig. 2.** Gonococcal purulent conjunctivitis.

**Fig. 3.** Glaucoma.
wavelengths of light, the retinal cones of other colors develop. The progression of eye function development is summarized in Table 1.

HEARING

Normal Development of Hearing

The most important aspects of the auditory system development take place during the second half of gestation. Babies born prematurely and exposed during this period to multiple potential adverse effects of life-sustaining therapies are at great risk for hearing deficits and secondarily speech delay. Among neonatal intensive care unit (NICU) graduates, the incidence of hearing impairment is estimated to be at least 10-fold greater than in their term counterparts.

Structure and Function

The auditory system comprises 3 related sets of structures: the peripheral components, including the outer, middle, and inner ears; the auditory nerves (cranial nerve VIII); and the auditory regions of the brain located primarily in the brainstem and left temporal lobe.

The outer ear of neonates features a narrow canal with thin cartilage, which is readily blocked and compressed. The shape, position, and peripheral tissue of the ear may provide clues to dysfunctional development. The classic low-set or malformed ear

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Gestational Age (wk)</th>
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<tbody>
<tr>
<td>Blink/squint in response to a bright light</td>
<td>26</td>
</tr>
<tr>
<td>Pupils constrict to light</td>
<td>30</td>
</tr>
<tr>
<td>Ability to fixate vision on a large object in close proximity</td>
<td>32</td>
</tr>
<tr>
<td>Track large moving objects</td>
<td>34</td>
</tr>
<tr>
<td>Color perception; red at first</td>
<td>34</td>
</tr>
</tbody>
</table>

is found in more than 120 well-characterized syndromes.\textsuperscript{11} For the ear to be considered low set, the entire ear must be below an extended line drawn from the inner canthus of the eye to the outer canthus (Fig. 5). A second criterion for low-set ear is the ear canal below an imaginary line drawn from the outer canthus to the base of the occiput. Posteriorly rotated ears or preauricular skin tags are more commonly seen in babies with syndromes.

The fluid-filled middle ear reaches adult size by 20 weeks’ gestation, but the middle ear ossicles remain cartilaginous until 32 weeks’ gestation. Cochlear structures, including inner and outer hair cells, are fully developed by 25 weeks’ gestation. This process extends to myelination developing from the brainstem to higher level auditory pathways. The cochlea transduces acoustic wave energy into electrical impulses, which occurs in the inner hair cells. Outer hair cells adjust reflexively to sound input by producing frequency-specific echo sounds called otoacoustic emissions (OAE).

Hearing is the first sense exposed to stimulation that promotes development of the neural pathways. Functional hearing in human fetuses develops at 25 to 27 weeks’ gestation. Low-frequency sounds, such as the mother’s heartbeat and speech, elicit physiologic responses that are consistently detectable. Maturing fetuses respond to a wider range of sound frequencies progressing through the third trimester and shortly after birth. The functional maturation of hearing in the newborn is caused by structural changes in the outer and middle ears. Progressive myelination of auditory axons results in a maturing brainstem evoked response (auditory brainstem response [ABR]) test because of increased conduction velocities and wave amplitudes.

The incidence of permanent hearing loss in neonates ranges between 1.4 and 3 per 1000 births in the United States.\textsuperscript{12–14} With progressive or new-onset hearing loss, the prevalence of permanent sensorineural hearing loss increases during childhood to estimated rates of about 2.7 per 1000 in 4-year-old children and 3.5 per 1000 in adolescents.\textsuperscript{10,14}

**Types and Causes of Hearing Impairment**

**Causes of hearing impairment**

Based on the anatomic location of the hearing dysfunction, hearing loss can be classified as conductive, sensorineural, or neural\textsuperscript{15–19}:

- Conductive hearing loss: blockage of sound transmission in the outer or middle ear caused by permanent conditions like anatomic malformations or transient problems such as fluid or debris.
- Sensorineural hearing loss: failure of sound transduction in the inner and outer hair cells of the cochlea, and of transmission through the auditory nerve.

- Neural hearing loss, also known as auditory neuropathy: dysfunction of the inner hair cells and auditory nerve, but OAE from the outer hair cells remain intact.

- Mixed hearing loss: combination of conductive and sensorineural hearing deficits.

In addition to the neurophysiologic classification described earlier, hearing loss can be further categorized according to severity (ie, mild, moderate, severe, or profound), based on the sound pressure level of the individual’s hearing threshold. In addition, hearing loss can be unilateral or bilateral.\textsuperscript{18,20}

About two-thirds of congenital hearing loss has an underlying genetic cause.\textsuperscript{10,14} Mutations in the connexin 26 gene (\textit{GJB2}), predominantly the 35delG point mutation, account for 20\% of congenital deafness. An additional 44\% of congenital deafness has other genetic causes; one-third of these being related to recognizable syndromes and two-thirds being nonsyndromic. Most nonsyndromic hearing loss cases follow an autosomal recessive inheritance pattern (\textit{DFNB}), whereas a minority are autosomal dominant (\textit{DFNA}); X-linked and mitochondrial inheritance is rare.

Although many gene mutations have been associated with hearing loss, about 95\% of congenitally deaf infants are born to parents with normal hearing, so a negative family history of deafness does not exclude the possibility of hereditary hearing loss. A newly diagnosed infant may serve as the index case to prompt genetic evaluation for the family.

The underlying pathophysiology of hearing dysfunction is complex. The 2007 Position Statement of the American Academy of Pediatrics Joint Committee on Infant Hearing\textsuperscript{21} outlined causes of hearing loss that can be congenital, delayed onset, and/or progressive. These categories include:

- Infections
  - Fetal: cytomegalovirus (CMV), varicella, syphilis, rubella, toxoplasmosis, and others
  - Postnatal infections: meningitis, otitis media, encephalitis

- Environmental and therapeutic toxicity:
  - Perinatal asphyxia, anoxia
  - Ototoxic medications (aminoglycosides, loop diuretics)
  - Mechanical ventilation, extracorporeal membrane oxygenation, sustained metabolic or respiratory acidosis
  - Severe hyperbilirubinemia requiring exchange transfusion

- Trauma: perinatal, child abuse, temporal bone fracture

- Familial hearing loss

- Craniofacial anomalies/syndromes:
  - Malformations of craniofacial structures derived from the first and second branchial arches, even without genetic associations, are embryologically related to the development of the inner ear, and are thus a risk factor for hearing loss. The many syndromes with craniofacial involvement include Waardenburg type I and II (white forelock), neurofibromatosis, and Alport.

Familial syndromes associated with progressive hearing loss include:

- Pendred syndrome accounts for only 3\% of deafness diagnosed from birth, but comprises 12\% of the cases of deafness in the preschool population.\textsuperscript{10,11} Although deafness occurs early, the other clinically obvious component of the syndrome is goiter, which does not present until late childhood.
Mitochondrial mutations or other neurodegenerative disorders, such as Friedrich ataxia, may first manifest beyond early infancy.

Usher syndrome is a familial disorder characterized by progressive hearing loss and progressive retinitis pigmentosa leading to blindness.

Jervell and Lange-Nielsen syndrome presents with cardiac dysrhythmias caused by a prolonged QT interval and should prompt reevaluation of hearing function.

Even if the newborn hearing screen is normal, a family history of hearing loss should trigger continued monitoring of the infant and formal audiological assessment should be repeated by 24 to 30 months of age.

Medical screening systems cannot detect hearing loss in many children in a timely manner. Concerns by the family members regarding hearing, speech, language, or developmental delay must be addressed to improve early detection of hearing loss. Despite early hearing screening, more than two-thirds of children with subsequent hearing loss were diagnosed following parental concern and school hearing screens.15,22,23

Conditions with combined auditory and visual impairment, such as Usher syndrome, are particularly devastating to development of communication and psychosocial function. Increased association exists with autism spectrum disorder, which occurs in about 7% of 8-year-old children with visual or auditory impairment. The clinical diagnosis of hearing or visual deficits resulted in a substantial delay in the diagnosis of the coexisting autism spectrum disorder.16

Functional Consequences

Congenital or neonatally acquired permanent hearing loss adversely affects expressive and receptive language development, resulting in diminished academic achievement and social development. These sequelae can be mitigated by diagnosis and appropriate therapeutic intervention within the first 6 months of life.10,18 Therefore, the age of 6 months represents a critical target for initial interventions in infants with hearing loss to optimize functional outcomes.19

The functional consequences of hearing loss depend on the age of onset and the specific subcategory of the hearing loss described earlier. Although bilateral deafness is most incapacitating, even unilateral hearing loss may affect language and educational performance.20 Minimal information is available regarding the persistent effects of milder transient or reversible hearing dysfunction, such as that related to external ear debris in newborns, persistent otitis media with effusion, or auditory neuropathy in severe hyperbilirubinemia.

Screening in Newborns and Young Children

Intervention for hearing loss is most effective when initiated early to salvage speech and language development. Because of vigorous advocacy by the American Academy of Pediatrics, newborn hearing screening is performed in most individual birthing hospitals in the United States. There is great variability in accuracy because of multiple testers. Table 2 summarizes the most useful hearing tests.

In newborn hearing screening, neither ABR nor OAE require an active response from the infant. The tests are more accurate when they are performed on sleeping infants in a quiet environment. Both tests are cost-effective for early universal screening, given the high incidence and consequences of neonatal hearing loss.22,23

Automated auditory brainstem response screening uses scalp electrodes to detect the eight cranial nerve and auditory brainstem pathway responses to sound stimuli, applying automated algorithms to define hearing thresholds. ABR screening
is sensitive to abnormalities in conductive, sensorineural, and purely neural hearing losses.

OAE tests detect the reflected echoes produced by the cochlear outer hair cells when stimulated by clicks across specified frequencies. These tests are abnormal in conductive and sensorineural hearing loss. However, they may be normal (ie, false-negative) in cases of purely auditory neuropathy. Because many of these cases are most likely to be found in the NICU setting, it is recommended that only ABR screening be used in NICUs. Because OAE testing is easier and less expensive to perform, it is often used to screen healthy newborns. Some hospitals use a 2-stage protocol with ABR screening following OAE test failures to minimize the rate of false-positive screens.

The Joint Committee on Infant Hearing promotes the 1-3-6 principle of screening, diagnosis, and therapy for neonatal hearing loss, recommending that all infants undergo hearing screening by 1 month of age and those who fail their screens should have a diagnostic evaluation by an audiologist no later than 3 months of age. Infants whose hearing loss is confirmed should receive therapy appropriate to their diagnosis by age 6 months. Neonatal hearing screening program failure rates range from 0.5% to 4%. Although false-negative neonatal hearing screens are exceedingly uncommon, such tests cannot detect progressive or later-onset hearing loss in childhood, which is as common as neonatal hearing loss.

The primary obstacle to neonatal hearing screening effectiveness in the United States is the 46% loss to follow-up of infants who fail their neonatal screen. Primary care providers in the medical home have a critical role in ensuring that these infants receive the appropriate diagnostic testing in a timely manner. Major barriers to follow-up include ineffective communication of screening results, inadequate education of families regarding the importance of screening failures (which may have been deemed referrals), and lack of access to hearing diagnostic services for infants.

Early hearing detection and intervention (EHDI) programs, promoted and driven by professional organizations such as the American Academy of Pediatrics and supported by state and federal health agencies, have been implemented by many state health departments in association with early intervention programs. EHDI programs serve as coordinating centers for gathering data (shared with the Centers for Disease Control and Prevention), communicating with major stakeholders, and providing resources to maximize the success of the screening programs. A major focus of EHDI programs presently is the minimization of losses to follow-up in screening programs. Follow-up could be improved using currently available technological solutions, including documentation of hearing screening results in electronic medical records,

### Table 2
**Neonatal hearing tests**

<table>
<thead>
<tr>
<th>Test</th>
<th>Loss Detected</th>
<th>Significance</th>
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<tbody>
<tr>
<td>Tympanometry</td>
<td>Conductive only</td>
<td>By changing pressure in the external ear canal, evaluates the intactness of the tympanic membrane and mobility with middle ear ossicles</td>
</tr>
<tr>
<td>Acoustic reflex</td>
<td>Conductive Sensorineural Neural</td>
<td>Measures stiffness of the middle ear Loud sounds trigger contraction of the tensor tympani and stapedius</td>
</tr>
<tr>
<td>ABR</td>
<td>Conductive Sensorineural Neural</td>
<td>Sound impulses of selected wavelengths generate electrical impulses that travel the auditory pathways to the brainstem and are reflected to the sensor</td>
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Clark-Gambelunghe & Clark
and automated communication of test results from hospitals to the medical homes and EHDI programs (preferably coordinated with other neonatal screening test results); also, teleaudiology could expand the availability of diagnostic testing for infants and their families.

The Joint Committee on Infant Hearing of the American Academy of Pediatrics has emphasized the need for a risk factor–based rescreening, even if the infants at risk passed the universal newborn screen. Although the timing and number of hearing reevaluations should be individualized, infants with a risk factor should have at least 1 postneonatal diagnostic audiologic evaluation by 24 to 30 months of age. Syndromic children or those infected with CMV may need more frequent reevaluation. Diagnostic services should be provided by audiologists with expertise and equipment appropriate for evaluating infants. When permanent hearing loss is confirmed, the primary care provider should coordinate further diagnostic evaluation, to define the cause of the hearing loss and possible comorbidities. This process should include consultation with genetics, an otolaryngologist experienced in pediatric hearing loss, and an ophthalmologist with expertise in evaluating infants.

Medical and Educational Interventions

Following diagnostic testing, the primary care provider needs to coordinate referrals for appropriate medical and surgical therapies, as well as community-based interventions. Input from multiple professionals, including audiology, speech-language pathology, and otolaryngology (ear, nose, and throat), as well as awareness of local community and school-based early intervention program resources, is needed to help families choose communication goals and interventions required to achieve them. Development of an individual family service plan is a first step in ensuring that the infant receives appropriate services no later than 6 months of age. In addition to preventing loss to follow-up before diagnosis, the medical home must also promote timely therapeutic follow-up, given that only 39% of infants with hearing loss were fitted with hearing aids by 6 months. It is important to establish appropriate initial therapy during the sensitive period for hearing development, to take advantage of plasticity of the auditory cortex, and to optimize cross-modal (eg, auditory and verbal) input of language acquisition. Aside from the coordinating functions, the medical home must provide continued surveillance for common conditions such as otitis media with effusion, which may affect hearing acuity and necessitate unplanned audiologic reevaluation, adjustments to existing amplification, or tympanostomy tubes. Close monitoring of developmental milestones is also essential.

Medical and Surgical Interventions

Ganciclovir
Detection of a CMV infection early allows the use of this antiviral medication to limit potential damage caused by the progressive CMV infection.

Surgical Implants

Bone-conducting miniature implants
SoundBite is a bone-conducting hearing aid inserted into the mouth. BAHA Attract is a magnet-based bone-conduction device inserted behind the ear.

Cochlear Implant

Electric and acoustic stimulation
Cochlear implants transduce sound waves into frequency-specific electrical signals that are delivered to residual functioning auditory nerve fibers. They can be placed
in children as young as 1 year of age with profound hearing loss and after 18 months of age in children with severe to profound bilateral sensorineural hearing loss, when amplification alone is inadequate. The device produces the greatest benefits in speech development when inserted by 7 years of age. Hearing loss caused by auditory neuropathy does not respond well to amplification alone, so early cochlear implantation may be advantageous in these situations.24,25 Patients with cochlear implants are at increased risk for bacterial meningitis, particularly with Streptococcus pneumoniae, and should be immunized according to a high-risk schedule and monitored for early signs of meningitis associated with otitis media or other infections.

**Modes of Communication**

The family’s choice of mode of communication may change over time, depending on the child’s functional hearing, development, available interventions, and social environment factors.26–30 Five options are currently available. The goal for the first 3 communication modes is spoken language, and the remaining 2 use sign language, with or without speech:

- Auditory verbal communication uses only optimized listening skills.
- Auditory-oral communication uses residual hearing with amplification, supported visually by speech reading.
- Cued speech combines listening with visual cues from 8 hand shapes near the face.
- American Sign Language (AMSLAN) can be learned by deaf children, with English or any other language as a second language.
- Total communication combines all modes of communication toward simultaneous use of speech and sign language.

**Continued Interventions: Family and School**

Parents and educators (including an expert in education of students with hearing loss) must develop an individualized education plan or section 504 plan to optimize student achievement, with support from the primary care medical home.28,29 Adaptations in the learning environment may involve the student, teachers, modes of communication, physical design of the classroom, and curricular modifications, including supplemental instruction. Examples of such adaptations include an optimal amplification system, visual assistive devices (eg, telecommunication device for the deaf), optimal seating arrangements, individualized communication with the student, use of an interpreter, assistance with asynchronous learning (eg, new vocabulary provided in advance of the session or a buddy system for note taking), and alternative testing methods. In addition, there are various options for supplemental instruction, including sign language and support from a deaf or hard-of-hearing role model. Continuous evaluation is essential for optimal adjustment and coordination of the educational accommodations over time.

Because hearing disorders and related comorbidities have varied causes, the outcome of an individual child is difficult to predict. Children with isolated auditory neuropathy treated by cochlear implantation performed comparably with age-matched peers with sensorineural hearing loss, but those with auditory neuropathy associated with a cognitive or developmental disorder had significantly less benefit and continued to rely on nonauditory modes of communication.25

**SMELL, TASTE, ORAL STRUCTURES, AND FUNCTION**

The ability of a baby to smell is an important component of the early infant-mother interaction. Components of the maternal diet reach the amniotic fluid, are swallowed,
and become familiar to the fetus. They may contribute to the scent of the mother, including her breast milk. By 5 to 6 days of life, babies preferentially choose the breast pad of their mother rather than that from another mother or an unused pad. Although the progressive development of smell is less well defined compared with vision and hearing, a few general observations have been made. Term babies prefer sweet odors such as lavender and vanilla and have a rapid avoidance response to foul odors like rotten eggs. Babies with choanal atresia or a tracheostomy have blunted development of smell, presumably caused by minimal airflow through the nose.

Taste development is likewise poorly understood compared with the other senses. Taste is supplied by the chorda tympani branch of the facial nerve (cranial nerve VII) on the anterior two-thirds of the tongue and by a branch of the glossopharyngeal (cranial nerve IX) over the posterior one-third of the tongue. Fetuses in the uterus continually swallow the components of amniotic fluid with proteins, carbohydrate, fat, and small molecules to initiate digestive enzymatic activity, so taste likely begins to develop in utero. Human neonates prefer sweet foods and can detect sour and bitter. Babies may not be able to detect salt until 3 to 4 months of age. They prefer breast milk to infant formula because the bovine alpha casein protein is more bitter than human beta casein. Hydrolyzed protein formulas are less savory. Sucrose in soy formulas is sweeter than lactose, thus soy formula should not be used as an adjunct to breast feeding.

The structures of the oral cavity derive from the first branchial arch. By the end of the fourth week of development, the frontonasal, 2 maxillary, and 2 mandibular processes are discernible. These tissues eventually fuse midline to form the face and palate at between 6 and 12 weeks’ gestation. Remnants of, and failure of, fusion can readily be detected on examination of newborns.

An appropriate neonatal oral examination includes both inspection and palpation. The provider should visually inspect the jaw and mouth size and shape, lips, gingiva, dentoalveolar ridge, palate, and mouth and tongue appearance and mobility. A gloved finger should be used to evaluate the sucking reflex and to palpate the hard and soft palates for a defect.

Drooping of the corner of the mouth at rest can result from facial nerve paralysis (Fig. 6). Normal appearance at rest, but failure of the affected side to move with crying, indicates hypoplasia or aplasia of the depressor anguli oris muscle. Facial nerve palsies are more like to occur with prolonged labor and compression of the facial nerve against the sacral bone or by use of forceps during delivery. The paralyzed side will have loss of the nasolabial fold, drooping of the mouth, and the mouth drawn to the normal side. It is important to determine whether there is branch 1 involvement of the facial nerve paralysis, because of the risk of corneal injury with improper eyelid closure. Most facial nerve palsies resolve spontaneously within days, but may persist weeks to months. Asymmetric crying facies caused by hypoplasia or aplasia of the depressor anguli oris muscle can be part of a genetic syndrome, the most significant association being congenital cardiac defects (Cayler syndrome).

Congenital soft tissue lesions of the oral cavity are common and practitioners must be able to distinguish normal findings from those that require intervention. There are 3 common types of oral inclusion cysts, which are epithelial tissue remnants. Inclusion cysts are small white or translucent papules or cysts noted in 75% of newborns, although the prevalence of inclusion cysts in premature infants is less than that of their term counterparts. Inclusion cysts are generally asymptomatic and require no further evaluation or management except for reassurance. Most cysts
resolve spontaneously by 3 months of age. Three types of inclusion cysts can be seen in newborns:

1. Epstein pearls are the most common of the congenital inclusion cysts, occurring in 75% to 80% of all neonates. They are found scattered along the midline raphe of the hard palate (Fig. 7).

2. Bohn nodules are heterotopic salivary gland remnants located on the buccal or lingual mucosal surface of the alveolar ridge (not the crest) or on the hard palate, away from the raphe (Fig. 8).

3. Dental lamina cysts are heterotopic salivary gland remnants located on the crest of the alveolar ridge.

Fig. 7. Epstein pearls.
Failure of midline fusion during embryogenesis can result in the spectrum of cleft lip, cleft palate, cleft lip and palate, or submucosal cleft palate. Lip closure occurs during week 5 to 6 of embryonic development, the hard palate forms during week 6 to 10, and the soft palate fuses during week 10 to 12. Cleft lip and palate are among the common congenital abnormalities, with a prevalence of 17 in every 10,000 live births, and cleft lip with palate is more common than isolated cleft palate (Fig. 9). Cleft lip can be unilateral, bilateral, or median, but midline lip cleft is rare.

The cause of clefts remains poorly understood, with genetic, syndromic, and environmental factors all being implicated. Teratogen exposures linked to cleft development include viral infections, metabolic abnormalities, medications, and drugs. Of inherited forms, the most common familial cleft lip and palate with no other anomalies is Van der Woude syndrome. Cleft lip and/or palate can be associated with genetic syndromes having other anomalies, including the spectrum of chromosome 22q11, oral-facial-digital syndrome, and Treacher-Collins syndrome. Micrognathia caused by mandibular hypoplasia can be isolated or associated with cleft palate, through the mechanical interference of the embryonic tongue with fusion of the 2 halves of the palate midline. This condition is termed the Robin sequence or Pierre Robin syndrome.

Care of neonates with cleft lip and/or palate requires aggressive feeding support and evaluation for airway concerns. Surgical primary lip repair is often undertaken at 3 months of age and primary palatal repair around 6 months. Management after discharge is often best coordinated through a multidisciplinary cleft and craniofacial team composed of experienced members of the medical, surgical, dental, and allied health disciplines.

Fig. 8. Bohn nodules.

Fig. 9. Cleft of soft palate.
Submucosal cleft palate is a milder or incomplete form of cleft palate. Examination may reveal a bifid uvula, muscular defect with overlying membrane, or a bluish mucosal line the length of the soft palate. This form is often clinically challenging to detect in the neonatal period. However, submucosal clefts are often functionally significant, because the levator muscles do not interdigitate across the cleft to form the normal levator sling. If the levator muscle cannot elevate and retract the posterior soft palate to divide the nasal and oral pharynxes, the result is velopharyngeal incompetence, which can affect both feeding and speech development.

Humans are usually born edentulous, with the first primary teeth emerging between 6 and 8 months of age. However, tooth eruption occurs before birth (natal teeth) or within the first month of life (neonatal teeth) at a rate of 1 in 2000 to 1 in 3000 live births. Central mandibular incisors are the most likely to erupt early and these are most often the primary dentition, not extra teeth, so should not be extracted without cause. Natal teeth are most commonly an isolated finding (Fig. 10), but can be associated with genetic syndromes such as chondroectodermal dysplasia (Ellis–van Creveld syndrome) and oculomandibulofacial syndrome (Hallermann-Streiff syndrome). Management is most often observation, although extraction may be considered if teeth are mobile and present an aspiration risk, interfere with breastfeeding, or lead to Riga-Fede ulceration.

Ankyloglossia (tongue-tie) is anchoring of the tongue anteriorly causing limited tongue movement, which occurs in 4% to 10% of babies. Ankyloglossia varies in severity and clinical consequence with considerable debate regarding optimal management. Restricted tongue movement can lead to a variety of problems in infants and children, such as inability to latch with breastfeeding, improper speech development, and compromised oral health.

Initial symptoms of ankyloglossia include maternal nipple pain with breastfeeding, loss of suction or a clicking sound while feeding, and poor latch. On examination, there is a frenulum inserted near the tip of the tongue, inability to extend the tongue to the lips or to the roof of the mouth, and notched or heart-shaped tongue on extension. Although interference with breastfeeding is the most common sequela, significant ankyloglossia is also associated with articulation difficulty, but not delay in onset of speech. Difficulty with oral hygiene increases the risk of dental caries and periodontal disease. Inability to lick and perform other activities that require tongue extension may have social consequences.

Fig. 10. Natal tooth.
Management of breastfeeding difficulties should include consultation with a lactation specialist, but frenectomy improves feeding for mother and baby significantly better than the intensive support of a lactation consultant.\textsuperscript{44} Indications and timing of surgical division for ankyloglossia have been investigated and in 1 randomized, prospective, but unblinded trial of neonates with feeding concerns and ankyloglossia, and feeding improved in all of the infants who received immediate division of the frenulum, but in only 1 infant who received intensive lactation support. Frenotomy was then offered and performed for the infants in the control group and all but 1 baby improved and fed normally after the procedure.\textsuperscript{48} Frenotomy (also called frenectomy) can be performed with blunt scissors, cautery, or laser if a simple membrane is present, but more complex anatomy should be referred for frenuloplasty. Absence of the inferior labial frenum is strongly correlated with infantile hypertrophic pyloric stenosis\textsuperscript{49} and absence of the inferior labial frenum and lingual frenulum is commonly noted in Ehlers-Danlos syndrome.\textsuperscript{50}

More recently, discussions have arisen around clinical consequence and management of a constricted maxillary frenum. The maxillary frenum normally extends over the alveolar ridge to form a raphe and persistence of this raphe during dental eruption may lead to widely spaced central incisors, termed a diastema. In addition to a cosmetic effect, there is concern that a prominent maxillary labial frenum can result in difficulty with plaque control and perhaps increased risk for dental caries caused by liquid trapping under the upper lip. Additional research is needed to better understand the appropriateness of performing preventive maxillary frenectomy on young children. At present, treatment is indicated in the rare cases in which the frenum attachment exerts tension on the gingiva of a permanent tooth or if the cosmetic appearance is unacceptable following orthodontic closure of the diastema.\textsuperscript{51,52}

**SUMMARY**

The development of each sense is crucial to successful interaction of babies with their mothers, ranging from bonding to feeding and eventually to capacity to learn. Primary care physicians have the greatest opportunity to detect visual and hearing deficits, to intervene, and to improve lifelong development and intellectual achievement.

**REFERENCES**


