13 Management of Otopathology and Hearing Loss in Children with Cleft Palate and Craniofacial Anomalies

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Children with cleft palate and other craniofacial anomalies are at risk for a variety of otopathologies including otitis media and structural deformities of the outer and middle ear. These conditions are usually associated with conductive hearing impairment; however, when hearing loss occurs as part of a syndrome, there may be sensorineural or mixed hearing loss. This chapter will review the etiology of these conditions and their medical management. We will also examine the assessment of hearing and various technologies available for treatment of hearing loss.

13.1 Etiology and Medical Management of Middle Ear Disease

Because of the shared embryological origins of the ear and other craniofacial structures, patients with cleft palate and craniofacial anomalies are at increased risk for a variety of otopathologies including otitis media, cholesteatoma, and structural abnormalities of the outer and middle ear. Otitis media is, by far, the most prevalent otopathology seen in patients with cleft palate (Paradise et al. 1969; Moller 1975; Hubbard et al. 1985; Rynnel-Dagoo et al. 1992; Sheahan et al. 2003, 2004; Flynn et al. 2009; Zheng et al. 2009); however, a few may require tympanoplasty, tympanomastoidectomy, or ossicular chain reconstruction (Goudy et al. 2006). The high prevalence of otitis media in this population is due primarily to poor Eustachian tube function during swallow (Doyle et al. 1980; Takahashi et al. 1994). In the normal swallow, the velopharyngeal sphincter, which includes the levator veli palatini and tensor veli palatini muscles, closes the nose from the oral cavity to prevent reflux of oral secretions and food into the nasopharynx. The tensor veli palatini, the primary muscle responsible for opening the Eustachian tube, originates from the Eustachian tube and inserts on the palate where it joins muscles from the contralateral side to form a supporting sling for the palate. In patients with cleft palate, the sling is poorly developed and follows a more vertical orientation resulting in greater risk of reflux into the middle ear. The outcome is poor aeration of the middle ear and a high prevalence of otitis media (Bluestone 1971; Fria et al. 1987; Flynn et al. 2009). Although the prevalence decreases with age, some patients continue to experience otitis media as adults (Handzic-Cuk et al. 1996, 2001; Sheahan et al. 2003).

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S. Berkowitz (ed.), *Cleft Lip and Palate*,
DOI 10.1007/978-3-642-30770-6_13,

Diagnosis of acute otitis media (AOM) requires (1) a history of acute onset of signs and symptoms; (2) middle ear effusion based on evidence of a bulging tympanic membrane, limited or absent mobility of the tympanic membrane, observation of an air-fluid level behind the tympanic membrane, or otorrhea; and (3) signs and symptoms of middle ear inflammation with evidence of otalgia (ear pain) or erythema of the tympanic membrane (AAP 2004). Otitis media with effusion (OME) is defined as the presence of fluid in the middle ear without signs or symptoms of AOM. It is more difficult to detect than AOM because in most cases, the only overt symptom is fluctuating hearing loss.

Otoscopy refers to examination of the eardrum or tympanic membrane using an otoscope and is part of any complete physical examination (Stool 2006). Tympanometry, an important adjunct to otoscopy, is a physiologic measure that provides information concerning the mobility of the tympanic membrane and middle ear system as well as middle ear pressure and estimates of equivalent volume, each of which may be associated with various middle ear conditions (Roush and Grose 2006). Figure 13.1 shows a series of tympanograms and their clinical interpretation.

Acute otitis media, when treated medically, is treated with antibiotics to cover for *H. influenzae*, *S. pneumoniae*, or *M. catarrhalis*. For most penicillin-resistant organisms, amoxicillin/clavulanate has proven effective (AAP 2004). When middle ear effusion persists for more than 3 months, as it often does in children with cleft palate, it is considered chronic. Tympanostomy tube placement is a common practice for those children, and improvements in hearing have been noted (Gould 1990) with relatively few complications (Curtin et al. 2009); however, multiple tube insertions have been associated with persistent conductive hearing loss (Goudy et al. 2006). Although it is unclear whether this is due to the placement of the tympanostomy tubes or to middle ear damage from the inflammation associated with otitis media, some clinicians have favored hearing aids over multiple tube placements (Maheshwar et al. 2002).

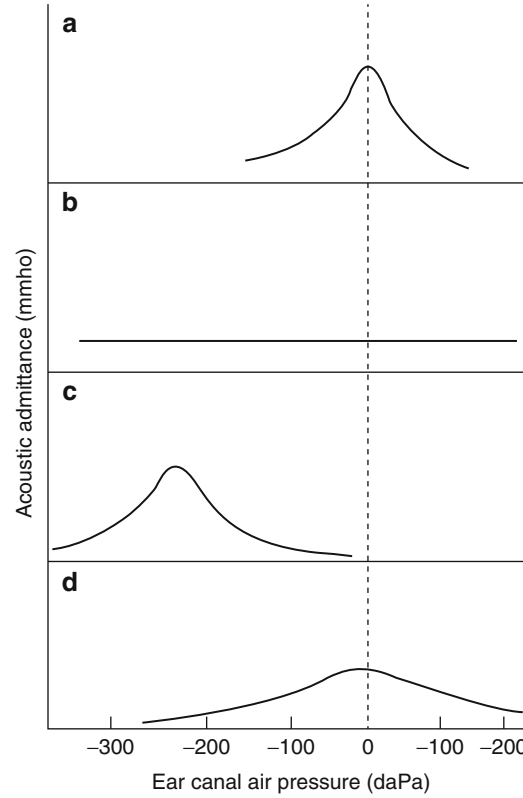

Fig. 13.1 Tympanometric patterns associated with normal middle ear function (**a**), middle ear effusion (**b**), negative middle ear pressure (**c**), and reduced middle ear mobility (**d**) (Reprinted with permission from Roush and Grose (2006), p. 378)

Complications from middle disease, although relatively rare, include perforation or retraction of the tympanic membrane, resulting in cholesteatoma. Cholesteatoma refers to a benign but locally erosive mass of squamous cells that often begins with a tympanic membrane retraction pocket or perforation. It is a potentially serious condition that warrants evaluation by an otolaryngologist and subsequent surgery.

Surgical repair of the palate has been shown to improve Eustachian tube function and decrease the frequency of otitis media and need for tympanostomy tubes (Bluestone 1971); however, it may take several years for Eustachian tube function to fully recover (Smith et al. 1994; Goudy et al. 2006). As with

the non-cleft palate population, the likelihood of hearing loss decreases with age (Gordon et al. 1988); however, hearing loss persists in adulthood for some patients.

13.2 Assessment of Hearing

Newborn hearing screening is now a standard of care throughout the United States (JCIH 2007). Two technologies are used for newborn screening and both employ physiologic methods; that is, they involve physiologic measurement of auditory function obtained without the infant's active participation. The first involves the measurement of otoacoustic emissions (OAEs) which are low-intensity sounds produced by the inner ear in response to acoustic stimuli (tones or clicks) and detected by a sensitive microphone placed in the ear canal. Successful recording of OAEs confirms a healthy inner ear (cochlea) and is consistent with normal or near-normal hearing sensitivity. Since the middle ear is involved in both the conduction of acoustic stimuli to the inner ear and reverse transmission of OAEs to the ear canal, their presence also confirms normal middle ear function. OAEs are "preneural" so their inclusion in the test battery makes it possible to evaluate the auditory system at the level of the inner ear without involving higher auditory centers. However, absent OAEs may be due to a variety of conditions ranging from middle ear dysfunction to profound cochlear hearing loss; other tests are needed to resolve their absence. OAE screening may be performed by a variety of professionals or support personnel, while diagnostic OAE testing is performed by an audiologist as part of a test battery. Another physiologic test used for both hearing screening and diagnosis of hearing loss is the auditory brainstem response (ABR). Like OAEs, ABRs are elicited by acoustic stimuli presented in the ear canal; however, ABRs are neurological responses obtained from surface electrodes attached to the head and provide information regarding the functional integrity of the auditory nerve and brainstem pathway. When used in a diagnostic test battery, audiologists use frequency-specific ABR testing to estimate hearing threshold levels.

Although OAE and ABR tests provide valuable information regarding the auditory system at the preneural, auditory nerve, and brainstem levels, they are not considered true tests of hearing in the perceptual sense. Comprehensive assessment of hearing requires behavioral tests, that is, procedures that involve observation of a listener's response to sound. Methods used for behavioral assessment of hearing vary depending on the child's age and developmental status. When infants reach a developmental age of approximately 6 months, most can be tested using an operant conditioning procedure known as visual reinforcement audiometry. By age 3–4 years, behavioral testing can be accomplished using conditioned play procedures, and by 5 years of age, typically developing children respond by raising their hand or pressing a response button.

To summarize, newborn hearing screening is conducted using physiologic measures: otoacoustic emissions or auditory brainstem responses, alone or in combination. Infants who do not pass the newborn hearing screening are referred to an audiologist for comprehensive assessment using these and other specialized procedures to determine if hearing impairment is present and, if so, to ascertain the type and degree of hearing loss. When infants reach a developmental age of approximately 6 months, they can be tested using behavioral methods. When permanent hearing loss is diagnosed, most children benefit from acoustic amplification (hearing aids). Those with severe-profound hearing loss are likely to benefit from a cochlear implant.

The initial goal of the audiologic assessment, whether conducted using physiologic or behavioral methods, is to obtain a frequency-specific estimate of the child's hearing thresholds for each ear, based on detection levels for air- and bone-conducted test stimuli. Air conduction audiometry involves the presentation of pure tones from an earphone or insert receiver; bone conduction audiometry involves the presentation

of signals through a bone vibrator placed behind the ear on the mastoid process. Testing is usually performed at octave intervals from 250 to 8,000 Hz for air conduction and 250–4,000 Hz for bone conduction. Thresholds are displayed on an audiogram which plots threshold levels in decibels hearing level (dB HL) as a function of frequency, using standard symbols (Fig. 13.2). The degree of hearing loss can be summarized by averaging the pure-tone air conduction thresholds in the mid-frequencies. Terms used to classify hearing levels include normal (0–15 dB HL), borderline normal (16–25 dB HL), mild (26–45 dB HL), moderate (46–75 dB HL), severe (76–0 dB HL), and profound hearing loss (>90+ dB HL). Borderline categories may be described using a combination of terms, such as moderate to severe (Roush and Grose 2006).

When pure-tone air conduction thresholds are abnormally elevated, bone conduction testing is performed to differentiate problems with sound transmission lateral to the inner ear. As shown in Fig. 13.2a, when air and bone conduction thresholds are equally elevated, the loss is described as sensorineural. Conductive hearing loss, illustrated in Fig. 13.2b, is characterized by normal or near-normal bone conduction thresholds with elevated air conduction thresholds. A mixed hearing loss, shown in Fig. 13.2c, is characterized by abnormal responses to both air conduction and bone conduction signals, with air conduction thresholds poorer than bone conduction thresholds.

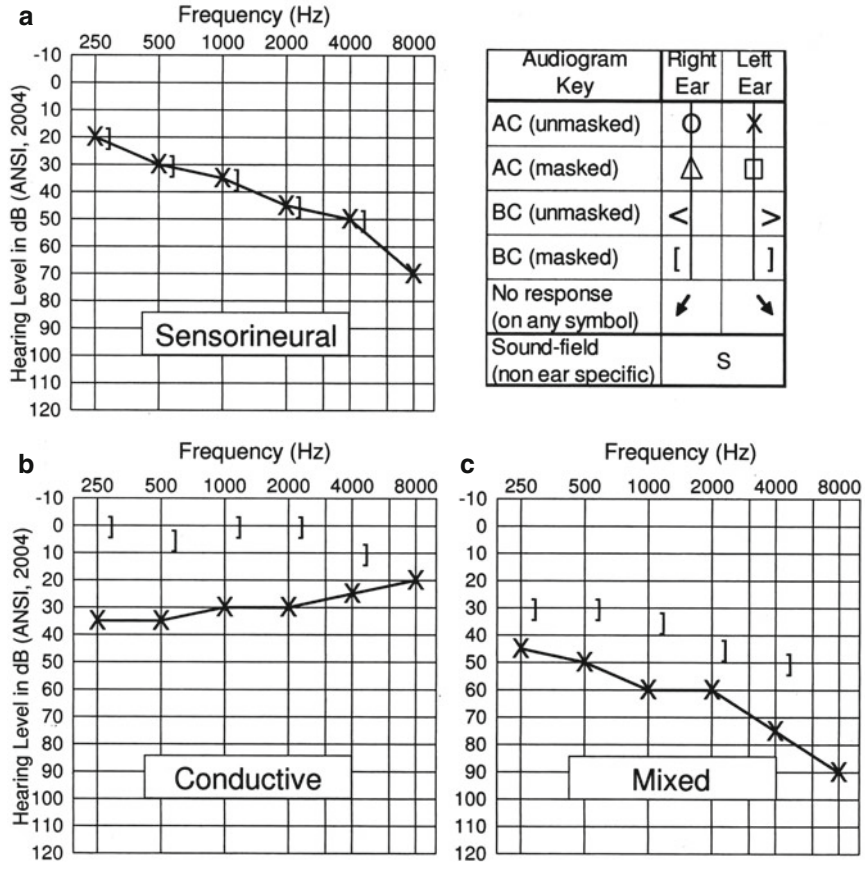

Fig. 13.2 Audiograms illustrating mild to moderate sensorineural hearing loss (**a**), mild conductive hearing loss (**b**), and moderate-to-severe mixed hearing loss (**c**) (Reprinted with permission from Kramer (2008), p. 153)

13.3 Hearing Loss in Children with Cleft Palate and Craniofacial Anomalies

As noted earlier, most hearing losses that occur with cleft palate and craniofacial anomalies are conductive in nature. That is, incoming sounds are attenuated by aural atresia or middle ear disease. When cleft palate or craniofacial anomalies occur as part of a syndrome, they may include cochlear or labyrinthine dysplasias that result in sensorineural or mixed hearing loss. These include Treacher-Collins syndrome, Goldenhar syndrome (oculoauriculovertebral or OAV spectrum), and craniofacial microsomia. CHARGE syndrome may involve abnormalities of the outer, middle, and inner ear, while Stickler syndrome, which is often identified by a Pierre Robin sequence, has been associated with progressive sensorineural hearing loss. Velocardiofacial syndrome (chromosome 22 deletion syndrome) may include sensorineural hearing loss, and when accompanied by cleft palate, Eustachian tube dysfunction, and otitis media, a mixed hearing impairment. Syndromes involving craniosynostosis may include auricular dysplasias, canal atresia, ossicular fixation, and sensorineural hearing loss. Tympanostomy tube placement may be indicated if feasible, and some patients will be candidates for ossicular reconstruction. Jarhsdoerfer et al. (1992) developed criteria to predict the success of atresia surgery based on otologic findings that include the presence of ossicles, an oval window, round window, facial nerve, and external ear. Unfortunately, only about half of patients with aural atresia are candidates for surgical repair, and many of those will require revision surgeries (Jarhsdoerfer et al. 1992).

13.4 Management of Hearing Loss in Children with Cleft Palate and Craniofacial Anomalies

The challenge of determining hearing status for these children often begins at birth. Many infants with cleft palates do not pass the newborn hearing screening because of middle ear effusion and require referral to a pediatric audiologist for follow-up assessment. Although most will not be found to have a permanent, underlying sensorineural or conductive hearing loss, it is important to determine the child's hearing status early as possible. For typically developing infants, the test used to make this determination, diagnostic ABR, can be easily accomplished in natural sleep. But for infants born with cleft palate, the test is often complicated by middle ear effusion. Although ABR testing can be completed using bone-conducted as well as air-conducted stimuli, it is preferable to perform the ABR when the middle ear is free of effusion. Another potential complication is the noisy breathing of some young infants with cleft palate, which may result in artifacts that affect the quality of the ABR when recorded in natural sleep. If an adequate ABR study cannot be performed in natural sleep, it must be completed with sedation or under general anesthetic. Sedated or operating room procedures add additional time and expense but enable the high-quality assessment needed for prompt, accurate diagnosis. The clinician must decide whether a separate procedure for diagnostic ABR is warranted or if the test can be performed at the time of cleft repair or tympanostomy tube placement, being mindful of the importance of avoiding excessive delay between screening and diagnostic ABR evaluation.

When hearing loss is diagnosed, timely and appropriate intervention are essential. In many cases, medical management will resolve a transient middle ear disorder and restore normal hearing. But hearing loss in children with cleft palate and craniofacial anomalies is often chronic, even with appropriate medical management. When conductive hearing loss cannot be resolved through medical intervention, hearing aid use should be considered. For children, hearing aids worn behind the ear are preferred whenever possible because they allow flexibility in programming, and they work well with assistive listening devices used at home and in the classroom. When acoustic amplification is precluded by aural atresia or chronic otorrhea, a bone conduction device is usually indicated. Most bone conduction hearing aids consist of an oscillator fitted to a headband that maintains pressure of the bone vibrator

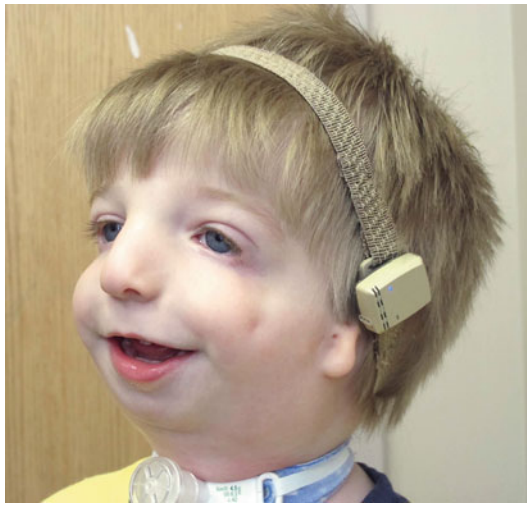

Fig. 13.3 A 3-year-old child with Treacher-Collins syndrome using a Baha softband bone conduction device. A vibrotactile transducer, held in place by an elastic strap, transmits vibrations from incoming sounds directly to the inner ear, bypassing the outer and middle ear

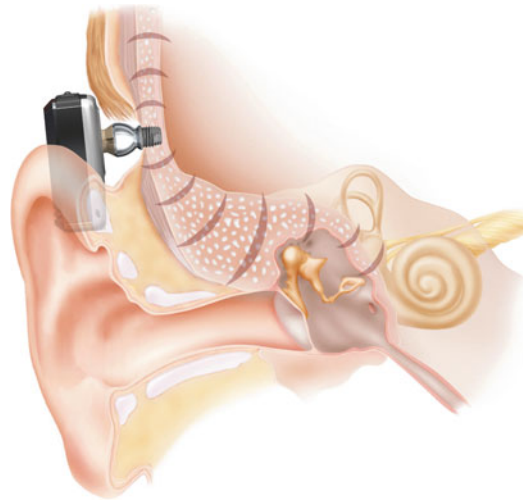

Fig. 13.4 The implanted bone-anchored hearing device consists of an external sound processor attached to a titanium abutment that is surgically implanted into the skull (Courtesy of Cochlear Corporation)

against the scull (Fig. 13.3). Because the inner ear is often unaffected by aural atresia, the bone oscillator is able to bypass the impaired outer and/or middle ear and deliver sound directly to the cochlea. Older children may be candidates for a percutaneous, bone-anchored device that delivers vibrotactile stimulation from a hearing instrument mounted to a titanium abutment surgically implanted in the temporal bone (Fig. 13.4). The surgically implanted, bone-anchored device provides stable retention and efficient transmission of vibrotactile signals, making it usable with mixed hearing impairment.

As noted earlier, although most hearing losses in children with cleft palate or craniofacial anomalies are conductive in nature, it is important to provide comprehensive audiological assessment to rule out a sensorineural or mixed impairment. When sensorineural hearing loss is identified, the audiologist and otolaryngologist will determine the best approach to hearing technology. An ear-level, air conduction hearing aid may be appropriate, or in cases of severe-profound hearing loss, a cochlear implant may be indicated. Cochlear implants bypass the inner ear, providing electrical stimulation from an external sound processer and coil that transmit digital signals across the skin to an internally implanted electrode that provides direct stimulation of the auditory nerve.

13.5 Summary

Hearing is vital for speech, language, cognitive, and social development. Nearly all children with cleft palate and craniofacial anomalies will experience middle ear disease and/or conductive hearing loss, and some are at risk for sensorineural or mixed hearing impairment. Careful monitoring of ear and hearing status by an otolaryngologist and audiologist are essential. With appropriate management, conductive hearing loss can often be avoided or ameliorated. When there is permanent (sensorineural) or chronic conductive impairment, several hearing technologies are available for delivery of sound via air conduction, bone conduction, or electrical stimulation.

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