Review article

Amblyaudia: A common diagnosis or a diagnosis of exclusion?

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ABSTRACT

Background: Amblyaudia represents the spectrum of auditory processing deficits (APD) that are felt to be associated with asymmetric hearing loss (AHL) and may have detrimental effects on hearing and language development. Findings: The hallmark finding in amblyaudia is an abnormally large asymmetry between the two ears during dichotic listening tasks with either normal or below normal performance in the dominant ear. Imbalanced signaling between the two ears during critical periods may result in altered or maladaptive patterns of neural connections that persist even after the secondary pathology has resolved.Amblyaudia frequently presents with difficulties in attention, speech comprehension, and reading. Dichotic hearing tests are used for screening and diagnosis because they put stress on the binaural auditory system and reveal any asymmetry between the auditory pathways. Interventions: The commonness and severity of amblyaudia in the population can be minimized by early identification and correction of AHL. Intervention may be in the form of reducing the intensity of input to the dominant ear during dichotic listening tasks or as a short- term intervention program called Auditory Rehabilitation for Interaural Asymmetry (ARIA).Conclusion: Amblyaudia is an important source of difficulties in attention, speech comprehension and reading, often difficult to diagnose and early diagnosis and implementation of corrective measures can make a significant difference in the development of these children.

KEYWORDS: Amblyaudia; auditory processing deficit; asymmetric hearing loss; conductive hearing loss

Introduction

A plethora of developments in the diagnosis and treatment of pediatric hearing lossin the recent past has as of yet failed to craft an adequate solution to the quandary of unilateral or asymmetric hearing loss. As a result, the greater part of the mass of patients with this condition is often misidentified, unidentified, mismanaged or lost to follow-up.1The common belief that the opposite ear with normal or near-normal hearing will compensate for a unilateral hearing loss, often leads to the undertreatment of asymmetric hearing loss in children. This factor of normal or near-normal hearing in the contralateral ear in cases of asymmetric hearing loss due to acquired or transient etiologies, often misleads parents and guardians, leading to a late diagnosis of the condition, and oftentimes, the child may not even be aware of the existence of the problem.2

Amblyaudiais a new diagnostic category within Auditory Processing Disorders (APD), characterized by deficits in the binaural integration of verbal information that is diagnosed by results from dichotic listening tests.3 The hallmark finding in amblyaudia is an abnormally large asymmetry between the two ears during dichotic listening tasks with either normal or below normal performance in the dominant ear.4Children who witness temporary

hearing loss, most commonly from ear infections, are at an increased risk of developing amblyaudia.5 The persistence of auditory perceptual deficits after the ear is audiometrically normal is similar to the poor vision through the "lazy" by structurally normal eye in amblyopia. Like the etiology of amblyopia, the source for amblyaudia may also stem from the maladaptive plasticity in the central auditory system during critical periods in development.6

Physiology of Binaural Hearing

The auditory system depends on the integration of afferent signals from both ears. Sound waves are collected by the external ear and transmitted inward for processing. When the sound waves reach the tympanic membrane, they are converted into mechanical vibrations which are then amplified by the ossicles and converted into fluid waves by the oval window. In the cochlea, the timing information is enhanced for sound localization.Sound input from the periphery is converted into neural signals in the inner ear and is transmitted by the auditory nerve to the cochlear nucleus, the first station of sound processing in the brain. The cochlear nucleus contains excitatory and inhibitory protrusions that innervate second-order auditory brainstem nuclei in both hemispheres of the brain.7 The integration of these bilateral excitatory and inhibitory signals is essential for complex auditory processing. A mismatch in bilateral auditory input during development can disrupt the integration of binaural cues at the level of the auditory midbrain and cortex.8

Definition of Amblyaudia

The existence of amblyaudia has been known for a while now, and consequently a number of definitions have been proposed. Amblyaudia represents the spectrum of auditory processing deficits (APD) that are felt to be associated with asymmetric hearing loss (AHL) and may have adverse effects on hearing and language development.9 The authors believe that the working definition for Amblyaudia proposed by Kaplan AB et al.10 to be the most appropriate, which states that amblyaudia is characterized by three key aspects: unilateral or asymmetric auditory dysfunction, hearing loss resulting in decreased afferent signal to the auditory cortex and which occurs during a critical period of development with subsequent correction or reversal.Asymmetry is critical to the pathophysiology of this disorder; if both sides are uniformly affected, the mismatch in binaural processing and neural maturation won't exist. Unfortunately our understanding of the condition is still far from complete and there does exist some ambiguity in the definition. Any peripheral etiology of hearing loss may put an individual at risk for amblyaudia, but the effect of different types, severity and duration of hearing loss is currently unknown. Also, the minimum threshold of hearing loss necessary to result in amblyaudia is still unknown. The hearing loss must occur during a critical period of early childhood development with subsequent rectification or reversal. The critical period is not yet well defined, and duration of asymmetric hearing loss may also be an important factor.

Pathophysiology Of Amblyaudia

During brain development, higher cortical sensory centers depend majorly on afferent input to form organized neural circuits. In the auditory system, early exposure to sound promotes the proper development and maturation of the auditory processing centers.11Imbalanced signaling between the two ears during critical periods may result in altered or maladaptive patterns of neural connections that persist even after the auxiliary pathology hasresolved.12As a result, neural transmissionfrom the affected side may be significantly weakenedas compared with the unaffected side. The overall effect of thisasymmetric signaling is the creation of a "dominant" ear athigher stages of the central auditory pathway, much like theemergence of a dominant eye at the level of the primary visualcortex, as described in amblyopia. During criticalperiods of neural plasticity, this imbalanced weighting of interauralcues may disrupt the representation of binaural stimuli ina manner that persists for an unknown length of time after thehearing loss is reversed in later life.13 Animal studies have shown that during critical periods of development, the central auditory pathways are extremely sensitive to peripheral deprivation, resulting in lasting abnormalities of binaural processing.14

Consequences Of Amblyaudia

A majority of individuals with amblyaudia are found to have normal or near-normal pure tone thresholds, but these individuals are predicted to have difficulty with auditory processing that depend on bilateral cues, i.e. sound localization and signal-to-noise suppression. These tasks contribute greatly to human communication by giving cues as to the sourceof sounds and by enabling the separation of specific sounds in

the setting of background noise.9The auditory system of a child with amblyaudia compensates for the imbalanced inputs between ears by adjusting sensitivity to binaural spatial cues. Because of this adaptation, amblyaudia is associated with degraded binaural spatial hearing and sound localization.Binaural spatial hearing and sound localization play key parts in speech comprehension, particularly in noisy surroundings.15 Difficulty with binaural spatial hearing may have secondary effects on educational development because of linguistic and cognitive problems. Linguistic difficulties result from the impairments in phonetic and phonological coding of sounds and articulation and syntax impairments. Cognitive difficulties are closely related with linguistic difficulties. The misperception of some phonemes produces a poor semantic database, which affects reading fluency and comprehension and can result in dyslexia. Because of the difficultyisolating auditory information, amblyaudia has been linked to attention problems that can affect educational development. Difficulties with adaptive skills and self- esteem have also been seen in children with amblyaudia.[16]

Clinical Presentation Of Amblyaudia

Amblyaudia often presents with difficulties in attention, speech comprehension, and reading. Inattention, such as inconsistent response patterns and shorter attention spans, is common in children with amblyaudia.9 Additionally, speech comprehension is made difficult, especially within noisy environments, due to the reduced ability to localize sound, comprehend verbal material, and follow verbal directions with multiple steps. The deficits in speech understanding also influence reading fluency and comprehension through a chain of reactions.There's mixed study regarding when the changes in neuronal sensitivity dissipate.[15]

The Association Of Otitis Media And Amblyaudia

Amblyaudia may theoretically result from any form of asymmetric auditory deprivation, including conductive and sensorineural causes, occurring during a critical period of development. Investigations on amblyaudia have thus far predominately investigated conductive hearing losses (CHL), mainly because, by definition, a diagnosis of amblyaudia necessitates complete reversal of hearing loss, which is most probable to occur in the setting of CHL through surgical correction.9

Amblyaudia has been correlated with early life auditory deprivation such as otitis media (OM) with prolonged periods of temporary hearing loss. Otitis media (OM) is a common childhood illness that is characterized by purulence and/or the accumulation of excessive mucin in the middle ear space. In some instances, the mechanical properties of the middle ear system can be altered by the presence of viscous fluid in the typically air-filled tympanum. In these cases, the auditory signal transmitted to the developing central nervous system is degraded, prompting many clinicians and researchers to hypothesize that OM may represent a common form of developmental auditory deprivation. 17Although OM is physically restricted to the middle ear space, it can interfere with the transmission of acoustic signals to the inner ear and, by extension, the entire auditory system. The middle ear pathology and accumulation of excess, viscous mucin that typically accompany OM can disrupt the acoustico-mechanical properties of the middle ear system, producing a CHL.18 In addition to attenuating the overall amplitude of the acoustic signal, viscous fluid in the middle ear space can also delay transmission of the transduced waveform. Differences in the timing (interaural time difference) and amplitudes (interaural level difference) of acoustic signals arriving at the two ears play an essential role in spatial hearing, particularly in the horizontal plane.19

Nearly 89% of cases of otitis media presenting with CHL resulted in amblyaudia.9 During periods of temporary hearing loss, the auditory system is unable to completely manipulate acoustic input from the affected ear. When temporary hearing loss occurs during early periods of auditory experience, the central auditory circuits that support sound location may be recalibrated.[20] These adaptations may be advantageous during the period of hearing loss, even so, they often become maladaptive once normal hearing is restored. Therefore, even after normal hearing has been restored there is a possibility of long lasting auditory perceptual deficits.7

Roughly 80% of children under the age of 3 experience otitis media making it the most commonly diagnosed illness among young children in the United States.[21] Approximately 75% of all infants have at least one episode during the first two years and 25 percent of infants develop recurrent or persistent OM.[22] OM can often go undiagnosed as it can occur without the presence of pain or infection.[19]

Assessment And Diagnosis Of Amblyaudia

Amblyaudiacan not be identified by traditional hearing tests because there are no current tests that place the two ears in competition. Instead, dichotic listening tests are used for screening and diagnosis because they put stress on the binaural auditory system and reveal any asymmetry between the auditory pathways.5 Some of the tests that are used are the Competing Words subtest of SCAN, staggered spondaic words test and the randomized dichotic digits test. Dichotic listening tests give the listener with two monosyllabic words or digits simultaneously and the listener is expected to repeat the words presented in each ear.[23] Following testing, if the child has poor performance, the child should be referred for further testing to rule out retrocochlear disorder, confirm the weaknesses, and receive a final diagnosis.

The diagnosis of amblyaudia should address the nature of the processing weakness, which ear it affects, and the severity.A suggested severity criterion for amblyaudia is based off of interaural asymmetry and is graded as borderline amblyaudiawith an interaural asymmetry of 10 – 19%, mild with 20 – 29%, moderate with 30 – 39%, moderately severe with 40 – 49% and severe with an interaural asymmetryof more than 50%.5

Prevention Of Amblyaudia

The prevalence and severity of amblyaudia in the population can be minimized by early identification and correction of AHL, ideally within the critical window of auditory development. An opportunity for early identification of unilateral hearing loss lies with a robust universal newborn hearing screening program.Unfortunately at the present time, national newborn hearing screening programs are yet to become a reality in several developing countries, including India. In countries with established universal newborn hearing screening programs, the mean age of diagnosis of hearing loss—particularly unilateral hearing loss—has decreased significantly.Even still, despite improvements in screening, patients with unilateral hearing loss still appear to be under-diagnosed and undertreated.24,25

Treatment And Prognosis Of Amblyaudia

As amblyaudia is a relatively newly identified phenomenon,there is no standard practice for management. The processing difficulties of amblyaudia can be improved through interventions. Reducing the intensity of input to the dominant ear during dichotic listening tasks in order to strengthen the affected ear is generally the first part of the intervention. Then the intensity to the dominant ear is systematically increased as long as the affected ear's performance remains high. This should be continued until both ears can perform equally during dichotic listening tasks at equal intensity.[26] Another option is a short- term intervention program called Auditory Rehabilitation for Interaural Asymmetry (ARIA), which follows a similar progression. The ARIA method corrects amblyaudia in four therapy sessions over four weeks. The ARIA auditory training method involves gradual adjustment of the relative balance of stimulus intensity in the two ears, until the asymmetry in listening accuracy scores between the two ears is reduced.[27] Remote microphone hearing aids would provide equivalent (low) levels of amplification to the two ears. It's recommended that amblyaudia is corrected before fitting remote microphone hearing aids.[28]

prognosis of amblyaudia

The timing and duration of hearing loss both play key roles in the severity and prognosis of amblyaudia. There's mixed research regarding when the changes in neuronal sensitiveness dissipate. Some studies implied that impairments are long lasting even after normal hearing has returned. However, other studies say that deficiencies largely disappear after a few years of typical auditory experience.[20,29,30] Even if amblyaudia resolves by late childhood, there's no way to determine where the child would be without the period of asymmetrical hearing. Additionally, it's hard to determine whether or not the individual will witness ripple effects on verbal, cognitive, and social functions developed during the periods of atypical hearing.[9]

Conclusion

Amblyaudia is an important source of difficulties in attention, speech comprehension and reading, and is often difficult to diagnose unless borne as a differential in mind while testing these children and appropriate dichotic testing is applied. An early diagnosis and implementation of corrective measures can make a significant difference in the development of these children. Concrete steps have to be taken to increase awareness of this condition.

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