

Rediscovery of hearing disorders with normal audiometry or hidden deafness

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Some subjects present isolated hearing disorders for speech discrimination in difficult auditory conditions with normal tonal and speech audiometry in quiet and normally synchronized auditory brainstem responses (ABR). They have a preserved audibility but degraded intelligibility. The prevalence of this hidden deafness is reported between 5 and 15% [1].

Many synonyms and confusions are related to this set of hearing dysfunctions: “Hidden deafness”, “auditory processing disorders (APD)”, “supraliminal hearing disorders”, “auditory synaptopathy” or even “Central” hearing disorders. Notably, this latter definition is not correct because the exact origin is unknown and may be localized wherever from the outer hair cells (OHC) to top of auditory pathway, involved in cognition-related mechanisms of auditory perception. Indeed, an abnormal peripheral cochlear coding of sound features (spectral, intensity, temporal) could be responsible of APD. This peripheral dys-coding will be more or less reprinted at the upper levels of the auditory pathways. It seems clear that not all APD have a central origin and that the notion of “central” auditory processing disorders should be reviewed [2].

There are still many confusions between peripheral and central APD as well as between APD and auditory neuropathy spectrum disorders (ANSD). ANSD subjects present also a degradation of speech intelligibility. However, unlike APD, ABR in ANSD show abnormal wave V with preserved cochlear microphonic and/or otoacoustic emissions (OAE). If diffuse cochlear synaptopathy or auditory neuropathy may produce ANSD, APD may result from limited dysfunction at any level of the auditory system [3].

Only the three possible peripheral origins of APD are discussed below OHC, inner hair cells (IHC) and synapses-cochlear neural fibers [4]. Partial OHC loss may show no impact on hearing perception and be undetectable on audiometry tests performed at low intensities and in quiet. The hearing thresholds may remain normal with a third of OHC loss (3rd row loss). However, this loss may affect the supraliminal auditory processing of speech, especially in difficult auditory conditions. It is also suspected to increase the sensitivity to ototoxicity (e.g. to noise or age) [5]. Acute partial IHC loss in animal models leads to a decrease in cochlear output and an increase in the hearing thresholds [6]. However, up to 80% IHC loss, the threshold shift may be less than 5 dB [7]. It is possible to have normal tonal audiometry thanks to an enhancement of the central auditory gain. A selective synaptopathy of the neural fibers, showing high responses thresholds and spontaneous low discharge, can lead to decreased cochlear output for high intensities but preserved hearing thresholds [4]. The origins of this synaptopathy are unclear: age, noise, genetic factors and other ototoxics like cisplatin have been suggested [8]. Synaptopathy may appear before the loss of IHC and may likely be related to at least some of hearing difficulties

experienced in noisy environments despite normal or near normal hearing acuity [9-12].

Various psychoacoustic tests are used for the assessment of hidden deafness. Standard tonal audiometry must be completed by extending the thresholds research to higher than 8 kHz frequencies or to frequency bands lower than 1 octave band [4,13]. Speech audiometry in noise remains a gold standard supraliminal test to diagnose this disorder [14]. Various supraliminal auditory tests, that have been used in the past, were supposed to mainly investigate the central auditory processing. It is now clear that this processing extends from the peripheral to the central audition. For instance, the so-called “Central” auditory battery (CAB) of Demanez is frequently abnormal. This test has been conceived to detect poor phonemic discrimination in quiet and in noise, abnormal dichotic and demasking abilities, limited detection of changes in frequencies, intensities or duration patterns [15-17]. Individuals with APD have less ability to detect spectral and temporal features, which may be evaluated by various temporal and localization tests.

Otoacoustic emissions (OAE) are abnormal in case of OHC lesions [18]. Distortion Products are more sensitive to detect OHC dysfunctions than extended high frequencies audiograms [4]. The contralateral median olivocochlear inhibition (Collet’s effect) is impaired in APD subjects showing no decrease of their OAE amplitude in presence of contralateral noise [19,20].

Many electrophysiological tests have been used for the investigation of this set of disorders, however, their real clinical usefulness is still in discussion [21]. For instance, a decreased wave I but a better-preserved wave V amplitude have been reported on ABR [22-25]. Increased ratio of SP (summing potential) /AP (action potential) on electrocochleography may be a sign of auditory neural fibers damage [26]. Speech ABR and frequency following responses may demonstrate damage to high threshold cochlear fibers with loss of the phase locking and therefore impairment in the coding of the envelope of sounds, especially in noise [27,28]. Abnormal cognitive and/or obligatory cortical auditory potentials may be disturbed in APD subjects [29].

The treatment of hidden deafness also remains very challenging. It is based on hearing training and connectivity use. The adaptation of

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subjects to their disorders is improved by auditory training in quiet and in noise, as well as by increasing their audiovisual integration of speech in congruent and incongruent conditions, in order to benefit from all the useful cues for communication. It is possible to act on the environment and the subject / environment interaction by promoting an articulated and slower speech flow, a decrease in the noise and reverberation levels, a placement in front of the speakers for a better exposition to the speech signal and less to noise, and by encouraging visual aids use. Finally, the incoming signal may be improved by receiving the signal without noise by Frequency Modulation (FM) System.

Conclusion

In conclusion, it seems important to get away from the concept of a systematic central origin of auditory processing disorders, and to valorize the concept of possible peripheral auditory encoding disorders. A functional loss of the cochlea does not necessarily mean a loss of hearing sensitivity. Supra-liminal tests are probably not so obsolete that we thought. Their place in the current practice is re-discovered and increasing.

Authors contribution

GG and ND wrote the original manuscript followed by all co-authors commenting on the original manuscript and approved the final work.

Conflicts of interest

No conflicts of interest.

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