"Oh Granny, What Big Two Ears You've Got!" "All the Better to Hear You with, My Dear!" (Neuronal Circuit Recovery with a Cochlear Implant)

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Abstract

Introduction: Having an auditory pattern of behavior based on two ears is essential for sound localization, quality of hearing, understanding in groups, and with ambient noise.

Aims and objectives: Describe and discuss: (1) The consequences of unilateral deafness. (2) The gradual recovery of failing neuronal circuits when stimulated with a cochlear implant. (3) The case of a blind patient with sudden unilateral deafness who required cochlear implantation that is used as a common thread for the subject.

Materials and methods: Forty-five-year-old blind woman with sudden unilateral deafness. With unilateral deafness, she could not localize the sound source in terms of side nor if the sound came from above/below, near/far, or from front/back. Her hearing in groups and with ambient noise deteriorated. As a result, she lost her autonomy and required and underwent cochlear implantation.

Results: It took her 2 years to recover full sound localization, to be able to discriminate in groups, and to recover binaural fusion. Recovery was gradual. Her abilities to localize sound source in terms of side, of being near or far, or coming from above or below recovered separately, that is to say, at different periods of time.

Conclusion: After losing functional neuronal circuits, early stimulation with a cochlear implant helped to fully recover these circuits. Neuronal circuits for sound localization for side, coming from above or below, near or far are seemingly different since they recovered at different times. Hearing with both ears is essential for sound localization, discrimination in groups and with ambient noise.

Keywords: Binaural hearing, Cochlear implantation in unilateral deafness, Neuronal circuits in lateralization, Neuronal recovery with cochlear implantation, Sudden unilateral deafness and sound localization, Unilateral deafness in the blind, Unilateral hearing loss.

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Introduction

The dialog between Little Red Riding Hood and the Wolf—written by Charles Perrault[1]—in which Little Red Riding Hood tells the Wolf: "What big two ears you have!" and the Wolf answers "The better to hear you with!" emphasizes the concept of better hearing with two ears, whether the ears are large and hairy or not.

The sense of hearing connects us with the outside world and an important part in this connection is sound localization.[2–6] Moreover, to be able to locate the sound source, it is essential to listen with two ears since with one ear (e.g., single-sided deafness) is not possible. While for humans, this information is very important, for some animals it is vital. Such is the case for avoiding a predator, for the predator itself, or for an animal in search of a mate.

Understood things well, having an auditory pattern of behavior based on two ears is a crucial need for our "long eared wolf", as it is for the owl that hunts mice in the darkness based on auditory clues.[7,8] In its proper proportion, sound localization is essential for all animals, including humans. However, given our difference in life forms with animals and our ability to adapt, it is difficult to realize the real magnitude of unilateral (single-sided) deafness in humans. On the contrary, the need of binaural hearing goes beyond sound localization. Since we live in an environment with multiple and simultaneous sounds, the situation is more complex and requires other capabilities, including focusing attention on one sound and blocking others at the same time. In order to have this ability, a hearing pattern based on both ears is essential.[2–5]

Since single-sided deafness disabilities are not that evident, it has been difficult to convince physicians of the importance of treating single-sided deafness and of doing it early.

In August 2017, a blind patient with a sudden single-sided sensorineural hearing loss from which she had not recovered, consulted us. Her clinical case constitutes a master class in the subject and is supportive of the concept of treatment in these cases.

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Materials and Methods

Clinical Case

P.B. is a woman who is 45 years old (born on October 30, 2017) who is blind due to retinitis pigmentosa. She was diagnosed at age 5 and from then her eyesight started to worsen gradually and went blind at age 24 during her third year in Law School at the University of Valparaíso, Chile. She graduated as a lawyer and as a professional she has dedicated herself to promote and defend the rights of people with disabilities. She works for the judicial assistance corporation of the Chilean Ministry of Justice.

She lives two blocks from her job in the heart of Santiago, in streets with high transit of vehicles and circulation of people. Every morning she walks seven blocks by herself, accompanied by her dog in a trip that covers a walk to a square (for her dog), and then to her workplace. She works 8 hours a day from Monday through Friday and every afternoon she goes back home with her dog to an apartment where she lives by herself. She has friends who visit her and is socially active.

On April 18, 2017, she developed a profound left-sided sudden sensorineural hearing loss with tinnitus. Complete hematological and imaging studies did not reveal any specific cause and her presumed diagnosis was a single-sided deafness of viral origin. She was treated accordingly (systemic and intratympanic corticosteroids vasodilators, etc.) but she did not recover her hearing and her tinnitus persisted. Her right ear had normal hearing and has remained normal until present (Fig. 1).

She consulted us for the first time in October 2017. When she came, she was well informed of her treatment alternatives. Between what she described spontaneously and what we asked her, the following observations came to light:

She could not localize the sound source in terms of side nor could she determine if the sound came from above or below, or near or far.

On one occasion, some friends came to visit her at her apartment. They rang the bell and she opened the door for them. They talked briefly before they entered. Thinking that they had already entered but before they did, she closed the door and left them outside. On another occasion, she heard a dog bark at her dog and quickly returned, without noticing that the dog was in a fourth floor on the left side. When she walked on the sidewalk, in addition to having lost the ability to locate sound sources and their distances, she could not determine if she was walking next to a building or an open space such as a street. In social activities outside her home like in a restaurant or bar, her hearing in groups and with ambient noise deteriorated to the point that she could not participate. In addition to noticing a decrease in the quality of the sound, the effort required to hear and understand increased significantly. As a result of these new deficits, she lost her autovalence and could not go out without human help.

With her medical history and background, our recommendation was a left-sided cochlear implant.

On December 14, 2017, she was implanted with a Cochlear Nucleus CI24 with a 422-electrode array device via an exploratory tympanotomy–mastoidectomy approach.[9] Recovery was very satisfactory, healing was uneventful and the implant was activated on January 17, 2018 with a Nucleus 6 Cochlear Processor. Mapping was done by subjectively measuring threshold (T) and comfortable (C) levels and balancing each electrode for equal loudness perception.

At activation, the tinnitus disappeared and remained that way provided that the implant was activated. If the implant was turned off, tinnitus appeared but different and milder than before.

One-week post-activation, she noticed that she was starting to locate distances from objects and to distinguish if her dog was walking on a ceramic or a wooden floor.

Two weeks post-activation, she could calculate distances from objects like she used to and could also tell if a sound source (e.g., her dog) was coming or going. She could not localize clearly if a voice was coming from left or right or from above or below, but she could identify some areas or points from which the sound did not come.

Four weeks post-activation, she could localize somewhat better if the sound was coming from left or right but not from above or below. Pure tones in the implanted ear were normal (Fig. 2).

At 7 weeks, the patient considered—in her terms—"that her ear was in clear resurrection" and she went out to the street with her dog but with human assistance.

Four months post-implantation and 3 months post-activation, she did her usual seven blocks walk by herself and her dog and then returned to her apartment on her own.

The following days she restarted her usual walk alone with her dog and kept doing it as in the past since she considered that she

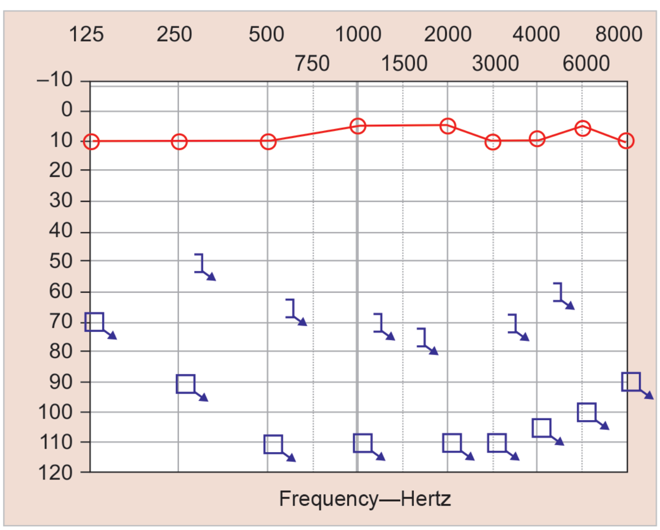

Fig. 1: Preoperative audiometry

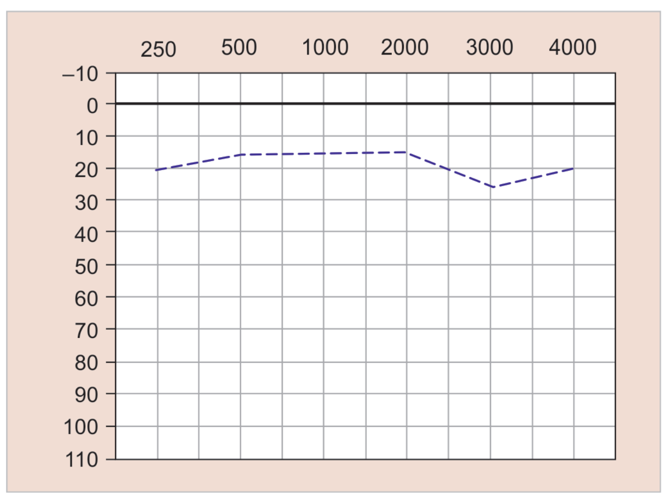

Fig. 2: Postoperative free field audiometry with cochlear implant in left ear (blue line). 100% word discrimination score

was recovering her autovalence. Although it was harder for her than before, she gradually improved and took her previous routine. At this stage, she could determine if she was walking next to a building or an open space such as a street. She could determine if a sound such as the sound of the motor of a car (her main sound reference for crossing streets) came from left or right, from far or near, or if it moved away or approached. She could also know if she was walking next to a building or an open space such as a street.

One-year post-activation, she told us that since her last checkup—3 months post-activation—great progress had occurred.

Her hearing in groups and with ambient noise has improved to the point that in a restaurant she can understand quite well and lately she can even understand what the people at the table next to her are talking about. The quality of the sound has improved and the effort to understand has decreased. However, the voices heard with the implant are still somewhat muffled and different from the voices heard with the normal ear. On the contrary, with the implant she could identify piano music very clearly and could differentiate the different musical instruments. Although she could determine if a sound came from left or right, from far or near, or if it moved away or was approaching, she was still not able to determine if the sound came from above or below. When the implant was deactivated, the tinnitus was very soft and on occasions, she could no longer hear it.

Two years post-activation, she considered that she had "practically recovered", that she was autonomous and that she could work as she did before her sudden hearing loss. Her listening in groups and with ambient noise had improved to the point that she could not only understand well but also perfectly identified the voices of those who spoke to her. The voices sounded natural and there were no differences between the normal and the implanted ear. She had also fully recovered her ability to locate sound. She could determine if it was coming from left or right, from above or below, from near or far, if it was approaching or moving away and at what distance. Her tinnitus completely disappeared (with or without using the implant). By now, she had recovered her previous autonomy, her social life, and her good character.

Discussion

Sound localization is processed by neural circuits between the cochlear nuclei and the geniculate body of the thalamus (where all impulses converge). This process is essential and complex since it involves not only determining if a sound comes from left or right but also if it comes from above or below, if it moves away or approaches, or if it comes from front or back and from how far.[2–6]

When our patient lost her hearing on the left side, she lost the capacity for all these components of sound localization.

To determine if a sound comes from the left or from the right, the central nervous system bases its analysis in disparities of time in sound arrival in both ears (interaural time differences) and interaural differences in intensity. The patient described this loss very clearly as well as not being able to tell if a sound came from above or below (e.g., a dog barking at her dog from a fourth floor).

She also lost her capacity to tell if a sound source was a close or a faraway source. This capacity is well developed in blind individuals.[5] Being blind she was accustomed to judge distances to static objects (e.g., walls) based on echo delays with her as the sound source. This is what she does in her house and also what she does on the street when she walks along the sidewalk and knows if she is next to a building (surface) or a street (open space).

On the contrary, for distances from sound sources in motion such as cars she also used these echo delays or Doppler effect.

By listening to the car's engines and analyzing the frequency of the pulses of these moving sound sources, she can determine distances and relative speeds. That is to say, if it moves away or approaches, and from how far. If the car (sound source) is approaching, the pressure pulses will arrive at her ear more frequently each time because they start closer to her, so the sound frequency rises. Once the car passes by, the reverse occurs.

Since in downtown Santiago, there are no sound or vibration transit signs, her main sound reference for crossing streets are the sounds of the motors of the cars. In the corners of the streets, she also uses as a timing reference the difference in sound of the motors that are stopped and of those that are about to advance or advancing.

The occasion in which some friends came to visit her at her apartment and she left them outside the door thinking that they had already entered, suggests that she did not have the Doppler effect capacity, and was acting using a pattern based on experience.

The principle is similar to the sonar (sound navigation and ranging) systems except that the echo sounding sonar systems are active and act as sound sources calculating distances and relative speed by echoing the sounds they generate.[5] The ultimate sonar is the bat, which is capable of modifying the frequencies of their output pulses in order to localize moving objects.[5] In daily practice, our patient uses many of the sonar elements to adapt and for this she requires both ears.

Interestingly, at 2 weeks post-stimulation, the first thing she regained was the ability to determine and calculate distances and if her dog was moving away or approaching. That is, if the sound approached or receded, it came from ahead or behind and from how far. Being able to determine with certainty if the sound came from left or right took her 3 months, and if it came from above or below it took her 2 years. The fact that the recovery of these different capacities occurred in different periods of time suggests that the neural circuits for them are not the same. This is suggestive that these are different neural circuits altogether, whose information merges at the central level.

In the end the information provided by these different circuits from both ears (binaural hearing) will converge at the geniculate body of the thalamus and end up fused in a single perception at a central level as binaural fusion. In other words, "binaurality" refers to the capacity of the brain to process the information of both ears. Bilateral hearing refers to listening with both ears and symmetry to listening with both ears in a symmetric manner.[3]

Although the stimulus and auditory mechanism is different in both ears, our patient has bilateral hearing and receives and uses symmetric information from both ears. Initially, the patient had binaurality (information from both ears was processed) but she was not able to fuse them centrally as a single perception (binaural fusion).

The sense of hearing is very complex and implies a processing scheme that goes beyond localization of the sound source. Since we live in an environment with multiple simultaneous sounds, the situation is more complicated, and our neural structure is prepared for that. There are other abilities like hearing with ambient noise and the capacity of focalizing attention toward a sound source and block other sound sources at the same time. For being able to do this, binaural hearing is essential.

Individuals who have only one hearing ear not only lack the capacity of localizing the sound source, they also have a "shadow effect" in which some information from the compromised ear (ear with hearing loss or deaf ear) is lost because the head causes a "shadow". There is also a "squelch" effect that relates to the central capacity of eliminating background noises of low intensity. Both ears are crucial for discrimination with ambient noise.

That is why individuals with an only hearing ear have great difficulties discriminating in groups and with environmental noise and are practically isolated in group activities.[2–5] This was the case of our patient in public places, such as, bars or restaurants prior to her implant and now she has gradually recovered such capabilities.

Recovery of listening ability in groups and with ambient noise was also gradual over time. It took a year for these capabilities to improve. However, despite having done it a year after the implant was activated, the voices with the implanted ear were muted and different from the voices heard with the normal ear. The sound quality had improved quite a bit, but it was not like before and her effort to understand persisted.

It was only after 2 years of stimulation that her listening in groups and with ambient noise had improved to the point that she could not only understand well but also perfectly identify the voices of those who spoke. Two years after implant activation, the voices seemed to have a natural sound and there were no differences between the normal and the implanted ear. Also, just after 2 years, the sound quality recovered, and she could listen effortlessly. That is, after 2 years, she was able to recover binaural fusion.

The rationale for less auditory effort in binaural hearing is found in functional studies of central responses with auditory stimuli evaluated with neuroSPECT.[10,11] These studies have shown that stimulating both ears at the same time (binaural stimuli), the central areas that are stimulated and inhibited are the same than those with monaural stimulation. However, with binaural stimulation, there is less inhibition and stimulation less intense. In other words, binaural stimulation requires less perfusion and less effort by the central nervous system. This translates in less effort to hear and possibly in a better quality of hearing.

Finally, it is remarkable that after 2 years of stimulation with a cochlear implant, the patient recovered the binaural fusion and along with it the functioning neural circuits prior to her sudden deafness. This is probably due to the fact that the neural circuits were functional prior to the episode of sudden deafness and that in this blind patient these circuits were fully developed. This opens the door to the concept that under certain conditions it is possible to recover deficient neural circuits.

On the one hand, Sharma et al.[12] reported the case of a child with unilateral deafness who was implanted at 9 years and 8 months of age who gradually developed cortical development according to his age and discrimination and sound localization at 33 months post-cochlear implantation. This took our patient a period of 2 years. Even considering that these are very different cases, both had neuronal circuit recovery post-stimulation with a cochlear implant. On the other hand, Ramos Macías et al.[13] implanted children with unilateral deafness and noted that children with acquired unilateral deafness who are implanted early (and with a short period of auditory deprivation) recovered binaural benefits, the same as our patient.

Although these cases are few, they are suggestive that regeneration of neuronal circuits may occur when early stimulation with a cochlear implant is used in some cases of unilateral deafness.

This does not seem to be the case in congenital bilateral deafness with cochlear implants, cases in which symmetry can be achieved and auditory deficits can be significantly compensated; however, some cortical processing deficits seemingly persist.[14,15]

This is also true for children with normal hearing but with language deficits due to early sensory deprivation because of lack of stimulation. With sensory rehabilitation, these children can have significant improvements, however, some central deficits persist.[16] The same occurs with children with unilateral deafness that also have deficits in vocabulary due to lack of full central stimulation.[17] Neuroplasticity studies that define the different types and degrees of neuronal circuit deficits and the critical periods for their stimulation and recovery are lacking. They will be essential to answer these questions and to develop early treatment schemes for adequate recovery of deficient neuronal circuits.

Conclusion

Early stimulation with a cochlear implant allowed a complete recovery of previously deficient neuronal circuits.

The neural circuits that determine the origin of a sound source in terms of side or if it comes from above or below, near or far, or from ahead or behind are apparently different since they recovered at different times and independently of each other.

Hearing with both ears is essential for sound localization, discrimination in groups and with ambient noise.

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