Deafness and Musical Perception –

Remarks from a Medical-Neuroscientific Point of View

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1.1 Introduction

In my lecture, I will discuss what we know from a medical-physiological point of view about music perception (i.e. the totality from sensory understanding via analysis to (re)production) in people with deafness or profound hearing loss. The term deafness will be used in the following, even though the concept of complete deafness is controversial, especially in people with acquired hearing loss. In the case of congenital deafness the situation is different again. This has been studied much less in research, also in musical terms.

If one examines the scientific literature for studies on music perception and deafness, one first finds that there are only a few, predominantly speculative, essays and, beyond that, very few independent experimental studies. If one analyzes the medical database MEDLINE, a total of 702 papers with the keywords "deaf*" and "music*" have appeared since 1893 (as of November 2020), of which 279 alone deal with music perception after cochlear implantation, 128 with amusia, 58 with Ludwig van Beethoven, and 55 with musical hallucinations (see below). Only a total of 47 articles have been found on deafness (without hearing aids) and music. This is surprisingly few in view of the abundance of medical literature. Unilateral deafness plays only a minor role in this context since the complex bilateral wiring of the auditory pathways stimulates all critical areas equally and the network functions largely unimpaired.

In addition, the phenomenon of amusia/tone deafness must be differentiated. This is a cortical processing disorder of music without the hearing ability itself being affected. For example, those affected cannot understand music as such, cannot express themselves in music, cannot read or write notes, although they can imagine music, etc. Amusias are mostly acquired through a stroke or other focal lesions of the brain, but can also be congenital (especially in Asian countries) or caused by systemic neurological diseases. However, for amusia to be functionally equivalent to deafness is extremely rare and only possible when both hemispheres are affected by the lesion. However, since this phenomenon is not relevant to the issue of sensorineural deafness (i.e., the inability to adequately pick up sound waves, which is the focus here), it will not be discussed in detail.

Sociologist Harriet Martineau, who herself had a form of progressive deafness, was one of the first in the 19th century to describe the personal/social consequences of deafness in her 1834 essay “Letters to the Deaf.” In addition to social isolation, due to impaired communication, she described limited enjoyment of music as a major consequence of deafness (Naples et al. 2020). From today's perspective, several factors influence the extent and quality of “enjoyment” of music:

- Deafness or marked hearing loss
- Deafness acquired pre-lingually or post-lingually
- Unilateral or bilateral deafness
- Hearing with or without hearing aids
- Additional amusic disorder.

In the so-called “deaf community” (which encompasses a wide range of hearing impairments), engagement in music perception has become very popular, especially among younger members. In fact, the majority of those affected now report some kind of involvement with music (Darrow 1993). There are various strategies for doing this, such as composing music that contains primarily vibrotactile and visual elements, to vibratory perception of singing in real time. There are now discos for deaf people that work primarily with light effects and vibration. Deaf professional percussionists and composers, such as Evelyn Glennie (*1965), achieve high musical performances solely through the vibration sensation in different parts of the body and, thereby, also achieve artistic and societal acceptance.

1.2 Music and Deafness – Brain Physiology/Pathology

The following will summarize what we currently know about cognitive music processing in people with deafness. Then, a special focus will be on the phenomenon of musical hallucinations, as this is one of the best studied areas and points to mechanisms as they occur in people with deafness.

When the deafness is acquired also plays a role. For example, one study showed that among people with congenital deafness, emotional appraisal of music remains impaired despite hearing aids, such as cochlear implants, while among people with acquired deafness (but the same degree of hearing loss), this ability reaches approximately the level exhibited by people with normal hearing (Mazaheryazdi et al. 2018).

Functional imaging has shown that the auditory areas of the brain learn to respond more strongly to tactile stimuli. However, these changes are probably of very little functional relevance, i.e. many deaf people do not have a noticeably better tactile processing ability in everyday life. However, this *is* true for simple stimuli. For complex processing of tactile stimuli generated from music, there seems to be a demonstrable superiority in deaf people, especially for vibration. In a study on identifying the emotional content of music via tactile stimuli on the finger, deaf people were superior to hearing people in sensing "feelings of happiness" (Sharp et al. 2020). In rhythm recognition, vibrotactile stimuli have been used with deaf people. In this regard, one study has examined the extent to which vibrotactile stimulation leads to synchronization with the rhythm of dance music (Tranchant et al. 2017). Thus, thanks to vibrotactile stimulation, deaf people were able to match their dance rhythm to the original music with the same degree of accuracy as people with normal hearing due to auditory stimulation. By contrast, for people with normal hearing, the accuracy of rhythm matching was better with acoustic stimulation than with vibrotactile stimulation.

In so-called "captioning", the visualization of acoustic signals, e.g. in a video, the entire amount of acoustic information and especially the emotional connotation is naturally shortened and simplified. Nevertheless, this method works for people with deafness. In a study that primarily encoded the emotional content of music visually, people with deafness showed similar localization of brain activity when analyzing captioning compared to people with normal hearing, but the intensity of activation was significantly higher. Thus, increased activity was found in voluntary attention areas, and language-related cortical brain areas were also more active (Revuelta et al. 2020). This captioning is also used in virtual reality. In this context, a method has recently been developed in which directional hearing and distance from acoustic sources are imitated by tactile stimulation at the ear itself in people with deafness or profound hearing loss. In studies, this method showed better understanding of virtual reality in deaf people and they could solve virtual reality tasks in almost the same time as people without hearing problems (Mazaheryazdi et al. 2020).

Functional imaging studies have also been conducted on music processing in deaf individuals. This is based on the finding that after sensory deprivation of one modality (e.g., deafness, blindness, loss of sensitivity), the cortex can reorganize in such a way that the stimuli of the other modalities are processed differently/better. This is called cross-modal reorganization. In blind people this is manifested in multiple and well known ways e.g. improved hearing or improved tactility. In humans with deafness, improved visual perception can also be found phenomenologically, but in humans (by contrast to the hereditarily deaf cat) this could not be associated with cross-modal reorganization in the auditory cortex until recently. For example, functional magnetic resonance imaging in humans with early acquired deafness showed greater cortex thickness in the right temporal planum but not in the Heschl gyrus and Heschl sulcus (responsible for primary hearing), the middle temporal area MT+, and the sulcus calcarinus (responsible for primary vision) (Shiell et al. 2016). This thickness was then correlated with the ability to perceive visual movements quickly and correctly. This showed that this visual ability is related to the reorganization of the planum temporale, although the planum temporale otherwise performs almost exclusively tasks in auditory stimulus processing.

Thus, the right planum temporale has been identified as a region that reorganizes in acquired deafness and becomes more sensitive to visual stimuli. This plasticity supports compensatory mechanisms in response to visual stimulation. Thus, the enhanced ability to perceive visual motion in early deafened individuals correlates with thickened cortex in a portion of the right planum temporale. The white matter (i.e., subcortical portions of the planum temporale) was also examined in this context (Shiell and Zatorre 2017). This showed that only in the planum temporale, but not in other parts of the brain involved in auditory processes, increased activatability is correlated with the ability to perceive and classify visual movements.

In summary, then, the following plastic brain changes were found in people with acquired deafness (Good et al. 2014):

- improved perception of visual stimuli in the peripheral visual field
- increased attention to visual and somatosensory stimuli
- increased activity in the secondary auditory cortex, following visual and vibrotactile stimuli
- increased activity in visual cortex and middle temporal cortex during perception of movement (dance)
- increased activity after non-auditory stimuli in the areas of multimodal integration, especially in the superior temporal gyrus et sulcus temporalis and the planum temporale.

As stated above, people with a cochlear implant represent a special group in this context. Here, it has already been shown in many cases that the cochlear implant can not only improve the verbal communication ability, but also that the perception of other acoustic stimuli, especially music, becomes possible again to a certain extent. In this regard, the timing of the cochlear implantation and also the timing of the onset of deafness do not seem to play a major role in the perception and emotional processing of music in people with cochlear implants (Fuller et al. 2019). The differences between unilateral and bilateral deafness in cochlear implant recipients also play a minor role.

However, there is also a consensus in research that a cochlear implant will never allow "normal" music perception and that even in people with acquired deafness, a cochlear implant will never restore the musical abilities as they existed before the development of deafness, which is not only due to limitations of today's technology, but also due to the remodeling processes of the brain caused by deafness.

Nevertheless, studies have shown that musical training with cochlear implants can lead to improved acoustic stimulus processing beyond the musical level. The reduced access to complex acoustic stimuli such as music, environmental sounds or voice emotions can lead to a significant impairment of the quality of life. Several studies have, therefore, investigated how musical training can lead to an improvement of other perceptions in people with a cochlear implant. It was found that even after reaching a plateau of language acquisition in people with a cochlear implant, musical training can still lead to perceptual improvement in speech prosody and in the perception of emotions (Jiam and Limb 2020). Also noteworthy is another study that demonstrated that enjoyment of music listening in people with unilateral cochlear implants is greater when the normal ear and the ear with a cochlear implant are musically stimulated together, although musical presentation at the cochlear implant alone was found to be unpleasant. Apparently, binaural listening to music is associated with greater enjoyment than monaural listening, even when it involves the healthy ear (Landsberger et al. 2020).

1.3 Musical Hallucinations

1.3.1 Introduction and Epidemiology

Since musical hallucinations are a more common phenomenon in people with deafness than in people who can hear normally, it will be explained in more detail here. Musical hallucinations, as a specific subtype of auditory hallucinations, represent a disturbance in the complex processing of acoustic stimuli in which perception is shaped by instrumental music, sounds, or singing; perception is judged to be external (Berrios 1990; Cerrato et al. 2001). They are less common than unformed auditory hallucinations, such as certain forms of tinnitus. Typically, there is insight into the nature of the hallucination but no possibility of volitional influence. "Earworms" are distinguished from musical hallucinations as internal musical imaginings that arise involuntarily, but can be influenced volitionally and are not perceived as external perceptions (Moseley et al. 2018). The different mental manifestations of musical perception are shown in Table 1.

Table 1: Different types of mental perception of music.

1) Voluntary music imagination: active imagination of music.
2) Involuntary music imagination ("earworm"): music arises involuntarily in thoughts, but is recognized as internal and can be suppressed volitionally
3) Continuous involuntary music imagination: chronic earworms
4) Musical hallucinosis: long-lasting musical imagination in patients with organic diseases such as hearing loss (mostly without dissociation)
5) Musical hallucination: music is present as if it were really (i.e. externally) playing (mostly with dissociation).

Fukunishi et al. (1998) reported musical hallucinations in 0.16% of a large collective in a hospital. By contrast, in a collective of elderly people with hearing disorders, the prevalence of musical hallucinations ranged from 2.5% (Cole et al. 2002) to 3.6% (Teunesse et al. 2012). In another study in patients with hearing disorders, 16.2% had auditory hallucinations (of which voices 51%, music 36%, and ringtones 24%), with the prevalence of hallucinations increasing with the severity of the hearing disorder up to a maximum of 24% (Linszen et al. 2019); no age dependence was found, and women with left-sided hearing impairment were most affected. In purely mathematical terms, the risk for musical hallucinations increased by 1.02 per 1 dB of hearing loss. Musical hallucinations have also been described in healthy children and adolescents (Aziz 2009). The infrequent occurrence of musical hallucinations may reflect their natural occurrence, but it is likely that they are rarely reported.

Probably the first reports of musical hallucinations were published by Coleman in 1849 and by Baillarger in 1846 (Berrios 1990). The first scientific descriptions were made by Petazzi (1900) and by Bryant (1907). Whether musical hallucinations could be a source of creativity in composers (Gordon 1996) has been controversial. This has been suggested for Joseph Haydn (Gordon 1999) and Gaetano Donizetti (Peschel et al. 1992). Maurice Ravel, who had a diagnostically unexplained neurodegenerative disease in the last years of his life, said of himself that he still "had so much music in his head" but that he could not play or write it because he could no longer translate his thoughts into speech and actions (Alajouanine 1948; Alonso and Pascuzzi 1999). This may also have been a manifestation of a musical hallucination. Bedrich Smetana reported his own musical hallucinations in the form of two male voices in G major (Neumayr 1991). In addition, he had a pronounced hearing loss later in life, very likely due to syphilis (Feldmann 1964). He also experienced musical hallucinations in the form of an A-flat major chord, which he illustrated in the last movement of his second string quartet.

1.3.2 Review papers

Reviews of case reports with musical hallucinations have been published several times to date (Berrios 1990; Berrios 1991; Keshavan et al. 1992). One review found a female preponderance of 80% and a mean age of 60 +/- 19 years. 67% of patients were deaf, and musical hallucinations were the only symptom in 40%. In 26% there was a psychiatric history, mostly depression. In 39% of cases, there was evidence of brain disease, such as tumor, epileptic focus, and stroke (Berrios 1991). It was concluded that musical hallucinations are more common in older women with deafness or structural brain disease. They confirmed previous assumptions that deafness, ear disease, advanced age, drugs, and brain disease are important factors in the generation and persistence of musical hallucinations. Based on the cases with brain disease, it was concluded that involvement of the right non-dominant hemisphere of the brain is more common as an underlying etiology than involvement of the left hemisphere. In another review (Keshavan et al. 1992), three subgroups were analyzed separately: one with hearing loss; one with circumscribed brain disease; one with psychiatric disease. Among these, deafness was most associated with musical hallucinations; nonspecific brain damage also occurred frequently with musical hallucinations. A female predominance and a tendency to occur in middle to older age were described. In addition, mental disorders were an additional contributor to the development of musical hallucinations; thus, depression or psychosis may be associated with musical hallucinations without the presence of deafness or circumscribed brain disease. In this review, right-sided brain lesions were only marginally more frequently associated with musical hallucination than left-sided lesions.

In a separate review of cases with musical hallucination described in the literature, five subgroups of causes of illness were differentiated (Evers and Ellger 2004). In summary, data from 132 cases could be analyzed. In patients for whom more than one cause matched, the most probable one was chosen, and the other pathologies were designated as co-diagnoses. This means that whenever more than one of the five pathologies was found, the diagnosis most closely related to the musical hallucination was chosen.

Ages ranged from 20 to 90 with an average of 62 +/- 19 years. 30% were male, 70% female. A brain lesion or epileptic focus could be localized in 27% (20 cases right, 16 left). Generalized brain atrophy was described in another 26%. Handedness was reported in 23%; all were right-handed.

Hearing loss was the most likely cause of musical hallucination in 52 cases. Generalized brain damage was also described in 18. In addition, 11 patients had psychiatric comorbidity and eight showed epileptic activity clinically or on EEG. In another 29 patients, hearing loss was present, but the musical hallucination was probably caused by other pathologies. In 31 patients, the musical hallucinations were a concomitant phenomenon of psychiatric disorders (including 14 depression and 11 schizophrenia). Twenty-one patients had a focal brain lesion. In 13 cases, the lesion was located in the right hemisphere, and in eight cases, in the left. The brain lesions were vascular or tumor (six cases each), in two cases they were infectious, and in the remaining cases the lesion could not be precisely identified. Diffuse brain atrophy was described in 34 patients, with no reason given in most cases. In 15 patients, epileptic activity was thought to be the trigger for the musical hallucinations. In one third of the patients each, the epileptic activity was localized in the right hemisphere, in the left hemisphere, or diffusely. In another 15 cases, epileptic brain activity was described but not assumed to be the main cause of the musical hallucinations. Intoxication or brain inflammation was the main cause of musical hallucinations in 13 patients. Here, in particular, intoxication with drugs had occurred.

In 73 cases details about the contents of the musical hallucinations were found. 57 patients perceived familiar sounds. Only in five cases was the sound completely unknown. The contents ranged from religious songs to children's songs to unspecific radio music. Classical music and folk music were perceived rather rarely. Music was presented vocally, purely instrumentally, or mixed vocal/instrumental. Unilateral perception of the music was reported by ten patients, bilateral by 21 patients. The emotions evoked by the perceived music were pleasant in seven cases and threatening/unpleasant in 30 patients; in four cases the emotions were neutral, and in the remaining cases no information was found. Gender differences were not significant in the individual subgroups, but women were more frequently affected overall than men. This was most evident in the group of patients with hearing loss (77% versus 23%).

1.3.3 Analysis of the Literature

Patients with musical hallucinations perceive mainly familiar songs and melodies that they know from the radio, from religious contexts, and from childhood. This relatively consistent pattern is often also associated with personal memories of this music. Thus, these hallucinations often stem from individual memory traces. In older affected persons, religious songs therefore very often appear as musical hallucinations (Warner and Aziz 2005). This possible mechanism is also called "concept of parasitic memory". However, it is still completely unclear why such memory traces come to consciousness, apparently spontaneously, without any specific stimulation of the brain. Another mechanism discussed in the literature (Keshavan et al. 1992) is “neuronal irritation.” Hallucinatory perception can be understood as a construction of a re-experience of stored perceptions by the excitation of relevant neural networks (e.g., hallucinations induced by epileptic discharges in the cerebral cortex). Voices, often in combination with instrumental music, are more frequently observed as musical hallucinations than purely instrumental music. Even more often, musical hallucinations are perceived as frightening and negative rather than pleasant or positive or neutral.

No unified concept is yet available about the influence of laterality of brain lesions associated with musical hallucinations. Previously, the right, non-dominant hemisphere was thought to play an important role in the development of musical hallucinations (Berrios 1990 and 1991). Interestingly, auditory hallucinations in the elderly are often associated with asymmetric hearing impairment and incomplete suppression of sensory input from the left ear (Cole et al. 2002). This would be consistent with the right hemisphere often being dominant for music perception (Zatorre 1984), at least in non-musicians. On the other hand, it must be taken into account that right hemispheric lesions are clinically only slightly more frequently associated with musical hallucinations than left hemispheric ones. In summary, there is no clear evidence so far whether laterality of brain lesions or brain dysfunction plays a role in the development of musical hallucinations.

There are only isolated descriptions of successful therapy, from which no general recommendations can be derived. It is possible that increased external auditory stimulation may reduce the severity of persistent musical hallucinations (Collins et al. 1989). There is no known causal treatment for the symptom “musical hallucination.” When successful therapy has been described in case reports, it has tended to be the elimination of the underlying cause or the improvement of hypacusis. In depressed patients or patients with schizophrenia, hallucinations sometimes disappeared with antidepressant or neuroleptic medication. In individual cases, the following medications were successful: *fluvoxamine, clomipramine, olanzapine, quetiapine, carbamazepine,* and *valproate.* The latter two medications were prescribed for mood stabilization and not for therapy of epilepsy. More recently, there have been several reports of effective use of acetylcholinesterase inhibitors for musical hallucinations, even in patients who had no evidence of dementia.

1.3.4 Pathophysiological considerations

No clear mechanism can be deduced as to how musical hallucinations occur just by analyzing case reports. In fact, musical hallucinations seem to be caused by different mechanisms, so that it is not possible to speak of one entity.

Hearing loss is frequently reported in the context of deafness and seems to be the most important/common factor associated with musical hallucinations. Also, musical hallucinations are more intense when ambient sounds are quiet (Hammeke et al. 1983; Terao 1995). In this sense, musical hallucinations can be interpreted as a deafferentation phenomenon, by which one means sensations even though the afferent nerve fibers are cut off, as in phantom pain. Gordon (1997) even considered that musical hallucinations must be exclusively associated with an inner ear disorder leading to a "hyperactive state of the ear." Accordingly, patients with a combination of tinnitus and auditory hallucinations report predominantly musical hallucinations. However, there are clear cases without any evidence of hearing impairment in the middle or inner ear that have nevertheless exhibited typical musical hallucinations (Erkwoh et al. 1993; Evers et al. 2002). In this respect, disorders of the ear represent a common but not a necessary condition for musical hallucinations. Furthermore, musical hallucinations have been suggested as being similar to visual hallucinations caused by focal lesions in the occipital lobe, called Charles-Bonnet syndrome, i.e., that a disorder in the cerebral cortex underlies these hallucinations. Furthermore, sensory deprivation in the environment has also been hypothesized as a trigger for musical hallucinations.

Cerebral imaging studies of patients with musical hallucinations showed dysfunction of the temporal cortex (Erkwoh et al. 1993; Kasai et al. 1999; Tanriverdi et al. 2001), with the left temporal lobe more commonly affected than the right. There are also old reports of initiation of musical hallucinations by direct stimulation of the superior temporal gyrus (Penfield 1958; Penfield and Perot 1963). Therefore, it is possible that some of the musical hallucinations, especially those with a focal brain lesion or epileptic activity on EEG, represent focal epileptic seizures in the temporal cortex. Recent research using functional brain imaging confirms the important role of the temporal lobe in the generation of auditory/musical hallucinations, but also shows that other brain areas may be involved, particularly circumscribed areas of the frontal lobe. In summary, the superior temporal sulcus seems to be the most important/frequent brain area activated in musical hallucinations (Bernardini et al. 2017), but beyond that there are individually very different additional activations.

Another type of deafferentation phenomenon may lie in a lesion of the central acoustic pathways/radiation. When analyzing individual cases of musical hallucinations due to a focal brain lesion, many of these lesions are located in the feeder acoustic pathways or in the connection fibers between the primary acoustic cortex and the association cerebral cortices. Hemorrhage between the left corpus geniculatum and the left acoustic cortex and brainstem lesions in the lateral or paramedian pontine tegmentum (Douen et al. 1997; Schielke et al. 2000) or disruption of the basal ganglia (Wodarz et al. 1995) have been described. In summary, the pons was one of the most common brain areas associated with musical hallucinations.

Musical hallucinations cannot only be explained as a purely psychotic state, since actually a dissociation from it is always possible. However, in some cases they can also be described by the concept of obsessive-compulsive disorders (Matsui et al. 2003).

Previous work on musical hallucinations has also developed other theories of their origin. For example, a neuropsychological approach has been proposed to explain musical hallucinations by spontaneous and non-dynamic activity in a cognitive network module. Interestingly, PET failed to find activity in the primary auditory cortex during musical hallucinations, whereas it revealed activation of the Heschl gyrus during verbal auditory hallucinations (Dierks et al. 1999). The brain areas involved in the module for generating musical hallucinations were the posterior temporal lobe, right basal ganglia, cerebellum, and inferior frontal lobe. However, these findings cannot explain how the spontaneous activation of this network occurs in the first place.

Disturbances of learning or memory must also be considered in explaining musical hallucinations. This has led to the concept of “parasitic memory” (Keshavan et al. 1992). This means that musical hallucinations in most cases represent an unlearned musical perception and are thus an autonomous, i.e., non-modifiable, memory function that can be retrieved purely by chance but also by external stimulation. Interestingly, almost all musical hallucinations consist of very familiar musical sequences and not of rarely heard or unknown sequences.

In summary, there are no biological findings to date that can fully explain the pathophysiology of musical hallucinations. Nevertheless, theories exist that can describe the various peculiarities of musical hallucinations. Most cases correspond to the concept of deafferentation phenomenon, which is also known from visual perception. In a smaller proportion of cases, measurable epileptic or dysfunctional activity in specific brain areas may be associated with musical hallucinations. Finally, the concept of activation of memory dysfunction, called “parasitic memory,” may also be used to explain musical hallucinations. In this respect, it is rather unlikely that musical hallucinations can contribute to the generation of compositional ideas.

Translated from the German by Elisabeth Hormann

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