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Auditory agnosia for environmental sounds in Alzheimer's disease: Effects on daily life

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ABSTRACT

Background: Auditory agnosia for environmental sounds is a type of agnosia attributed to central auditory dysfunction. It is common in Alzheimer's disease, and is associated with peripheral hearing loss, although independent of it, and presumed independent of language deficits. The effects of this type of agnosia on daily life in Alzheimer's disease are unknown.

Objective: We aimed to assess the impact of auditory agnosia for environmental sounds in people with Alzheimer's disease while also exploring the role of unrecognized hearing loss.

Methods: We tested 34 home-dwelling people with Alzheimer's disease and a mean MMSE of 21.9 with the aid of a sound naming and recognition test, the tailor-made EESAA (*Experiencing Environmental Sounds in Auditory Agnosia*) questionnaire, the ADQRL (*Alzheimer's Disease-Related Quality of Life*) scale, and speech and tone audiometry.

Results: Some 57 % of our 34 participants showed clinical signs of auditory agnosia for environmental sounds, and 47 % had undetected hearing loss to such an extent that it made them eligible for a hearing aid. Although the two factors appear to be independent, their joint effect can impact people's daily functioning. Nonetheless, we found them to have only little impact on the participants' quality of life as measured by the ADQRL, possibly because most of them lived in a sheltered environment, and some moreover showed anosognosia for their agnosia.

Conclusion: Difficulties recognizing environmental sounds in daily life are very common in people with Alzheimer's disease. Although we found no direct relation with quality of life as measured by a questionnaire, awareness of auditory agnosia for environmental sounds is still important since it may help explain why function declines. The additional finding that 47 % of people in this group had unrecognized hearing loss shows that self-assessment of hearing is often inaccurate in Alzheimer's disease, with implications for daily practice where clinicians might only explore hearing loss when acknowledged by their patient. On the basis of our findings we advise further longitudinal, multi-year studies of hearing screening and rehabilitation in Alzheimer's disease, if possible starting during its prodromal stage, something supported by findings in a large trials suggesting that hearing interventions might be slowing cognitive decline in an older population at risk of this.

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Introduction

We humans have an uncanny ability to grasp the meaning of environmental sounds. Footsteps in a hallway, oncoming traffic, water coming to a boil, these are only a few of the myriad sounds that we recognize in a heartbeat. In Holland, people can put this ability to the test by participating in the popular radio programme *What's the Sound* where the producers present an exotic sound and offer callers the opportunity to guess what it is. The game is played for laughs and for a modest financial reward (for those who guess correctly), but in fact it capitalizes on our brain's capacity to quickly disambiguate environmental sounds. It also confronts us with the limits to that capacity, and in that sense may aid to empathize with people who suffer from a condition called *auditory agnosia for environmental sounds*. This type of agnosia is characterized by the inability or strongly diminished ability to make sense of sounds despite hearing them. It is also independent of deficits in the processing of spoken language (Polster and Rose, 1998), although combinations do occur in clinical practice. Auditory agnosia for environmental sounds is conceptualized as an auditory processing disorder and attributed to cortical dysfunction. In this context environmental sounds are defined as sounds produced by events that take place in the world around us (Ballas and Howard, 1987), or, alternatively, as 'all naturally occurring sounds other than speech and music' (Gygi et al., 2007), which has the advantage of including sounds emanating from within the body. Environmental sounds are dynamic in that they can convey action and movement-related information, i.e. 'news that something is happening' (Jenkins, 1985). From an evolutionary and a practical point of view, the capacity to make sense of such sounds is of utmost importance: to warn of danger (e.g., car horn, rattlesnake), to signal presence (e.g., humming, bicycle bell), to indicate the correct or incorrect functioning of devices (e.g., clicking of a stapler or water dripping), to indicate food crispness (e.g., crunch of celery), to locate and orient to an event (e.g., an explosion to the right), to monitor change in status (e.g., chiming of a cuckoo clock), to communicate information about emotional (e.g., scream) or physical (e.g., a burp) states, and so on. In short, our uncanny ability to grasp what we hear is more than just a gimmick that allows us to play games. On the contrary, it is an evolutionary-driven ability without which the world around us loses much of its significance and becomes substantially more dangerous.

Relation with dementia

Auditory agnosia for environmental sounds is primarily associated with Alzheimer's disease and other types of dementia (Rapcsak et al., 1989; Eustache et al., 1995; Coebergh et al., 2020). Rapcsak et al. studied 18 people with Alzheimer's disease who had a mean score of 14 on the *Mini-Mental State Examination* (MMSE). They asked these participants to match sounds to pictures, offering them four to choose from. They thus found frequently impaired accuracy with almost no mistakes in a group of healthy controls (Rapcsak et al., 1989). Eustache et al. compared 15 people with Alzheimer's disease (with a mean score of 21 on the MMSE) to controls while studying their ability to match sounds to written descriptions thereof, also offering them four possibilities to choose from. In the patient group they found a significant impairment in the recognition of environmental sounds, but no relation to the participants' MMSE scores (Eustache et al., 1995). Brandt et al. found a poor performance in 28 people with Alzheimer's disease in an environmental sound naming and picture recognition test. In their case, this was strongly correlated to lower MMSE scores (Brandt et al., 2010), something that was also found by Dietz et al. in a group of 18 patients with either mild cognitive impairment, Alzheimer's disease, or frontotemporal dementia (Janine Diehl-Schmid Birgit Dietz et al., 2017). Our group (Coebergh et al., 2020) explored the neurological basis of auditory agnosia for environmental sounds, and found that hearing loss is a significant risk factor for its development in the context of Alzheimer's disease. We also found that self-reporting on hearing loss is often inaccurate in this group, which is in line with an older study by Gold et al (Gold et al., 1996). In the present study, we seek to expand these insights by assessing the effects of this type of agnosia on people's daily lives, taking into account the role played by risk factors such as hearing loss and lack of insight.

Clinical relevance

The impact of auditory agnosia for environmental sounds on activities of daily life (ADL) and quality of life (QOL) is potentially substantial. That said, in people with Alzheimer's disease the impact of auditory agnosia has never been studied systematically. Even case reports on this topic are rare. In several of these - including a novel - people struggle to distinguish between the sound of their telephone and that of the doorbell (Uttner et al., 2006; Marshall et al., 1985; Bernlef, 1989). In another rare study, Saygin et al. describe a man who recognized less than a quarter of environmental sounds due to a temporal lobe stroke. During testing he often replied, 'All I know is there was sound there'. Nonetheless he did not seem to have any severe functional impairments. The authors proposed that their patient probably compensated for his handicap by making clever use of contextual cues and information from the other sensory modalities (Saygin et al., 2010). Another reason why people may seem to tolerate their condition rather well is anosognosia for their agnosia (Klarendić et al., 2021). It is known that agnosia in general can affect QOL in unexpected ways; however, caregivers will often testify that the dictum 'ignorance is bliss' only goes so far, since anosognosia is associated with a greater discrepancy between patient and caregiver ratings of QOL, and with greater caregiver burden. Taking these different viewpoints into account, it is important to understand how QOL in Alzheimer's disease is operationalized. Lawton's general model of QOL has been influential in the conceptualization of QOL in dementia, defining QOL as 'a multidimensional construct that should include not only objective (observable) indices of well-being judged against socio-normative criteria, but also the individual's own subjective perception of his or her position in life.' Lawton also identified four broad dimensions that contribute to QOL: (Polster and Rose, 1998) psychologic well-being (e.g., positive and negative affect), (Ballas and Howard, 1987) behavioural competence (e.g., cognitive and functional abilities), (Gygi et al., 2007) objective environment (e.g., caretakers and living situation), and (Jenkins, 1985) perceived QOL (Lawton, 1997; Powell Lawton, 1994). In the present study we use these dimensions (with the exception of psychological well-being) to explore the effects of auditory agnosia for environmental sounds on QOL. In addition, we assess the possible impact of hearing loss on QOL in this context (Park et al., 2016).

Materials and methods

We draw on the data that we collected in 34 people diagnosed with mostly mild (based on mean MMSE 21 (range 10–26)) probable Alzheimer's disease, as reported on in Coebergh et al (Coebergh et al., 2020). Inclusion criteria for participation in this and the present study were: age $\geq$ 55 years, being home-dwelling, and having a diagnosis of probable Alzheimer's disease based on NINDS-ADRDA criteria (McKhann et al., 1984), with participants needing to be Dutch- or English-speaking. All participants were identified through the outpatient memory clinic of the Haga Hospital, The Hague. The diagnosis was made by a neurologist with experience in dementia, based on clinical assessment, neuropsychological testing, brain MRI (1.5 T unit (Philips, Best, the Netherlands)), electroencephalography (20-minute EEG with NicoletOne (CareFusion), analyzed with Studyroom), and laboratory testing for renal and liver function, full blood count, thyroid tests, electrolytes, ESR, and Hba1C. A lumbar puncture was only performed on indication ($n = 6$, all consistent with Alzheimer's disease). Each participant also received neuropsychological testing with (informant) history and clinical examination. This involved cognitive, memory, and language screening, executive function tests, and tests of working memory, attention, and concentration. Tests included (Dutch translations of) the MMSE (Folstein et al., 1975), the *Kaufmann-Short Neurological Assessment Procedure* (K-SNAP, a test for general cognitive abilities), the *Verbal Learning and Memory Test*, the *Kaufmann Adolescent and Adult Intelligence Test* (KAIT), the *Word Fluency Test*, *Ruff's Figure Fluency Test*, the *Boston Naming Test* (BNT), the *Number Series Test*, the *Modified Card Sorting Test*, the *Incomplete Letter Test*, the *Reverse Digit Span*, the *Visual Recognition Test* (Warrington and James, 1991), and the *Sound Naming and Recognition Test*. The study was approved in 2008 by the Local Research Ethics Committee of the Haga Hospital, The Hague (number: 08 – 098), and considered exempt from independent ethics review. It was carried out in accordance with the revised conditions of the Declaration of Helsinki.

Participant consent

After diagnosis, the participants and one of their caregivers were approached. Together, they provided written informed consent.

Sound naming and recognition test

For the present and our previous study, borrowing from Marcell et al. (2000) (Marcell et al., 2000), we created a sound naming and recognition test with 24 sounds appropriate for Dutch culture, and with high accuracy and familiarity for matched controls. To increase task difficulty and investigate naming ability, we included a test in which the participant had to name a sound without receiving any other cues. To resemble daily life more closely, we then provided multiple cues consisting of written words and three pictures of similar items, either semantically and/or acoustically. Sounds were played in a quiet environment at a volume of 75 dB SPL. When there was hearing loss, they were amplified in 5 dB SPL intervals until the participant confirmed they could hear a sound and/or their hearing aid was adjusted. None of the participants complained that sounds were too quiet to hear. For further details regarding the sounds used, the types of test and their limitations, see (Coebergh et al., 2020).

Assessment of environmental sound recognition in daily life

To assess the phenomenology of the behavior of people with Alzheimer's disease in response to environmental sounds, we used the *EESAA Questionnaire* (Dealing with Environmental Sounds in Auditory Agnosia), a tailor-made, non-validated semi-structured questionnaire, developed after prior testing and consultations with local dementia specialists. This is a 12-item questionnaire that focuses on such diverse issues as the way people deal with these sounds in daily life, ENT visits, perceived hearing loss, tinnitus, auditory hallucinations, and auditory localization difficulties (see Supplementary Material).

QOL

To assess QOL, we used the caregiver-rated version of *Alzheimer's Disease-Related Quality of Life* (ADQRL) 40-items scale, which allowed us to also explore people's social interactions and their responses to their surroundings (Rabins et al., 1999).

ENT examination

All patients received a clinical ENT examination. Speech and tone audiometry was performed in a soundproof room with the aid of an audiometer AC40200 (Emid, Doesburg, the Netherlands) through headphones (Telephonics C69313) and bone conduction (RadioEar B-71). For analyses, the worst score of bone or air conduction was used. The main parameter used was pure-tone audiometry (PTA) at 0.5, 1, 2, and 4 kHz (PTA4). Hearing loss was rated in accordance with WHO criteria as: none (0–25 dB), slight (26–40 dB), moderate (41–60 dB), severe (61–80 dB), or profound (>=81 dB). In addition, we analyzed the PTA3 (average of 0.5, 1, and 2 kHz) and the Fletcher index (average of 1, 2, and 4 kHz).

Statistical Methods

For our analyses we used *Stata Statistical Software: Release 17* (SPSS 17.0, StataCorp, 2021). We used the *t*-test for independent samples to compare characteristics between participants with and without auditory agnosia for environmental sounds. Analyses were repeated while considering several covariates, using ANOVA and ANCOVA. Most data were normally distributed. Upon inspection of a histogram of the unadjusted residuals, no profound deviations from normality were seen. If data was not following the normal distribution after being transformed, we performed the Mann-Whitney *U* test. We used the chi-squared test and the Fisher's exact test to assess categorical data, with a significance level set at.05. Many of the factors analyzed were highly correlated. Therefore it was deemed inappropriate to correct p-values for multiple comparisons (for example, by a Bonferroni correction) since (Polster and Rose, 1998) this would increase type II errors, (Ballas and Howard, 1987) there was no 'universal null hypothesis', (Gygi et al., 2007) it was not imperative to avoid type I errors, and (Jenkins, 1985) there were not a large number of tests carried out without pre-planned hypotheses (Knudson and Lindsey, 2014).

Results

Of the 34 people included in this and our previous study (Coebergh et al., 2020) (Coebergh et al., 2020) all completed all tests analysed in this study. Demographic data and MMSE scores can be found in Table 1.

Sound naming and recognition

Table 2 shows the results for testing on auditory agnosia for environmental sounds, while Table 3 summarizes the number of errors made per sound category. Overall, participants had the most difficulty recognizing bagpipes (51.6 % making an error), the sound of the telephone ringing, ducks quacking, and harmonica sounds (for each of these, 48.4 % made an error). Musical instruments were most difficult to *name*, with people often answering along the lines of, 'It's a musical instrument, but I don't know which one'. Most difficult to *identify* were the telephone ringing (38.7 % wrong) and ducks quacking (32.3 % wrong). Two participants gave a sound the name of the previous sound (perseverative error), even though objectively, it was very different. Two others sometimes copied the sounds presented rather than naming them (imitation error). Five participants copied sounds played to them. We interpreted this as signs of executive dysfunction, which is common in Alzheimer's disease.

Dealing with environmental sounds in daily life

We here describe incidences of likely impact on daily life identified on the *EESAA Questionnaire*. A total of nine participants (27 %) did not answer the phone themselves anymore when they did use to. One of

Table 1
Demographics and MMSE scores.

	Participants
Total number	34
Sex (%)	63 % female
Age range	59–88 years
Mean age (SD)	74 (± 7) years
Mean MMSE score (SD)	21.9 (± 3.5)
MMSE range	10–26

Table 2
Test results for auditory agnosia.

Sound naming and recognition scores	Participants (n, %)
Sound Naming < = 83.3 % (20/24 points)	22 (65 %)
Sound Naming Mean (SD)	73.8 % (± 16.7 %)
Sound Naming Range	37.5–100.0 % (9–24 points)
Sound Recognition < = 87.5 % (21/24 points)	12 subjects (35 %)
Sound Recognition Mean (SD)	91.3 % (± 9.2 %)
Sound recognition Range	54.2–100.0 % (13–24 points)

Table 3
Total and mean errors made per sound category in those struggling with Agnosia for Environmental Sounds (AES) naming and recognition on AES test.

	In people with AES Naming problem	In people with AES Recognition problem
Animal sounds (n = 5)	32 (6.4)	16 (3.2)
Musical instruments (n = 5)	63 (12.6)	12 (2.4)
Animate (n = 4)	16 (4)	9 (2.25)
Inanimate (n = 10)	76 (7.6)	30 (3)

Legend: n = number of sounds presented per category)
Mean in (.) can be higher than total sounds since multiple attempts allowed

them said that his wife had always been the one at home who answered the phone, and another one that she was too insecure. Of the 17 participants who named the telephone sound incorrectly, 12 (71 %) did answer the phone at home. Of the 11 who did not recognize the telephone sound, nine (82 %) answered the phone at home. Two participants confused phones with doorbells. One of them said that she often goes to the front door, but does not see anyone, or answers the phone while nobody is calling. This can be interpreted either as agnosia or as an indication of nonverbal auditory hallucinations. One person got a louder doorbell, but it did not help with recognizing it. An audiologist found normal hearing. One of the participants answers the door and says, 'Miss A speaking'. Another one thinks the phone ringing is music playing. He moreover does not hear the doorbell. Yet another one describes not recognizing the doorbell when there are several people in the room, which we attribute to a lack of attention (on testing, the person did not have auditory agnosia for environmental sounds). Interestingly, two out of seven people with reported problems in identifying the sound of the telephone or the doorbell in daily life had no auditory agnosia for environmental sounds on testing. Two out of seven recognized problems themselves (both with auditory agnosia for environmental sounds) whereas five did not. In all, we found clinical auditory agnosia for environmental sounds to be marginally more common in people with self- or caregiver-described problems (72 %) than in the total group of people with Alzheimer's disease (57 %). Upon testing, this difference was not significant. Three other participants could not use the telephone anymore for other reasons (e.g. they picked up the remote control instead or had difficulty with the buttons). Another person suggested she did not answer the phone out of apathy. Two participants (without peripheral ENT pathology) noticed difficulty locating the direction of sounds. Three people said they sometimes got sounds mixed up. Three others said they never confused sounds, whereas their partners said that they did. There was one more discrepancy between patient and partner, with the patient saying he answered the phone and the partner denying this. So, clearly discrepancies exist, but there was no evident relation to the scores on testing for auditory agnosia for environmental sounds. Six people still drove a car, of which three had auditory agnosia for environmental sounds. None of the participants had had a car accident in the past five years. Sixteen participants still rode a bicycle. There were no accidents reported that were related to misinterpreting sounds.

QOL

We found no significant differences in the ADRQL scores for participants with and without auditory agnosia for environmental sounds (Table 4).

Hearing loss

Speech and tone audiometry yielded the following results: 16 (48 %) people showed virtually no hearing loss (0–25 dB), 14 (42 %) slight hearing loss (26–40 dB), three (10 %) moderate hearing loss (41–60 dB), and no one severe or profound hearing loss. These levels of hearing loss did not interfere with any of the other tests presented. Five participants disagreed with their partner on whether they had hearing problems, with one of them claiming to have problems while the partner denied that. People with auditory agnosia for environmental sounds had a tendency not to be aware of their hearing loss, but numbers were too small for further analysis.

Other auditory symptomatology

Six participants heard sounds that were not real, of whom five had auditory agnosia for environmental sounds; one heard his deceased wife, one people walking outside, two others unknown people in the house, and one noises upstairs. Also, six people reported tinnitus. However, their mean naming and recognition scores did not differ significantly from those of participants without tinnitus.

Discussion

As reported in Coebergh et al (Coebergh et al., 2020)., in our group of 34 home-dwelling people with Alzheimer's disease and a mean MMSE of 21.9, more than half of the participants showed signs of auditory agnosia for environmental sounds on a validated test: 37 % for recognizing sounds correctly, and 67 % for naming them correctly, whereas controls rarely make any mistake. The present study shows that those who reported problems in daily life tended to have higher scores on tests for this type of agnosia. The other way around, we did not find a clear relation between this type of agnosia and self- or caregiver-reported *problems* with environmental sounds. This may be since the majority of the participants lived in a supported environment, where the wrong identification of sounds does not tend to have grave consequences, and potential hazards are often detected in time by caregivers. So despite the relatively high prevalence of auditory agnosia, practical consequences tended to be mild in this group, even for those who still got into traffic.

Table 4
ADRQL data Social Interaction (A), Awareness of Self (B), Feelings and Mood (C), Enjoyment of Activities (D), and Response to Surroundings (E); percentage correct of total 40 questions; 100 % is no problem. Difference between those with AES naming and AES recognition and those without.

	Median, 25th, 75th percentile				
	AESn+		AESn-		P
A	100 %	92.6, 100 %	100 %	86.1, 100 %	0.325
B	90.7 %	87.4, 100.0 %	90.7 %	63.8, 90.7 %	0.241
C	100 %	94.2, 100 %	94.6 %	92.6, 94.6 %	0.164
D	100 %	78.0, 100 %	100 %	63.2, 100 %	0.855
E	100 %	100 %	100 %	100 %	0.638
Total	95.7 %	93.3, 98.4 %	96.6 %	79.8, 98.4 %	0.619
	Median, 25th, 75th percentile				
	AESr+		AESr-		P
A	100 %	92.3, 100 %	100 %	91.7–100 %	0.794
B	90.7 %	90.7, 100.0 %	90.7 %	82.8, 100 %	0.180
C	100 %	94.0, 100 %	100 %	93.1, 100 %	0.648
D	100 %	64.2, 100 %	100 %	78.0, 100 %	0.587
E	100 %	90.7, 100 %	100 %	100 %	0.322
Total	95.1 %	92.9, 98.4 %	96.7 %	92.1, 98.3 %	0.910

The lack of accidents being reported may be because sometimes all we need to detect is something loud coming our way, without knowing exactly what it is. Moreover, in real life additional cues are often available, such as contextual ones and deductive ones. We did not find any significant differences between sound categories, although sounds with a melodic quality (doorbell, telephone, bagpipes, other musical instruments) were mistaken somewhat more frequently than others. The finding that agnosia for musical instruments did not seem to be problematic is probably explained best by the fact that music rarely serves as a sign of imminent danger. We did not investigate higher-order semantic features such as causal ambiguity, ecological frequency, or importance to the listener. This could be relevant since one's own phone or doorbell, for example, has a higher familiarity (and importance) than a test one, and familiarity and importance may influence how well people with Alzheimer's disease do on this test (Roye et al., 2010). Evidence for the importance of context for identification comes from Gygi and Shafiro, who found that sounds embedded in an inappropriate context (e.g. the sound of a horse galloping in a restaurant) are identified better than in an appropriate context (e.g. a dog barking in a playground) (Gygi and Shafiro, 2011). And yet, in general, commonly encountered sounds are more easily identified. Also, hearing a sound can facilitate the identification of a subsequent sound that is related (Ballas and Howard, 1987; Shafiro et al., 2016). It would therefore be interesting to study how priming affects recognition of environmental sounds in Alzheimer's disease. Another topic for future study is the issue of whether more recently acquired memories of (personally) meaningful sounds, like a ringtone,('auditory templates'; (Griffiths and Warren, 2002) are lost sooner than those acquired earlier in life (e.g. the sound of a bicycle bell, or of a baby crying). We also did not consider how problems with sound *localization* affect daily life. Sound localization can be deficient in people with Alzheimer's disease, and this could influence QOL measures (Kurylo et al., 1993).

Other auditory phenomena

It might also be interesting to explore whether (environmental) sound identification problems correlate to positive auditory phenomena such as auditory hallucinations and tinnitus. In our study, five out of six participants with (possible) nonverbal auditory hallucinations had auditory agnosia. These numbers are too small to allow for any conclusions, but severe hearing loss does predispose towards psychotic symptoms and delusions in dementia (Ballard et al., 1995) and auditory hallucinations correlate to severity of hearing loss (Linszen et al., 2018). Of our participants, 18 % had tinnitus (severity not specified), and according to caregivers 24.2 % were frightened by sounds (considered indicative of hyperacusis). Nondahl et al. report a tinnitus prevalence of 8.2 % in an elderly population (defined as moderate in severity or causing difficulty falling asleep) (Nondahl et al., 2011), and in a cohort of people with semantic dementia, 32 % had either hyperacusis or tinnitus, which was suggested to be due to functional and structural brain network changes characteristic of the underlying condition (Mahoney et al., 2011).

Hearing loss and hearing aids

Since 47 % of the participants to our study showed unrecognized hearing loss, one might be tempted to think that that would explain the relatively high rate that we found for auditory agnosia. However, since we made sure to present sound fragments at levels appropriate for each participant, that conclusion cannot be drawn. That said, in real life, hearing loss can be expected to lead to even higher rates of misinterpreted sounds than can be attributed to agnosia alone. In the general population, hearing loss is found in 27 % of the elderly with a mean age 78 years (George et al., 2003). The rate that we found is not as high as that reported on by Gold et al. in people with Alzheimer's disease (92 %), but we used a more practical definition, i.e. eligibility for a hearing aid rather than > 25 dB loss at 1, 2, or 4 kHz, which has no clear clinical consequences. In our study, of those eligible for a hearing aid, 21 % did not have one. Like Gold et al., we found two types of anosognosia for hearing loss, in the sense that some participants thought they had a hearing problem when they did not, and, more commonly, that they did not think they had a hearing problem when they did. In the literature on Alzheimer's disease this is attributed to a loss of self-monitoring (Gold et al., 1996). Mild hearing loss (25–40 dB in either ear) in older adults is associated with reduced QOL, with a clear relationship between severity of hearing loss and QOL (Dalton et al., 2003). In such cases a hearing aid may be of help, although for people with Alzheimer's disease this is uncertain since previous studies on this topic are inconsistent. For example, a multicentre, double-blind, randomized, placebo-controlled trial of hearing-aid use in people with Alzheimer's disease, with an average age 83 years, showed no significant effects after six months, although declines in both groups were smaller than expected (Nguyen et al., 2017). At 12 months follow-up the hearing-aid group did significantly better on the ADRQL, although not on other QOL tests. The study also demonstrated that many people do not persist with wearing hearing aids (Adrait et al., 2017). In another study, Mamo et al. reported on communication techniques and over-the-counter amplification devices for people with hearing loss and dementia, and found a caregiver-reported reduction in depression and neuropsychiatric symptoms for people with high symptom scores at baseline (Mamo et al., 2017). The question whether treating hearing loss early reduces the risk of auditory agnosia in people with Alzheimer's disease much later is an interesting one, especially since we found the severity of hearing loss to be a major risk factor (Coebergh et al., 2020).

Limitations

The present study has several limitations. First, our sample size was relatively small. Consequently, we were unable to analyze in detail differences between those with and without auditory agnosia for environmental sounds regarding issues such as anosognosia for hearing loss and other complex auditory phenomena. Secondly, we did not correct for variables that Léon-Salas et al. found to influence QOL subscales in Alzheimer's disease as assessed with the aid of the ADRQL, such as household income (lower response to surroundings), instrumental ADL (less awareness of self), mood, caregiver caring for another dependent person (higher social interaction), and caregiver burden (worse feelings and mood) (León-Salas et al., 2011). Thirdly, likely by studying an outpatient group attending a neurology clinic with caregivers we found baseline high rates of quality of life on the ADQRL, which likely reduced the chance of finding differences on this scale related to the symptoms described (and/or alternatively scales like this are not sensitive to picking up impact of individual symptoms of agnosia).

Fourthly and last, we did not use the *Hearing Handicap Inventory for Elderly - Screening* (HHIE-S) questionnaire. This could have been useful, since the HHIE-S focuses more on emotional and social problems because of changes in hearing (embarrassment, frustration, etc.) whereas our tailor-made *EESAA Questionnaire* looks more at functional limitations.

Conclusions

Our ability to recognize environmental sounds is more than just a parlor trick that allows us to be successful at playing sound-guessing games. It is a hard-wired, evolutionary driven capacity that increases our well-being and our chances of survival. In dementia, this ability may decrease independently of hearing and linguistic capacities. Thus, in the group of 34 people with Alzheimer's disease here described, with a mean MMSE of 21.9, some 50 % showed signs of auditory agnosia for environmental sounds. Practical consequences and impact on QOL were negligible, but this was probably mainly since the participants led rather sheltered lives, had few responsibilities, and could count on caregivers to keep their affairs in order, with resultant high QOL scores. Nonetheless, awareness of auditory agnosia for environmental sounds is still important for people with Alzheimer's disease and their caregivers since it may help explain why levels of functioning decline. An additional finding here presented is unrecognized hearing loss in 47 % of the group under study, to the extent of eligibility for a hearing aid. From this we conclude that self-assessment of hearing loss was often inaccurate in this group. This may have important implications for daily practice where clinicians might only explore hearing loss when their patient acknowledges this or asks for a test. Based on our findings and previous studies aimed at linking hearing and QOL in Alzheimer's disease, we advise longitudinal, multi-year studies of hearing screening and rehabilitation, also in prodromal stages of the disease.

Author contributions

J.A.C. contributed to the conception and design of the work, collected data for the work, contributed to the analysis and interpretation of data for the work, drafted and revised the work, gave his approval for the final version to be published, and agreed to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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Declaration of Competing Interest

We confirm that all authors of this paper had access to all study data and are responsible for the contents of this article. All authors contributed and had authority over manuscript preparation, as well as approved of the submission of this manuscript. We confirm that this is an original paper, which has not been published, nor is it currently under consideration for publication elsewhere. We have no conflicts of interest to disclose.

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Appendix A. Supporting information

Supplementary data associated with this article can be found in the online version at doi:10.1016/j.ibneur.2025.01.006.

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