



Music, memory, and Alzheimer's disease: is music recognition spared in dementia, and how can it be assessed?

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Summary Despite intriguing and suggestive clinical observations, no formal research has assessed the possible sparing of musical recognition and memory in Alzheimer's dementia (AD). A case study is presented of an 84-year old woman with severe cognitive impairment implicating AD, but for whom music recognition and memory, according to her caregivers, appeared to be spared. The hypotheses addressed were, first, that memory for familiar music may be spared in dementia, and second, that musical recognition and memory may be reliably assessed with existing tests if behavioral observation is employed to overcome the problem of verbal or written communication.

Our hypotheses were stimulated by the patient EN, for whom diagnosis of AD became probable in 2000. With severe problems in memory, language, and cognition, she now has a mini-mental status score of 8 (out of 30) and is unable to understand or recall standard instructions. In order to assess her music recognition abilities, three tests from the previous literature were adapted for behavioral observation. Two tests involved the discrimination of familiar melodies from unfamiliar melodies. The third involved the detection of distortions ("wrong" notes) in familiar melodies and discrimination of distorted melodies from melodies correctly reproduced. Test melodies were presented to EN on a CD player and her responses were observed by two test administrators. EN responded to familiar melodies by singing along, usually with the words, and often continuing to sing after the stimulus had stopped. She never responded to the unfamiliar melodies. She responded to distorted melodies with facial expressions – surprise, laughter, a frown, or an exclamation, "Oh, dear!"; she never responded in this way to the undistorted melodies. Allowing these responses as indicators of detection, the results for EN were in the normal or near normal range of scores for elderly controls. As well, lyrics to familiar melodies, spoken in a conversational voice without rhythmic or pitch clues, often prompted EN to sing the tune that correctly accompanied the lyrics.

EN's results provide encouraging support for our hypotheses that sparing of musical memory may be a feature of some forms of dementia and that it may be reliably and quantitatively assessed through behavioral observation. The contrast between EN's response to music and her mini-mental status is dramatic.

The article concludes with several considerations why music may be preserved in dementia and suggestions to guide future research.

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Introduction

The powerful methods of cognitive neuropsychology have revealed much about the functional architecture of the brain through case studies of both the sparing and the loss of musical abilities following brain damage. For example, studies have demonstrated dissociations between music and language abilities: on the one hand, language skills may be impaired with musical abilities spared (aphasia without amusia); on the other, musical abilities may be impaired with language skills spared (amusia without aphasia). At a finer grain of analysis, the various components of music itself – such as the melodic versus the temporal dimensions – may be selectively spared or lost. For collective summaries of this research, see [1–3].

In contrast, the assessment of musical skills in dementias such as (probable or possible) Alzheimer's disease (AD) has received comparatively little study. Because cognitive research in dementia faces additional challenges, it is perhaps not surprising that research findings are scarce. First, the diffuse location of specific brain pathology and the multiple domains of cognitive deficit make it difficult to predict and to account for patterns of loss and sparing of function. Second, the progressive nature of the disease means that the encephalopathy is rarely stable, implying little time for a reliable and extensive assessment of a given state. Third, current tests and procedures in music perception and cognition – those that have been employed with brain damaged patients – require fairly intact memory and cognitive processing skills to follow test instructions. They seem not to be suited to the testing of dementia.

Yet, despite the scarcity of research findings, considerable anecdotal evidence from caregivers and family members of AD patients suggests that music is appreciated even in later stages of the disease. Many reasons for understanding this appreciation can be offered, but not the least is that music may be a welcome tool for improving the quality of life in institutions and private homes [4]. However, it is not clear whether anecdotal descriptions reveal the sparing of musical abilities in AD or a more general response to environmental stimulation. Several investigators report specific sparing of musical abilities [5–8]. Unfortunately, the lack of normal control data makes interpretation unclear; factors such as age, education, and musical experience have not been taken into account. In contrast, a few other investigators have compared AD patients with age-matched controls and reported difficulties with musical recognition – even

in those patients with relatively high cognitive function [9,10]. Familiarity ratings of traditional tunes showed mild impairment and naming the titles of the tunes showed moderate impairment, with respect to age-related controls, although patients did retain some knowledge of tune identity [10]. The discrepancy between case study reports of sparing and laboratory reports of difficulties presents an apparent contradiction.

Despite these challenges, we hypothesize that musical abilities, such as memory for familiar music, may be spared in dementia. That is, we propose to add musical recognition and memory to the list of differential degrees of AD impairments in domains such as language and visuospatial function [11]. Moreover, we hypothesize that musical recognition and memory may be reliably assessed with existing tests if behavioral observation is employed to overcome the problem of verbal or written communication.

In this paper, we describe an AD patient, EN, who provides an initial test of our general hypotheses. Although she suffers severe dementia with marked deficits in cognition and memory for events, language, and physical activity, EN was thought by caregivers to have normal memory for music. We assessed this claim by adapting three tests from the previous literature for behavioral observation. We report that, compared to controls, EN's test scores were in the normal or near-normal range. We then discuss possibilities for future research.

Case history

EN, born in 1920 in London UK, is a retired (1985) high-school teacher. In 1998, there was increased concern about confusion because of an episode of delirium occurring with pneumonia. The diagnosis of AD became probable by 2000. She managed with minimal help in her own home until October 2002, when she was moved to a small institution near family.

Surgical antecedents: Two normal pregnancies and deliveries (1950 and 1951); cervical cone biopsy, ca. 1965; bilateral cataract surgery, 2002.

Medical antecedents: None

Family history: Mother died in 50s of cervical cancer; father died in 70s of cerebral thrombosis. Only brother died age 54 of cerebral hemorrhage. An older sister is alive and well with normal cognition at age 89. A non-identical twin sister is alive with macular degeneration and no other medical problems; the twin's husband is living and well. Both sisters reside in UK.

Medication: None until donepezil from 2000 to 2002; rivastigmine for two months in 2002; and galantamine since early 2003. The patient has also been taking Vitamin E, glucosamine, and Ginkgo biloba since 2001.

Personal, education, work history: The patient completed high school and business college and worked as a stenographer in the British forces during the war. She emigrated to Canada as a war bride in 1946, and worked as a homemaker until her husband's sudden death in 1963. To support her family, she returned to work and completed a BA in 1973. She then trained and worked as a high school teacher until legal retirement age in 1985. In retirement, she enjoyed travel, hiking, cycling, and racquet sports, as well as music.

Music history: As a child, the patient accompanied her father, who was reputed to have perfect pitch, as he practiced the organ and rehearsed his church choir. Music was a part of the family's social life, and provided EN a rich childhood experience. While serving in the armed forces in WWII, she served as a disc jockey and music librarian for her unit. In the mid 1950s, she began formal piano training, receiving her grade 8 piano, music theory, and harmony in about 1960. EN never learned to play "by ear": she could play the piano only while reading music and her sight-reading skills were limited. She also learned to play recorder with an amateur society and was an avid concert-attendee, with a taste for early and baroque music. She was the volunteer choir leader at her local church for approximately six years. Rather than being formally educated, or musically talented, EN was always a deeply devoted amateur.

Present response to music: EN has not played an instrument in many years, but she enjoys listening to music, especially choral music and that which was once familiar. She still sings spontaneously in a given setting, songs that seem to be stimulated by her environment: "Oh what a beautiful morning!" "Blue moon," "In the bleak midwinter," "Blow blow thou winter wind." Given a few simple notes of a song or a hymn, without words, she can recall the words, sometimes in multiple verses, sometimes in foreign languages that she never spoke. She also appears to enjoy — foot tapping, smiling — music that she might once have scorned, such as cowboy ballads and salsa. (The latter observation is consistent with suggestions that listeners' musical taste may change over the life span [12]).

Physical examination: BP 120/70. Pulse 72 regular. No abnormalities of chest, heart, abdomen. Minimal degenerative arthritis of fingers. All findings are neurological and symmetrical. Mild cog-wheel rigidity. Reflexes difficult to elicit. Plantars

response normal. The patient has been completely incontinent since January 2003. Her walking was steady and firm in October 2002 for 2–3 km a day, but it became increasingly unsteady, with tendency to lean to the side or forward, and frequent falls, thought perhaps to represent an instance of Pisa syndrome [13,14]. By January 2004, she was confined to a wheelchair and could stand or walk only with support. The motor problem is thought to represent a form of motor apraxia consistent with the diagnosis of AD. She converses readily with a large vocabulary and responds appropriately to questions of social convention, but a long-standing problem with nominal dysphasia has degenerated into severe problems with speech. Although speech consists of well articulated words, her sentences are often garbled, incomprehensible, and incomplete. As she hesitates to search for a word, the original purpose of the sentence seems to be forgotten, and she abandons the effort. She sometimes displays mild frustration with this problem, but her mood is generally cheerful.

Mini-mental status examination: From the time of diagnosis, EN's mini-mental status examination (MMSE) [15] declined slowly over two years in the 25–20 range (normal 30) while on Aricept. In March 2000 her score was 23, in October 2002, it was 18, declining to 14 by April 2003; from January 2004 to the present (August 2004) it has been 8. In the early stages of her disease, the patient's verbal scores on MMSE testing were consistently higher than her orientation, short-term recall, and calculation scores. She could "write a sentence" without difficulty, but rarely knew a date, a location, nor was she able to draw a clock properly, to calculate or to recall names of more than a few vegetables or animals when asked to do so within 1 min. Nevertheless, she could recognize and name them or point to them when shown images. In sum, she has severe dementia of the AD type.

Music perception tests

EN was given several tests of music perception and memory during the fall of 2003 and winter/spring of 2004. Testing sessions were conducted in a quiet room in her residence. Each session lasted about 10–20 min.

EN was unable to remember the standard instructions for the tests and unable to make sense of printed response sheets. She cannot write. Nevertheless, she appeared to respond to the test materials by her behavioral and facial expressions, as described by the test administrator's observations

below. Without understanding their purpose, she clearly enjoyed the sessions.

Familiarity Decision test

This test is the first test of the 1998 version of the University of Montreal Musical Test Battery [16]. It consists of 10 familiar excerpts, from vocal (e.g., "Happy Birthday") and instrumental (e.g., "Blue Danube") pieces previously calibrated as familiar, mixed with 10 unfamiliar excerpts. The unfamiliar excerpts were the reverse in pitch and time of the familiar excerpts, i.e., "retrogrades". Excerpts were generated as synthesized piano tones, and were recorded on tape. The task is to determine which excerpts are familiar.

This test is intended as a "warm-up" for the battery itself; normal controls find it very easy. For 20 normal controls with mean age 66 years (range 59–71) and music background similar to EN, Steinke, Cuddy and Jakobson [17] reported a mean score of 96% correct (range 80–100). However, the test is sensitive to musical memory loss. Peretz et al. [18] have reported specific failures of familiar melody recognition following bilateral lesion of auditory cortex.

EN responded to familiar melodies by singing along, often with some of the words, and often continuing the melody after it stopped. She never responded to the unfamiliar melodies. The only trial missed was trial 10, as EN did not respond to the French song "Il est né le divin enfant". Having been raised in the UK, it is possible EN had never encountered the song. She did, however, sing along in French to the tune of "Frère Jacques". With trial 10 discounted, EN's score was perfect.

These same findings, with the same scores, occurred on several repeat testings from October 2003 to May 2004. While the scores remained unchanged, the test administrator noticed that the recognition responses became slower during this time period.

Distorted Tunes test

The Distorted Tunes test (DTT) [19] contains 26 popular melodies such as "Silent Night" and "Pop goes the Weasel". Seventeen of the melodies were distorted by a pitch change of two to nine notes, generally within one or two semitones of the correct note. Rhythm and contour were unaltered. Melodies contained synthesized complex tones and were recorded on CD. The task is to detect which melodies have been altered in pitch and which are correct.

In their study of genetic correlates of musical pitch recognition, Drayna et al. tested 284 pairs of twins aged 18–74 on the DTT [19]. The distribution of scores was markedly skewed. Although scores ranged from 9 to 26 correct, about half (46%) of the sample obtained scores of 25 or 26. A score of 23 was considered indicative of some deficit in pitch recognition.

EN responded to the familiarity of the melodies with apparent enjoyment and again hummed or sang along with the tunes long after the test music had stopped. However, she responded to distorted melodies with facial expressions – surprise, laughter, or a frown – or exclamation "Oh, dear!" Allowing these responses as indicators of detection, the result for EN was a score of 25/26. A replication of the test two days later also yielded a score of 25/26. The one melody that she missed on both tests was a distorted rendition of the "Battle Hymn of the Republic," in which the error occurred early in the short melody. As with the French song "Il est né le divin enfant", EN may not have encountered this song in her youth.

Famous Melodies test

The Famous Melodies Test was developed by W. Steinke [17]. It is conceptually similar to the Familiarity Decision Test in that participants are asked to indicate the familiarity of, and to give the name of, a series of melodies. However, it is also intended to assess recognition of song melodies ($N = 68$) separately from recognition of instrumental melodies ($N = 39$). There are also eight novel melodies in the series, for a total of 115 melodies. Song and instrumental melodies were intended to be highly familiar for a Canadian sample of normal controls.

Given the results for the first two tests, we were concerned whether all the melodies of the Famous Melody test would have been familiar to EN before the onset of AD. We obtained from a family member a selection of 11 test melodies that EN was not likely to have ever encountered. These 11 melodies were presented but not included in the final score (EN did not recognize them).

With behavioral observation as the recognition measure, EN's score for song melody recognition was 86% correct and for instrumental melody recognition was 64% correct. She falsely recognized only one of the eight novel melodies. Comparable results from normal controls, amateur musicians aged 59–71, were 98% correct for song recognition (range 91–100%) and 89% correct for instrumental

melody recognition (range 74–98%). Controls typically falsely recognized two of the eight novel melodies. Results for KB, an amateur musician aged 66 years, following a right-hemisphere stroke, were 88% for song melody recognition but only 18% correct for instrumental melody recognition. KB never falsely recognized a novel melody [17].

EN thus revealed some difficulties with music recognition on the Famous Melodies test, but her scores are close to the normal range. It must be noted that the culling of melodies thought to be unfamiliar to EN was conservative and EN may never have encountered other melodies. Thus her scores may be under-estimated. In any event, she did not show the failure of instrumental recognition revealed by KB, who, apart from his song melody recognition ability, fulfils the description of amusia.

Reliability check and final observations

The Familiarity Decision and the Distorted Tunes tests were re-administered in June 2004 in the presence of an independent observer. Complete agreement of scoring was attained between the test administrator and the observer. It was also noted that, despite exposure through repeated testing, the unfamiliar melodies of the Familiarity Decision test never elicited a recognition response from EN.

Although normative data are lacking, we tried an exploratory test of whether EN could correctly reproduce a familiar tune (such as those on the Familiarity Decision test) if prompted by spoken lyrics. The spoken lyrics of 12 tunes (e.g., “Au Clair de la Lune”, “Silent Night”, “Happy Birthday” and “Frère Jacques”) elicited correct vocal renditions of the tune, though only some of the words were recalled correctly. Two lyrics (“Frosty the Snowman”, and “Santa Claus is coming”) did not elicit a sung response, nor did two thought by the caregiver to be unfamiliar to EN (“Il est né” and “The Drummer Boy”).

In contrast to the production of correct vocal renditions, EN was unable to complete the lyrics of the tunes. However, the text of a tune could often act as a recall cue for the tune itself. This observation is consistent with an associationist account of song memory ([17], see also [2]). According to the associationist account, the melody analysis system and the speech analysis system for song are distinct, but interconnected through repeated coincidental activation. Activation of one system influences the level of activity in the other. In the case of EN, the speech analysis system is impaired, but activation of this system, even if

degraded, is nevertheless sufficient to co-activate an internal melodic representation. The melodic representation is relatively spared; thus recognition and recall of tunes are possible.

Discussion

EN's results provide encouraging support for our hypothesis that sparing of musical memory may be detected in dementia and may be reliably and quantitatively assessed through behavioral observation. We note dramatic contrast between EN's response to music and her mini-mental status. We also note that evidence of sparing would not have been recovered with conventional assessment – conventional assessment requiring verbal communication would suggest severe musical difficulties.

Many questions then arise: Is music preferentially spared in other cases of AD? Is EN typical of AD patients who once loved music, or is she an exception? And further: if such sparing is found among AD populations, is it an example of the sparing of an over-learned complex cognitive skill or is music “special”?

We propose several research directions to answer these questions. The efforts of test developers to design reliable, quantitative, music memory tests for AD patients should be supported [20]. As well, the repertoire of tests should be flexible for different applications. For example, the English-born EN's lack of familiarity with the American “Battle Hymn” or the French carol “Il est né” raises an important point: tests must respect the patient's date and place of birth, culture, and taste.

The case-study approach is instructive and should be pursued. Since the time of Galen, and probably earlier, experiments in physiology rely on animal models: a precise lesion – be it surgical or chemical – is deliberately created so that its effects can be studied; the loss of function, which results from the lesion, elucidates the role of the damaged part in the normal setting [21,22]. These methods were codified by Claude Bernard in the mid-19th century to create a paradigm of experimental surgery that became the founding principle that demarcated the academic discipline of physiology [23,24]. However, when it comes to control of the more subjective aspects of existence, those that rely on language for recognition – emotion, sensation, feelings, and memory – animal models are limited. Researchers must often rely on “accidents of nature,” provided by disease or trauma (e.g., [1,2,17]). We hypothesize that AD provides

yet another "accident of nature," for the study of music memory in dementia and, indeed, all forms of memory. The case-study approach will detect patterns of loss and sparing, if they exist – patterns that may be quite specific to an individual. Where sparing is suspected, a comparison of the patient's data with normal controls will reveal whether sparing is complete or whether the cognitive activity is nevertheless compromised [9,10].

We conclude with several considerations why music may be preserved in dementia. It may be that the presence of music, through enhancing a general level of activation, may prompt motor activity or memory recall. Two analogies spring to mind. Could music, with its regular rhythm, act quasi-mechanically on the verbal and motor systems, in a manner similar to kinesia paradoxa: the movement response elicited in a person with Parkinson's disease when wooden rungs, placed on a level, help overcome the extreme, immobilizing bradykinesia [25–28]? Or, are the words that come along with a tune evoked by an effect of music on emotional state, one that both heightens comfort and decreases anxiety thereby eliminating a basic form of verbal apraxia which wells up under stress?

How does music sparing relate to the sparing of other cognitive skills? A recent study reported sparing in AD of the skills involved in playing social games such as bridge and in solving jig-saw puzzles [29]. Musical memory is another example of a complex skill – a skill integrating pitch, rhythm, timbre, dynamic, linguistic, visual, kinesthetic, and emotional components. At early stages of auditory processing, hemispheric specialization may operate; the right auditory cortex, for example, is specialized for spectral (pitch) analyses, while the left auditory cortex is specialized for rapid temporal resolution [30]. However, complex musical tasks engage many processes, so that cortical activation patterns indicate widely distributed and partly interchangeable brain substrates [31]. In the case of AD, weakened components may be supported and reinforced through co-activation processes. As well, subcortical structures, spared the progressive destruction of cortical tissue, may play an important role.

Music may be "special", or at least different from many non-verbal cognitive skills, in that these distributed neuronal networks are acquired without formal training and intervention [32]. Children and many, if not most, adults enjoy music without possessing declarative knowledge of the rules of composition and vocal production. Much musical understanding, unlike the specific knowledge required for expertise at bridge or chess, is held in

common by members of a culture. Thus music sparing may be the most available and accessible form of the sparing of a complex skill in dementia.

When the enormous burden of dementia in our aging population is considered together with the pleasure music provides to all people, our hypotheses should suggest a host of research opportunities to those who are interested in music and memory.

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