

GENERALIZED AUDITORY AGNOSIA WITH SPARED MUSIC RECOGNITION IN A LEFT-HANDER. ANALYSIS OF A CASE WITH A RIGHT TEMPORAL STROKE

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ABSTRACT

After a right temporoparietal stroke, a left-handed man lost the ability to understand speech and environmental sounds but developed greater appreciation for music. The patient had preserved reading and writing but poor verbal comprehension. Slower speech, single syllable words, and minimal written cues greatly facilitated his verbal comprehension. On identifying environmental sounds, he made predominant acoustic errors. Although he failed to name melodies, he could match, describe, and sing them. The patient had normal hearing except for presbycusis, right-ear dominance for phonemes, and normal discrimination of basic psychoacoustic features and rhythm. Further testing disclosed difficulty distinguishing tone sequences and discriminating two clicks and short-versus-long tones, particularly in the left ear. Together, these findings suggest impairment in a direct route for temporal analysis and auditory word forms in his right hemisphere to Wernicke's area in his left hemisphere. The findings further suggest a separate and possibly rhythm-based mechanism for music recognition.

Key words: auditory agnosia, cortical auditory disorders, amusia, word deafness

INTRODUCTION

Generalized auditory agnosia, or cortical auditory disorder, is a rare condition usually associated with bilateral temporal lesions (Bauer, 1997). Auditory agnosia refers to an impaired capacity to recognize sounds in the presence of otherwise adequate hearing as measured by standard audiometry. The term has been used broadly to refer to impaired capacity to recognize sounds in general (generalized auditory agnosia) and, in a narrower sense, to a selective deficit in recognizing nonverbal sounds only (selective auditory agnosia).

In addition to generalized auditory agnosia, specific lesions can produce other "cortical auditory deficits." Selective auditory agnosia for meaningful sounds may result from lesions in the right hemisphere, especially the temporoparietal area (Fujii, Fakatsu, Watabe et al., 1990; Nielsen and Sult, 1939; Spreen, Benton and Fincham, 1965; Wortis and Pfeffer, 1948). Isolated verbal agnosia or pure word deafness (PWD) may result from bilateral or from isolated left temporal lesions that presumably disconnect Wernicke's area (Coslett, Brashear and Heilman, 1984; Shoumaker, Ajax and Schenkenberg, 1977). Patients with PWD cannot understand spoken language but they can read, copy and write spontaneously, and speak in a relatively normal manner

(Buchman, Garron, Trost-Cardamone et al., 1986). Isolated receptive amusia, or the loss of musical ability or appreciation, can follow right, left, or bilateral temporal strokes (Griffiths, Rees, Witton et al., 1997; Mavlov, 1980; Sparr, 1999). These different auditory syndromes imply that lesions in the left temporal region are necessary for agnosia for verbal sounds, lesions in the right temporal region are necessary for agnosia for non-verbal sounds, and either left or right-sided lesions may result in various disturbances in musical ability.

This paper presents a left-handed patient whose cortical auditory syndrome deviated from these patterns in several respects. He is unique because of the presence of both verbal and non-verbal auditory agnosia from a stroke involving the right temporal region, along with an increased interest and appreciation of music. The aim of this study was to clarify the underlying mechanism for his unusual combination of neurological auditory manifestations.

CASE REPORT

NS, a 68-year-old left-handed man, had difficulty understanding speech after a stroke sustained during coronary artery bypass surgery. When he awoke from anesthesia, he could not understand what people were saying, as if they were "speaking too fast or in Chinese." His own speech was not affected, and he could read and write. In addition, environmental sounds became indistinct and difficult to understand. The initial evaluation did not reveal focal abnormalities on his neurological examination, and his neuroimaging disclosed a right temporoparietal stroke.

Over the course of the next twelve years, the patient continued with difficulty comprehending speech and environmental sounds. His wife reported that people resorted to writing notes to him because he could not follow oral conversations. In general, communication was achieved through writing. In this way, NS was able to maintain his lifestyle, including reading multiple newspapers, following the stock market, and maintaining his finances. He gravitated to reading and to "listening" with closed captions to television programs. Moreover, his appreciation of music increased over the years. Previous to his stroke, he had not shown a prominent interest in music. After his stroke, he spend much of his day listening to music. NS and his wife became avid attendees at concerts and other musical performances.

Twelve years after his stroke, the patient underwent further evaluation for his auditory disorder. On interview, he was aware of his auditory difficulty and reported that speech still sounded fast and foreign and environmental sounds muffled. He endorsed a greater appreciation of music and would illustrate this by breaking out into song. The patient was a retired accountant who did not smoke and only occasionally used alcohol. He was strongly left-handed all of his life and was "left-footed" in sports. His past medical history was significant for coronary artery disease and benign prostatic hypertrophy, and his family history was negative for known familial illnesses and positive for left-handedness.

On examination, this older man appeared much younger than his age. He was alert, attentive, and well oriented to place and time. NS was very talkative and had a slightly empty but otherwise fluent output without paraphasic errors. When spoken to, however, he appeared confused and perplexed. He could not comprehend most simple, verbal, one-step commands or yes/no questions but had no difficulty comprehending written commands. For example, he could not understand the verbal command "touch your chin" but immediately understood it in writing, and when asked "are we in New York?" he replied "Yes, I was born in New York." Access to lip reading, careful attention to facial expression, and slowing of speech helped his auditory comprehension and allowed him to follow many simple commands. He attempted to visualize spoken words by spelling the words aloud or with his hands. Seeing a spoken word simultaneously written resulted in instantaneous auditory recognition with an expression of sudden discovery. The Token Test confirmed verbal, but

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