Receptive amelodia in a trained musician

Steven A. Sparr, MD

Clinical case studies of patients with isolated disorders of musical perception are exceedingly rare, and help elucidate the localization of various components of music in the brain. We report a highly trained musician who experienced profound inability to discern melody due to right temporal lobe injury.

Case history. A 91-year-old retired musicologist of great accomplishment with a history of diabetes mellitus and coronary artery disease suddenly developed difficulty reading the left side of his newspaper, unsteady gait, and difficulty dressing.

Initial examination showed no evidence of aphasia or dementia. He had a dense left homonymous hemianopia, mild left hemiparesis, and elements of left hemispatial neglect. On the evening of admission he experienced auditory hallucinations of a choir singing. All of these deficits cleared over the next 24 to 72 hours.

During his hospitalization his auditory abilities were studied in detail. He was unable to identify the melody of any of a wide variety of well-known musical pieces presented by recording or live by piano. He did not recognize relatively simple tunes played on a single instrument or vocal music, which contained the additional clues of verbal lyrics. He could repeat a series of three notes or fewer, but consistently failed to reproduce a series of four notes or more. He made errors in identifying instruments playing; at one point he identified the horn section as a harp.

At the same time, he had no difficulty humming a tune from memory. He was able to replicate the pitch of single notes. Given two notes, he had no difficulty indicating which note was higher. He readily distinguished consonant from dissonant chords. He could replicate the rhythm of a series of handclaps. Indeed, with many of the recorded pieces that he could not recognize, he was able to establish the rhythm and accurately pretend to lead the orchestra in time.

When presented with sheet music, he was immediately able to discern the melody of the composition, and readily categorized its style. He was able to explain the melodic lines and interactions of various instruments in a Stravinsky score.

With respect to other nonlinguistic auditory functions, he was unable to identify recorded sound effects, spoken or singing voices of famous personalities, or the emotional tone of a voice (receptive aprosodia). He made errors of up to 30 degrees in attempting to point to a sound source while blindfolded.

Carotid duplex revealed a hemodynamically significant stenosis of the right internal carotid artery. CAT scan and MRI of the brain failed to show an infarction. SPECT scan revealed decreased perfusion of the right temporal lobe (see the figure).

On follow-up examinations his ability to identify melodies improved to approximately 20% correct by 1 month after discharge, and ultimately to 70% accuracy by 3 years. However, even when he recognized a composition, recognition was not immediate. Throughout this time period he remained unaware of his deficits and denied any difficulty with musical perception.

Discussion. Our patient was found to have profound inability to discern melody despite intact perception of pitch, rhythm, and harmony. The cause was likely an ischemic injury of the right temporal lobe.

Clinical case studies have reported patients who developed receptive amelodia after unilateral ischemic 1,2 or hemorrhagic 3 in injury of the right temporal lobe. None of these patients had formal musical training. An amateur musician 4 experienced distortions in musical timbre and impaired recognition of the identity of voices and environmental sounds after right temporal lobe infarction, although he continued to recognize melodies. Another patient with some musical training 5 developed progressive loss of melody recog-

Figure. SPECT scan shows diminished perfusion of the right temporal lobe (left side of image).

References
ition due to a neurodegenerative disorder with focal onset in the right temporal lobe as demonstrated by SPECT scan.

It has been suggested that highly trained musicians utilize left hemispheric systems to perceive melody, whereas the musically naive process melody in the right hemisphere. In contrast, our patient, who was certainly a highly sophisticated musician, remained dependent on right hemispheric systems for auditory perception of melody.

This case corroborates the important role played by the right temporal lobe in the perception of music, particularly its melodic aspects. Even in highly trained musicians, the right hemisphere may remain critical for the perception of melody. In addition, the right temporal lobe may be essential in mediating other nonlinguistic auditory facilities such as decoding environmental sounds, discerning emotional prosody, and identifying voices. Finally, severe amelodia can occur without patient awareness of deficit. It is therefore incumbent on the examiner to probe the musical functions of patients with suitable lesions.

From the Stern Stroke Center, Montefiore Medical Center, Albert Einstein College of Medicine, Bronx, NY.

Refractory neurosarcoidosis responding to infliximab

J.A. Pettersen, MD, MSc; D.W. Zochodne, MD, FRCP; R.B. Bell, MD, FRCP; L. Martin, MD, FRCP; and M.D. Hill, MD, FRCP

Sarcoidosis is an idiopathic inflammatory disease characterized by granulomatous infiltration of multiple organs including the brain. The natural history is highly variable and neurologic involvement has been associated with greater resistance to treatment and increased overall morbidity and mortality.1,2 Corticosteroids are the mainstay of treatment but adjuvant approaches with antimalarials, cyclosporine, cytotoxic agents, and the anti–tumor necrosis factor-alpha (TNFα) agents thalidomide and pentoxifylline have been associated with some degree of disease response.3 Increasing evidence suggests that TNFα plays a pivotal role in the inflammatory cascade of this disease4,5 and recent observations suggest a beneficial response of refractory systemic sarcoidosis to infliximab, a chimeric monoclonal human-murine antibody directed against TNFα.6,7

We describe a case of refractory neurosarcoidosis in which the patient responded dramatically to infliximab therapy.

Case report.

Case presentation. A 46-year-old man developed biopsy-proven sarcoidosis 16 years previously initially involving the skin (lupus pernio) and subsequently the liver (granulomatous hepatitis), knees (synovitis), lungs (hilar lymphadenopathy), and brain (left temporal lobe lesion with associated focal seizures). His neurosarcoidosis was complicated by an episode of prolonged status epilepticus in 1997. Despite undergoing radiation therapy to

Figure. Initial and post-treatment brain MRI studies. Axial fluid-attenuated inversion recovery (FLAIR) (A) and gadolinium-enhanced T1-weighted images (B) at baseline reveal multiple supratentorial granulomas and a large left temporal lobe lesion with associated vasogenic edema. Post-treatment axial FLAIR (C) and gadolinium-enhanced T1-weighted images (D) show reduced volume of lesions and improvement of surrounding edema.
Receptive amelodia in a trained musician

Steven A. Sparr

*Neurology* 2002;59;1659-

This information is current as of July 24, 2007

Updated Information & Services

including high-resolution figures, can be found at:

http://www.neurology.org/cgi/content/full/59/10/1659

Subspecialty Collections

This article, along with others on similar topics, appears in the following collection(s):

All Neuropsychology/Behavior

http://www.neurology.org/cgi/collection/all_neuropsychology_behavior

Aphasia

http://www.neurology.org/cgi/collection/aphasia

Permissions & Licensing

Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at:

http://www.neurology.org/misc/Permissions.shtml

Reprints

Information about ordering reprints can be found online:

http://www.neurology.org/misc/reprints.shtml