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Postmorbidity learning of saxophone playing in a patient with frontotemporal dementia

Hanna Cho^a, Juhee Chin^b, Mee Kyung Suh^b, Hee Jin Kim^b, Yeo Jin Kim^b, Byoung Seok Ye^c, Na Kyung Lee^d, Eun Joo Kim^e, Sang Won Seo^b and Duk L. Na^{b*}

^aDepartment of Neurology, Gangnam Severance Hospital, Yonsei University College of Medicine, Seoul, South Korea; ^bDepartment of Neurology, Samsung Medical Center, Sungkyunkwan University School of Medicine, Seoul, South Korea; ^cDepartment of Neurology, Yonsei University College of Medicine, Seoul, South Korea; ^dDepartment of Health Sciences and Technology, SAHST, Sungkyunkwan University, Seoul, South Korea; ^eDepartment of Neurology, Pusan National University Hospital, Seoul, South Korea

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Some patients with frontotemporal dementia (FTD) show an artistic enhancement of musical abilities. However, no patients with FTD, to date, have been reported to be able to learn how to play a musical instrument after disease onset. Herein we describe a patient (J. K.) who had never played any musical instruments premorbidly, but who learned to play the saxophone after being diagnosed with a behavioral variant of FTD. He mastered a repertoire that consisted of 10 pieces of Korean folk songs over a period of three years. Furthermore, his saxophone skills were high enough to outperform other students in his class that consisted of ordinary healthy individuals without cognitive impairment.

Keywords: musical abilities; musical instrument; artistic talent; saxophone; frontotemporal dementia

Despite the progression of the disease, previously learned skills such as playing cards, dominoes, and board games can be preserved in dementia patients (Beatty & Greiner, 1998; Beatty et al., 1994; Greiner et al., 1997). Interestingly, some patients with frontotemporal dementia (FTD) even show an artistic enhancement of visual or musical abilities (Finney & Heilman, 2007; Hailstone, Omar, & Warren, 2009; Liu et al., 2009; Mell, Howard, & Miller, 2003; Miller, Boone, Cummings, Read, & Mishkin, 2000; Miller et al., 1998; Weinstein et al., 2011). Indeed, several reports have described patients who continue to play musical instruments skillfully even after being diagnosed with FTD (Hailstone et al., 2009; Miller et al., 2000; Weinstein et al., 2011) while another report described two FTD patients who began to compose music in the presence of disease (Miller et al., 2000).

However, no patients with FTD have been previously reported to be able to learn how to play a new musical instrument after disease onset. Here, we describe a businessman who had never played a musical instrument premorbidly, but who learned to play the saxophone after being diagnosed with a behavioral variant of FTD (bvFTD). His saxophone skills were high enough to outperform other students in his class that consisted of ordinary healthy individuals without cognitive impairment.

Case report

Patient J. K. was a right-handed businessman with 12 years of education. At the age of 58, changes in his

personality as well as abnormalities in his behavior became apparent to his wife, to whom he had been married for over 32 years. During conversations at work, J. K. abruptly stated his thoughts without considering the feelings of his fellow coworkers. Additionally, when his wife was hospitalized for an illness, he displayed an indifferent attitude towards her condition throughout the whole period of hospitalization. Concurrently, he also experienced language problems such as word-finding difficulties.

A year later, at the age of 59, he began to exhibit aggressive and impulsive behaviors. Premorbidly, he was very gentle and introverted, but at this stage of disease progression, he often became angry over trifling matters, spoke ill of others, and even became both verbally and physically violent. He also showed loss of decorum for he tended to speak impolitely and rudely to someone he met for the first time. In the past, J. K. enjoyed spending time with his friends and coworkers, but now he disliked participating in social gatherings. He used to voluntarily help his wife out with the household chores but now was unwilling to perform unless asked. He began to show a tendency to perseverate during conversations, often repeating the same phrases multiple times regardless of what the question was at that point. His language difficulties gradually increased to the point where he showed signs of trouble with naming and comprehension of familiar words. His wife noticed lapses in his memory evidenced by his frequently missed appointments and repetitive questions. At this time, his job performance was heavily affected, and he was unable to cope with social engagements.

*Corresponding author. Email: dukna@naver.com

Eventually, he became withdrawn socially, and soon he ended up quitting his job as a businessman. His past medical history was positive only for hypertension with no previous history of neurologic or psychiatric diseases. There was no other family history of mental or neurological disorders including dementia.

Patient J. K. first visited a university hospital in Seoul in January 2009 (at the age of 59 (Year 1)) due to changes in his behavior and language abilities. At the first neuropsychological assessment, his Mini-Mental State Examination (MMSE) score was 26 out of 30, and he displayed predominant impairments of frontal/executive functions as well as verbal memory (Table 1). In contrast, his attention, visuospatial function, visual memory, and ideomotor praxis had been spared. Despite the symptom of language impairment reported by his wife, he scored 42 out of 60 on the Korean version of Boston Naming Test,

which was within normal ranges according to age/education-matched norms (Kim & Na, 1999). Brain magnetic resonance (MR) images revealed mild atrophy in the bilateral dorsolateral and medial frontal cortex. The larger anterior horn of the left lateral ventricle was suggestive of greater atrophy on the left frontal lobe than on the right. Anterior temporal atrophy was also noted with the left being more severe than the right, and the temporal horn atrophy was also greater in the left than the right (Figure 1 (A) and (B)). Brain 2-[¹⁸F]fluoro-2-deoxy-D-glucose positron emission tomography (FDG-PET) scans also displayed significant levels of hypometabolism in the bilateral dorsolateral and medial frontal area and anterior temporal lobe, both of which were more severe on the left than on the right (Figure 2 (A) and (B)). His elementary neurological examination, including cranial nerve function, motor and sensory examinations, cerebellar function, extrapyramidal signs, and reflexes, was all found to be normal.

After this diagnosis, in February 2009, J. K. began to receive saxophone lessons according to his wife's suggestion. His wife thought playing a musical instrument would soothe his abnormal behaviors. He had not received any formal musical training before, had never learned how to read musical notes, and most importantly had never played any musical instrument in the past. Beginning around the time of his diagnosis, he participated in a saxophone class for 2 hours a day. At first, it took a long time for him to learn how to read musical notes and to play the saxophone. However, his skills progressed, and soon he was able to play new, unfamiliar songs every 2-3 months.

In November 2011, at the age of 61 (Year 3), he was referred to our clinic. The patient showed widespread cognitive impairments with profound deficits in memory, language, and visuospatial function, and he also showed signs of apraxia as well as behavioral problems. When the patient visited our clinic at Year 3, during examinations at the outpatient clinic, he was polite but was rather impatient or distracted, for he kept trying to leave the examination room and repeatedly expressed his wishes to go home. Frequent bursts of inappropriate laughter were also observed. Apathy, loss of emotions, and disinhibition were also noted; however, aggressiveness and anxiety had reduced in comparison with his condition at Year 1. No formal language evaluation was performed, but on bedside evaluation, he seldom spoke spontaneously if not spoken to. When questioned, he only answered with short phrases such as "I don't know." His auditory comprehension and naming seemed to be impaired. In contrast to these deficits, however, his repetition of four- to five-word sentences was preserved, suggestive of a language deficit close to mixed transcortical type of aphasia. He manifested mild utilization behavior and stereotypy of speech during the interview. His MMSE score was 7 out of 30, and he did not show parkinsonism or abnormalities in extraocular

Table 1. Neuropsychological test results.

| Tests (possible maximum score) | Year 1 | Year 3 |
|---|------------|----------|
| MMSE | *26 (3) | 7 (<1) |
| <i>Attention</i> | | |
| Digit span: forward (9) | 6 (34) | 6 (39) |
| Digit span: backward (8) | 2 (8) | 0 (<1) |
| <i>Language and related functions</i> | | |
| K-BNT (60) | 42 (25) | 7 (<1) |
| Calculation (12) | 6 (<5) | 0 (<5) |
| Ideomotor limb apraxia (5) | 5 (≥15) | 0 (<5) |
| <i>Visuospatial function</i> | | |
| RCFT (36) | 36 (84) | 1 (<1) |
| <i>Memory</i> | | |
| SVLT: sum of three free recall trials (12 + 12 + 12 = 36) | 17 (8) | 0 (<1) |
| SVLT: 20 min delayed recall (12) | 0 (<1) | 0 (<1) |
| SVLT: recognition (12) | 0 (<1) | 0 (<1) |
| RCFT: immediate recall (36) | 16 (36) | 1 (<1) |
| RCFT: 20 min delayed recall (36) | 14.5 (27) | 1 (<1) |
| RCFT: recognition (12) | 0 (<1) | 0 (<1) |
| <i>Frontal/executive function</i> | | |
| Contrasting program | Abnormal | Abnormal |
| Go/no-go test | Abnormal | Abnormal |
| Fist edge palm | Normal | Normal |
| Alternating hand movement | Borderline | Abnormal |
| Alternating square triangle | Normal | Deformed |
| Luria loop | Normal | Normal |
| Motor impersistence | Normal | Normal |
| COWAT: animals | 9 (3) | 0 (<1) |
| COWAT: supermarket items | 3 (<1) | 0 (<1) |
| COWAT: phonemic fluency, <i>three</i> letters | 15 (5) | 0 (<1) |
| Stroop test: color reading in 2 min (112) | 24 (<1) | 0 (<1) |

Note: *Values are raw score (percentile).

K-BNT, Korean version of Boston Naming Test; COWAT, Controlled Oral Word Association Test; MMSE, Mini-Mental State Examination; RCFT, Rey-Osterrieth Complex Figure Test; SVLT, Seoul Verbal Learning Test.

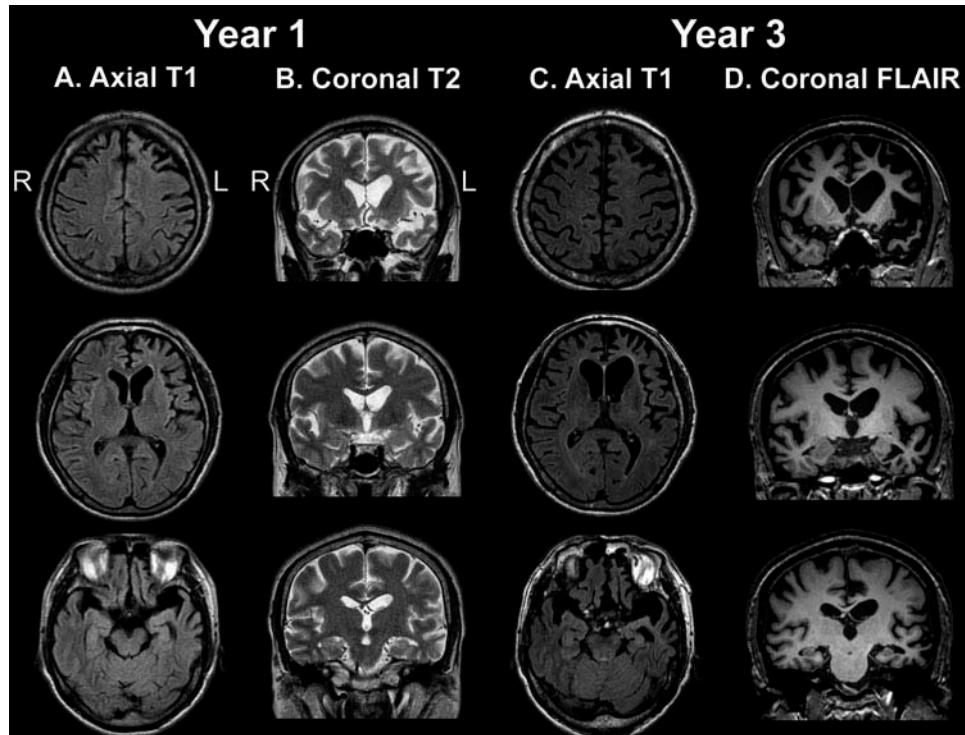


Figure 1. Brain MR images of patient J. K. at Year 1 (A and B) and Year 3 (C and D): In Year 1 (age 59), brain MR images reveal mild atrophy in the bilateral dorsolateral and medial frontal cortex. The larger anterior horn of the left lateral ventricle is suggestive of greater atrophy in the left frontal lobe than the right. Anterior temporal atrophy was also noted with the left being more prominent than the right, which was further supported by the greater temporal horn atrophy in the left than that of the right (A and B). Brain MR images in Year 3 (age 61) show more marked atrophy in the bilateral anterior temporal areas as well as dorsolateral and medial frontal lobes with the left being more severe than the right. Atrophy in the left insula and anterior temporal lobe (predominantly involving the left temporal pole) worsened in comparison with the MR images acquired in Year 1 (C and D).

Note: R, right side; L, left side.

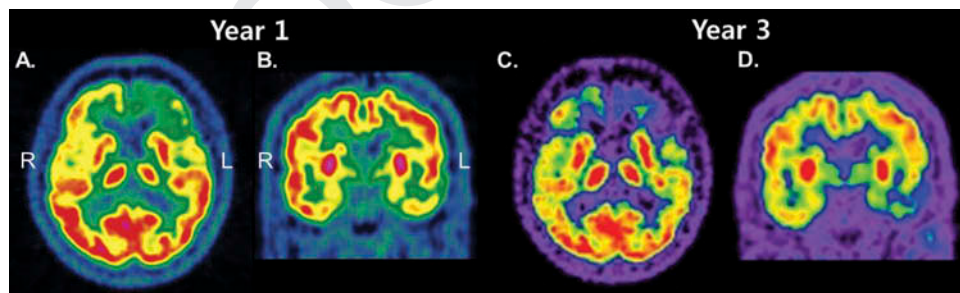


Figure 2. Brain FDG-PET scans of patient J. K. at Year 1 (A and B) and Year 3 (C and D): Axial (A) and coronal (B) scans at Year 1, and axial (C) and coronal (D) scans at Year 3. At the age of 59 (Year 1), FDG-PET scans reveal significant hypometabolism in the bilateral dorsolateral and medial frontal area, and anterior temporal lobe, both of which are more severe on the left than on the right side (A and B). At the age of 61 (Year 3), FDG-PET scans display aggravated hypometabolism patterns in comparison with scans acquired at Year 1 (C and D). [To view this figure in color, please see the online version of this journal.]

Note: R, right side; L, left side.

movements. Upon neuropsychological assessment, J. K. displayed impairments across all cognitive domains except for attention (Table 1). Brain MR images showed a more marked atrophy in the bilateral anterior temporal areas as well as the bilateral dorsolateral and medial frontal lobe

with the left being more severe than the right. From the MR images, atrophy in the left insula and anterior temporal lobe (predominantly involving the left temporal pole) was greater at Year 3 in comparison with Year 1 (Figure 1, (C) and (D)). FDG-PET scans also displayed

aggravated hypometabolism patterns in comparison with scans acquired at Year 1 (Figure 2(C) and (D)).

Despite these cognitive and behavioral abnormalities, the patient continued to practice his saxophone daily for 2 hours over a 3-year period. From the time of diagnosis, he had mastered a repertoire that consisted of 10 Korean folk songs. In comparison with other members of his class (all of whom were healthy and possessed no notable memory problems or brain damage), his teacher considered him an excellent saxophonist with advanced skills in accuracy, rhythm, and tempo. According to the teacher, J. K. was a good sight-reader, and due to his virtuosity, he was even able to represent his class in a contest during which he played two full pieces by himself (Supplementary audio clip). In September 2012, at the age of 62 (Year 4), he became almost mute with some echolalia. He was still interested in saxophone and enjoyed playing the instrument daily for two hours. Although he could no longer learn new songs, he was still able to play previously learned songs by sight-reading.

Discussion

The clinical features of patient J. K. in the earlier stage of the disease were characterized by a gradual progression of behavioral disinhibition such as loss of decorum and impulsive actions (including early decline in social interpersonal conduct and impairment in regulation of personal conduct), apathy, loss of empathy or emotional blunting, loss of insight, and perseveration of language. Therefore, patient J. K. satisfied both the Neary (Neary et al., 1998) and International diagnostic criteria (Rascovsky et al., 2011) for bvFTD. Moreover, severe atrophy of the bilateral dorsolateral and medial frontal lobes, the left insula, and the left anterior temporal lobe evident from MR images and FDG-PET scans was also consistent with the diagnosis of bvFTD. However, J. K.'s disinhibition was not as florid as can be seen from typical bvFTD patients. Furthermore, language problem was one of his early symptoms which first started with naming and comprehension difficulties and which soon progressed to mixed transcortical aphasia (Year 3) and muteness (Year 4) over time. Therefore, J. K. had some clinical features that were characteristic of a language variant of FTD, especially semantic variant, given the marked left temporal atrophy based on MR images. Thus, although we've made the final diagnosis of J. K. as bvFTD, J. K. had an FTD syndrome that was difficult to characterize.

Interestingly, J. K. learned to play saxophone for the first time after the diagnosis of FTD, and continued to learn new songs every 2–3 months, despite progressive cognitive decline and overall apathy. These findings suggest that it is possible to learn how to play a new musical instrument even with FTD.

Prior case reports have described several patients with dementia who continued to skillfully play musical instruments; however, these patients were professional musicians premorbidly and had preserved musical performance even after the diagnosis of FTD (Hailstone et al., 2009; Miller et al., 2000; Weinstein et al., 2011). In contrast, J. K. had never played a musical instrument and had received no formal musical training before his diagnosis of FTD. To our knowledge, J. K. is the first patient ever reported who, despite no prior musical training, learned how to play a new instrument and performed around 10 new musical pieces after the onset of FTD.

How saxophone playing was possible in this particular case, even after the onset of FTD, is an intriguing question with several possible explanations. First, learning how to play a new instrument may require basic motor skills. Motor skills for playing a musical instrument can be attributed to procedural, non-declarative forms of memory (Crystal, Grober, & Masur, 1989; Polk & Kertesz, 1993). Non-declarative memory includes different forms of learning and memory abilities, including the perceptual and motor skills involved in musical performance. The basal ganglia, cerebellum, and supplementary motor regions play a collective role in procedural memory (Daselaar, Rombouts, Veltman, Raaijmakers, & Jonker, 2003; Exner, Koschack, & Irle, 2002). Since these areas are relatively spared in early FTD (Broe et al., 2003; Kril & Halliday, 2011), J. K. might have been able to acquire and maintain the procedural memory skills required for playing a musical instrument at the early stage of the disease. Second, visuo-perceptual skills are required to read musical scores (Miller et al., 2000). Patients with FTD often preserve visuo-constructive abilities because pathology is minimal in the posterior parietal regions (Brun, 1993). Despite progressive FTD, the sparing of these areas might have allowed J. K. to sight-read and play music. J. K., in fact, showed preserved visuospatial functions at the Year 1 evaluation. Third, relative sparing of the right hemisphere which was exhibited from J. K.'s brain images might have contributed to his musical talent. Previous studies have proposed that a relative sparing of the right hemisphere allows preservation of musical skills in degenerative diseases of the brain (Polk & Kertesz, 1993), and a right-lateralized working memory system is important for playing music in normal subjects (Marin & Perry, 1999). Moreover, as reported from previous studies (Miller et al., 2000, 1998), asymmetric hemisphere degeneration may have predisposed J. K. to develop artistic talents. According to the "paradoxical functional facilitation" hypothesis (Kapur, 1996), an unexpected occurrence of enhanced artistic skills can arise from brain injury. Loss of inhibitory activity due to a loss of function in the left anterior temporal lobe could have enhanced the right-hemisphere activation resulting in manifestation of musical talent.

In conclusion, the present study showed that it is possible for a patient with a left-hemisphere-predominant FTD syndrome to learn how to play a musical instrument, which may have implications for the use of music therapy and music-based cognitive rehabilitation in FTD patients.

A limitation of our case report is that we could not make a pathologic diagnosis of the patient. Regarding the patient's possible underlying pathology, however, TAR DNA-binding protein 43 immunoreactive inclusions would be more likely than tau pathology because of the absence of parkinsonism (Josephs et al., 2011). Additionally, J. K. went through genetic tests such as microtubule-associated protein tau (Hutton et al., 1998), progranulin (Baker et al., 2006), and chromosome 9 open reading frame 72 (DeJesus-Hernandez et al., 2011), which all turned out to be negative.

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Supplementary material

Supplementary content is available via the "Supplementary" tab on the article's online page (<http://dx.doi.org/10.1080/13554794.2014.992915>).

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