

**Modified electroconvulsive therapy for the treatment of refractory schizophrenia-like psychosis
associated with Huntington's disease.**

Takeshi Nakano, Shinji Ono, Junji Yamaguchi, Ryu Sugimoto, Naohiro Yamaguchi, Yoshiro Morimoto,

Tatsuya Kubo, Hiroki Ozawa, Naohiro Kurotaki.

Department of Psychiatry, Nagasaki University Hospital, Nagasaki, Japan

Correspondence:

Shinji Ono, MD, PhD.

Sakamoto 1-7-1, Nagasaki 852-8523, Japan

Tel: +81-95-8197293 *Fax:* +81-95-8197296 *E-mail:* shinji1231zzz@nagasaki-u.ac.jp

Dear Sirs,

Huntington's disease (HD) is a progressive neurodegenerative disorder characterized by involuntary movements, dementia and psychiatric symptoms [1]. Psychiatric symptoms in HD resemble those of schizophrenia; therefore HD is sometimes misdiagnosed as schizophrenia clinically [1-3].

The efficacious effect of modified electroconvulsive therapy (mECT) has been reported in both psychiatric disorders and in movement disorders including dyskinesia and Parkinson's disease [4, 5].

However, there have been few reports of mECT for the treatment of schizophrenia-like psychosis in HD.

Here, we describe a case of a patient with HD who had psychiatric symptoms refractory to neuroleptics, whose symptoms were dramatically improved by mECT.

A 59-year-old man was admitted to our hospital because he had auditory hallucinations and delusions of persecutions. His mother had been diagnosed with dementia resulting in long-term hospitalization. His clinical characteristics were mostly unremarkable except for imperceptible involuntary movements of the lip. Initial treatment using risperidone (to 6mg/day) was ineffective and just caused extrapyramidal side effects; subsequently his medication was changed to olanzapine (20mg/day). However, his psychiatric symptoms showed no improvement.

Based on treatment-refractory psychosis we then introduced mECT. After the fourth treatment with

mECT, his delusions and hallucinations were dramatically improved and he regained fluent speech, although his involuntary lip movements were still present. During serial mECT treatment, his elder brother had visited the department of neurology in our hospital complaining of involuntary movement and was subsequently diagnosed with HD by a genetic test. This finding led us to surmise that the patient's psychiatric symptoms were associated with HD. The genetic test for HD revealed that the patient had 44 CAG repeats. Thus we recognized the patient's clinical symptoms as psychiatric features of HD.

Subsequently, we have continuously performed mECT and the patient has shown no psychotic exacerbation. Single-photon emission computed tomography (SPECT) scans, brain MRI images, mini-mental state examination (MMSE) scores, positive and negative symptom scale (PANSS) and brief psychiatric rating scale (BPRS) scores before and after mECT are summarized in Fig. 1.

Several studies have reported that patients with HD frequently have accompanying psychiatric symptoms [1-3]. The frequency of schizophrenia-like psychosis ranges from 3% to 12% of patients with HD [6]. Before the identification of the *HTT* gene, dementia or schizophrenia-like psychosis associated with HD that displayed little involuntary movement would have been misdiagnosed as Alzheimer's disease or schizophrenia, and would have led to long-term hospitalization as in the case of the patient's mother. Our case suggests that a renewed focus on taking a detailed family history into account should be

applied when considering the possibility of HD, especially if early-onset dementia and/or late-onset psychosis exists in other family members.

To date, there is just one report of mECT for the treatment of schizophrenia-like psychosis associated with HD [7]. Neuroleptics are a standard treatment of psychosis in HD; however, the effect of pharmacotherapy on psychosis in HD remains unclear. Furthermore, neuroleptics cause abnormal movements due to extrapyramidal side effects [8]. In this particular case, mECT dramatically improved psychosis associated with HD without worsening involuntary movements. The fact that mECT does not cause extrapyramidal side effects raises the possibility that mECT could be a first-line treatment of schizophrenia-like psychosis in HD (Table 1).

Recently, Mughal et al. demonstrated that ECT could protect neurons against mutant huntingtin protein resulting in improved functional outcome, leading to slow disease progression [9]. In agreement with Mughal et al., the results of brain MRI and MMSE scores of before and after treatment, showed that the cognitive function of our patient remained clinically unchanged. Several studies have indicated that the changes in SPECT reflect progression of the disease [10]. Furthermore, van den Bogaard et al. suggested that it is difficult to detect the visible structural changes of HD patients at 2-year follow-up [11]. Unchanged MMSE scores and MRI images may not be due to the mECT treatment, but be a natural

course of the disease. Together with the SPECT results, we conclude that the patient's pathological state

may progress, which may indicate that mECT does not arrest the progression of HD.

Conflicts of interest: The authors report no disclosures related to the current case presentation.

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Figure legend

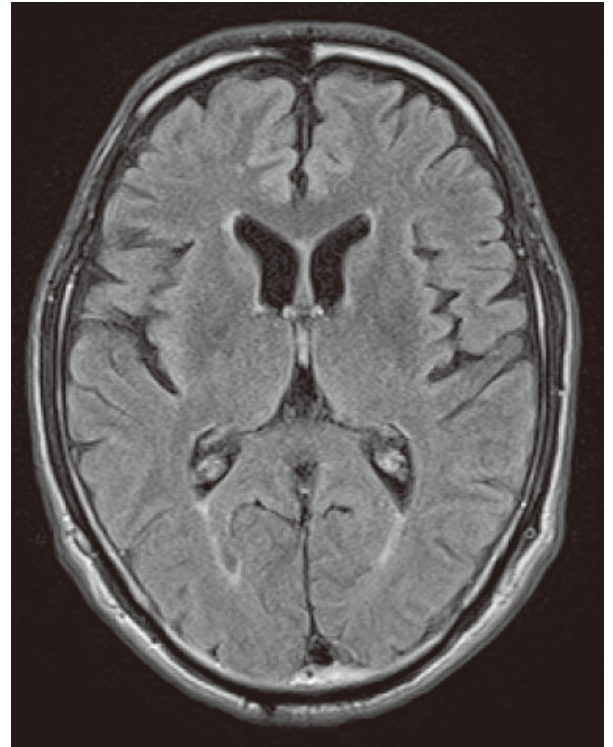
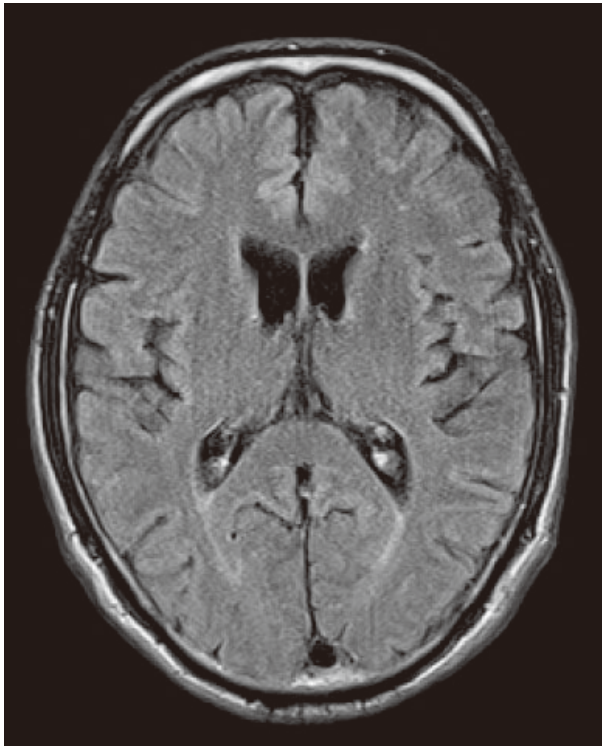
Fig. 1. Comparison of brain MRI, SPECT and psychiatric rating scales before and after mECT. All data from the SPECT images were analyzed using an easy Z-score imaging system. The brain MRI before mECT that showed slight atrophy of the subcallosal gyrus was similar to that of after mECT treatment. The SPECT after mECT treatment that showed a decrease in ^{99m}Tc -uptake in the basal ganglia, cingulate gyrus and thalamus was greatly decreased compared with the analysis before mECT.

Fig. 1

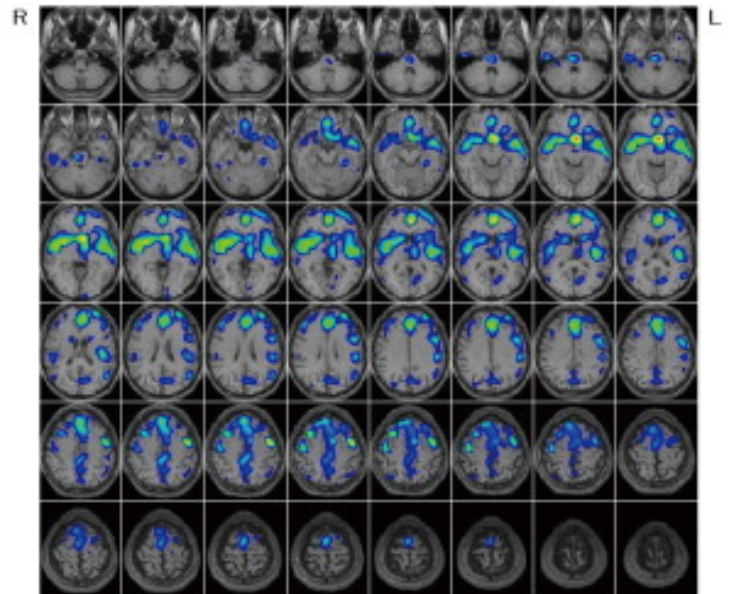
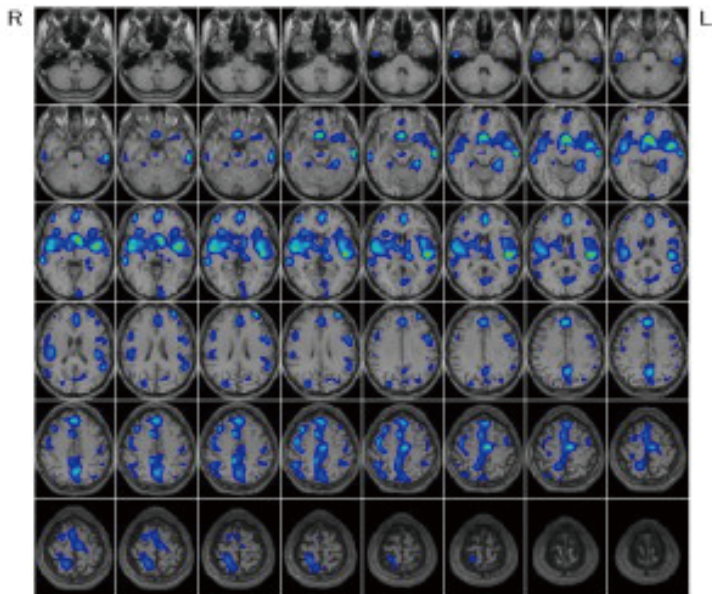
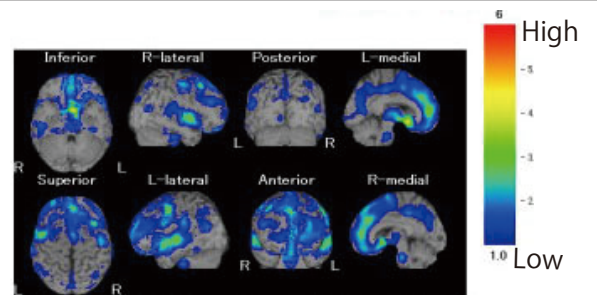
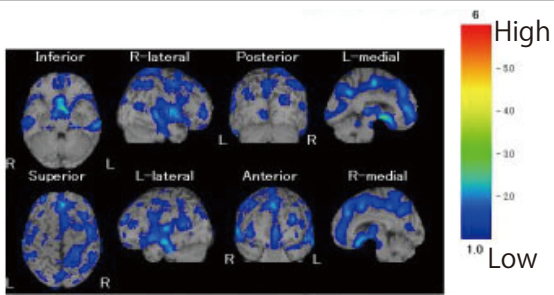
Before mECT

After 21st mECT
(six months had passed since the first-time mECT)

Brain MRI



SPECT



Psychiatric rating scales

MMSE 27
PANSS 139
BPRS 84

26
68
36

Table 1. The pros and cons of mECT treatment versus neuroleptics treatment for psychosis in Huntington's disease.

	Merit	Demerit
mECT	<p>More potent effect</p> <p>Fewer side effect</p> <p>No absolute contraindications</p>	<p>Cumbersome introduction</p> <p>Need particular equipment</p> <p>High cost</p> <p>Antero/Retro-grade amnesia and transient cardiovascular complications as side effects</p>
Neuroleptics	<p>Easy introduction (easy-to-take)</p>	<p>Lethal side effect including malignant syndrome and cardiovascular complications</p> <p>Worsening abnormal movement due to extrapyramidal side effect</p>