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MATTERS ARISING

Transient musical hallucinosis

Paquier *et al* reported a patient who, following subarachnoid haemorrhage, developed musical hallucinosis. Based on a literature review, they suggested that musical hallucinosis, formed auditory perceptions that occur in the absence of external acoustic stimulus while the patient is aware of their non-real nature, may result from lesions of either side of the brain, and not necessarily from the non dominant hemisphere, as previously proposed.¹ A patient recently seen by us reinforces the authors' conclusion.

A 75 year old right handed woman had been suffering from severe hearing loss due to streptomycin use for about 30 years. Her past history revealed non insulin dependent diabetes mellitus, ischaemic heart disease, peripheral vascular disease and paroxysmal atrial fibrillation. In September 1992, she suddenly developed right hemiparesis and dysphasia which recovered within a few weeks. Her CT scan revealed a left thalamic infarction, mild cortical atrophy and ventricular enlargement. A few days after the event, she started hearing a melody, which

seemed in the first days to originate externally and was heard bilaterally. The melody she heard was extremely loud, leading her to ask surrounding people to turn off the radio, which she believed to be the source of the tune. The music began suddenly, was slow, clear and reminiscent of popular songs that she had heard in her youth, but were still unknown to her. She was able to sing this melody. Shortly after the onset of this phenomenon, she gained full insight into the problem and realised that this incessant tune originated in her own mind. The volume was variable and sometimes the melody was enjoyable; the volume was mostly high, especially during the night, disturbing her sleep, and severely interfering with her daily activities. Amitriptyline partially helped her sleep. The intensity of the music diminished during the following weeks, but the same melody persisted.

Musical hallucinations after stroke are reported rarely. Only three cases, all with right hemispheric pathology, were quoted in a recent review.¹ Our patient illustrates the fact that dominant hemispheric stroke can also result in musical hallucinations.

As with several other reported cases, including that of Paquier *et al*, our patient had suffered from hearing loss for many years. Berrios in a review, pointed out that musical hallucinations are far more common in elderly, hearing impaired, female patients.¹ It is possible that musical hallucinations represent a "deafferentation" phenomenon, reminiscent of visual hallucinations in the blind, thalamic pains or phantom limb. It appears that both central and end organ pathology contribute to the appearance of musical hallucinations.² The prolonged lack of normal input to cortical areas involved in hearing, due to peripheral disease, might cause a specific vulnerability which results in the generation of this abnormal sensation following a central insult. Appropriately, Wengel *et al* entitled their manuscript "musical hallucinations, the sounds of silence?",³ as they occur when the mind is chronically deprived from music and sound.

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Inhibition of motor unit discharge in humans evoked by transcranial stimulation

Transcranial magnetic stimulation of the motor cortex can elicit contraction of contralateral muscles but, until recently, there had been no reports that transcranial magnetic stimulation could suppress muscle contraction. We have shown¹ that inhibition of voluntary contraction can be elicited by transcranial magnetic stimulation and have presented evidence^{2,3} that the mechanism is likely to be supraspinal, presumably involving inhibition of corticospinal neurons with consequent disfacilitation of motor neurons.

The inhibition was revealed in our studies during a period of voluntary contraction by stimulating the motor cortex at a strength lower than that required to produce excitation under the same conditions. A recent study has reported that the discharge of motor neurons in the first dorsal interosseus muscle of the hand of a patient with multiple sclerosis could be suppressed by transcranial magnetic stimulation of the motor cortex, but this was not observed in normal subjects.⁴ In our previous studies¹⁻³ we averaged the rectified surface electromyogram (EMG) to reveal inhibition of voluntary contraction in a number of different arm and hand muscles. We have now re-investigated one of our subjects to examine the effect of transcranial magnetic stimulation on the probability of discharge of single motor units in the first dorsal interosseus muscle. We can confirm that transcranial magnetic stimulation at a strength which causes a reduction in gross surface EMG, and is sub-threshold for excitation, does lower the probability of discharge of individual motor units in normal humans.

The subject was a right handed male (age 49 years) with no history of neurological illness. Local ethical approval for the study was obtained and the subject gave his informed consent to the procedures. Two forms of electromyographic recordings were made from the first dorsal interosseus muscle of the right (dominant) hand. To record gross EMG, surface electrodes were placed over the belly of the muscle and at an indifferent point over the proximal interphalangeal joint of the first digit. A concentric needle electrode was inserted percutaneously into the first dorsal interosseus muscle to record the discharges of single motor unit. The subject was required to make a weak voluntary contraction of the muscle. Auditory feedback of the signal was provided to enable the subject to recruit and maintain the discharges of a motor unit that could be reliably identified and selected for peri-stimulus time histogram analysis. Transcranial magnetic stimulation was delivered from a Novamatrix 200 stimulator using a 9 cm round coil centered over the vertex. The initial direction of current flow in the coil was anticlockwise and adequate stimuli preferentially excited muscles on the right side.

The threshold transcranial magnetic stimulation required to produce an initial excitatory response, gauged from the surface EMG recording, was 40% of maximum output. The response had a latency of 23 ms and was followed 5-8 ms later by a period of suppressed EMG lasting 30 ms and culminating in a late period of increased EMG activity. Part A of the figure shows the average of the full-wave rectified surface EMG response to 50 magnetic stimuli at 37% of output. At this strength of stimulation no initial excitation occurs but suppression of EMG is evident with a latency of 29 ms and duration 26 ms. The peri-stimulus time histogram in part B of the figure is constructed from the discharges of a single motor unit (average frequency 9.1 impulses/s) during 100 magnetic stimuli also at 37% of maximal output. A profound depression of firing occurs also lasting 26 ms and with a latency of 29 ms. Within the period of suppressed firing the unit discharges on only 8 occasions during the 100 trials. The number of counts expected for that period of the peri-stimulus time histogram in the absence of stimulation was

Figure A Averaged surface, rectified EMG from the first dorsal interosseus muscle, during weak voluntary contraction, in response to 50 magnetic stimuli at a strength (37% of output) below threshold for direct excitation. **B**: Transcranial magnetic stimulation (below) and cumsum (above) of the discharges of a single motor unit in the first dorsal interosseus during 100 magnetic stimuli at 37% output under identical conditions to those in A.

65. Thus the depression represents an 88% reduction in probability of discharge. The period of suppressed firing is followed by a pronounced increase in the probability of discharge lasting about 10–12 ms. There is no evidence in the peri-stimulus time histogram of a period of facilitation before the depression and this is emphasised by the flat time course of the cumsum.

The responses to transcranial magnetic stimulation of three other motor units were examined in the same subject. All showed the same qualitative pattern of response to transcranial magnetic stimulation at a strength (37%) just below threshold for exciting contraction of first dorsal interosseus. The periods of suppressed firing of the other three units started 31, 29, and 29 ms after the stimulus and lasted for 27, 23 and 21 ms respectively. On reducing the strength of stimulation to 35% of maximal output suppression of motor unit discharge was still evident starting at 29 ms after transcranial magnetic stimulation but lasting only 12 ms.

The results show that substantial suppression of motor unit discharge in normal humans can occur in response to transcranial magnetic stimulation in the absence of an earlier period of excitation. This was to be expected from our previous findings^{1–3} and shows that the suppression is not restricted to instances of neurological disorder.⁴ The finding expands the realm of transcranial magnetic stimulation in the investigation of cortical motor function in normal humans. It also means that abnormal responses to transcranial magnetic stimulation in cases of disordered brain function may reflect disturbance to either excitatory or inhibitory inputs to corticospinal neurons.

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Boniface and Mills reply:

We note with interest the results of Ellaway *et al* on the inhibition of motor unit discharge by transcranial magnetic stimulation in the intact human nervous system. This raises important issues related to the mechanisms involved and to our own findings in a patient with multiple sclerosis (MS).¹

We have studied five single motor units from the first dorsal interosseus muscle of four healthy subjects at stimulus intensities just below that required to produce a short latency excitatory response. A partial but significant reduction in firing probability was produced in three of these five motor units which was confirmed on cumsum analysis (Mills and Boniface, unpublished observations). Complete suppression of discharge as in the patient with MS, however, was not seen.

The prominent late peak evident in the peristimulus time histogram of Ellaway *et al* has a latency similar to the secondary peak described in histograms obtained at higher stimulus intensities.^{2,3} This may reflect not only an accumulation of delayed discharges from the inhibitory period but also a late excitatory event. This was not encountered in the three motor units from the patient with MS suggesting a different mechanism for the observed changes in firing probability.

High stimulus intensities are sometimes used when studying MS patients because of the high motor threshold, which is a common feature in this condition. The effects of transcranial magnetic stimulation on firing probability therefore are not necessarily directly comparable between patients and healthy subjects. Problems also arise from the selective nature of single motor unit recordings with transcranial magnetic stimulation which sample a small number of low threshold motor units. Despite these difficulties, it is clear that transcranial magnetic stimulation produces some suppressive effects in healthy subjects, but so far descriptions of the complete silencing of motor unit discharge in the absence of peaks in the peristimulus time histogram are confined to one patient with MS. It is conceivable that this phenomenon is a function of stimulus intensity, both in absolute terms and in relation to motor threshold. The evidence, however, suggests that changes in the balance of descending excitatory and inhibitory influences on the motor neuron by transcranial magnetic stimulation, whether by transient withdrawal of excitatory drive or by active inhibition, are different in the patient with MS when compared with healthy controls. These differences can be exploited to study the disordered central mechanisms of motor control in neurologi-

cal disease and the role of inhibitory processes.

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MRI in a case of rigid Huntington's disease

In a recent paper Savoiardo *et al*¹ report MRI findings of 18 patients with Huntington's disease (HD), seven of whom had the rigid variant. They found increased signal intensities of the caudate and putamen in all the rigid patients in intermediate weighted images and in all but one in T2 weighted images. All but one hyperkinetic patient had no signal abnormalities on MRI. Sethi² and Kang *et al*³ also report putamen hyperintensities in single cases of rigid HD. In contrast, Rutledge *et al*⁴ reported hypointensities of the caudate and putamen in four choreatic patients. Several authors either did not report on intensity changes or explicitly did not find any. We report a case of rigid HD with a pattern of MRI findings, which has not been described previously.

At the age of 18 our patient developed clumsiness of her hands. There was a rapid progression with development of gait disturbance, dysarthria, loss of spontaneous movements, urinary dysfunction and hyper-salivation. When referred to our hospital at the age of 26 she had orofacial apraxia, aphonia, spastic gait, severe bradykinesia with bamboo pole-like posture, rigidity of all extremities, dystonia of the upper extremities with hyperextension of the fingers, abducted arms and flexed elbows. Tendon reflexes were exaggerated with bilateral Babinski signs. Blood smears were negative for acanthocytes. Four other members of the family were also affected by either choreatic or rigid movement disorders. Pathological investigations were not carried out on any of these cases. Mode of inheritance seemed to be autosomal dominant. Our diagnosis was rigid HD.

MRI (1.5 Tesla) in our patient showed bilateral caudate atrophy (figure). Additionally, on T2-weighted images (SE 2.1/90) there was mild hypointensity of the putamen and the globus pallidus, which did not involve the caudate. The same constellation was found in one of the male relatives, who was hyperkinetic and the only clinically affected family member that we were able to study.

Our findings do not agree with Savoiardo's study in three ways: 1) instead of hyperintensities we found hypointensity in the basal ganglia; 2) in our case the puta-