Epileptic seizures evoked by the Rubik's cube

Sir: The magic cube, invented by the Hungarian architect Ernő Rubik, is a three dimensional puzzle requiring the restoring of the scrambled coloured pieces of a $3 \times 3 \times 3$ cube to their proper positions. We report on a patient whose seizures were precipitated predominantly and consistently when playing with the Rubik's cube.

A 30 year old businessman had experienced sudden transient jerky movements of the body, the right arm in particular, since the age of 15 years. The jerks occurred when he concentrated on a problem; doing arithmetic, checking cash accounts, and making decisions on business matters, were some common situations. An activity which consistently produced the jerks was playing the Rubik's cube. Often the cube dropped out of his hands. The attacks were also associated with transient thought block. The jerks and the mental block made it difficult to proceed with a game. Following repeated jerks, on four occasions, he had become unconscious for periods lasting 15–20 minutes. After these “major” attacks he gave up playing the cube. Card games and draughts (checkers) which he played occasionally did not cause jerks as far as he could remember. Neither did other activities such as reading, writing or watching television.

He was taking phenytoin 300 mg/day regularly and diazepam 2–6 mg/day intermittently. With this treatment he had been free of major attacks for 4 years. The diazepam prevented the jerks but he did not take the full dose regularly because it made him drowsy. Instead, he had trained himself to disengage his mind from a problem and “let the mind go blank” when he developed a jerk. This avoided repeated jerks and possible major attacks. He was a right-handed man with normal intelligence. No abnormalities were found on physical examination.

The electroencephalogram (EEG) at rest (30 minutes) and on photic stimulation showed no abnormality. Hyperventilation (3 minutes) produced a few episodes of generalised bilaterally synchronous sharp and slow complexes. Playing the cube, within a minute, produced generalised synchronous symmetrical atypical spike and wave discharges of 3 Hz, each lasting 1–3 s (fig). Twenty such discharges and 11 minor spikes or sharp wave discharges occurred during the test period of 15 minutes. Myoclonic jerks corresponding with the EEG discharges were noted on six occasions. With each jerk the play was interrupted and the patient appeared dazed. He later volunteered that he had mental block many times during the play. Looking at the cube while turning it around, but not manipulating the pieces, did not produce any abnormality. The following tests were then carried out. Against each test is the number of epileptiform discharges recorded over a 15 minute period: written arithmetic 7, mental arithmetic 23 (jerks +), draughts 17 (jerks +), card game 11, reading 3, verbal test on reading material 3.

The basic seizure disorder in this patient is typical of “impulsive petit mal” or “juvenile myoclonic epilepsy”. The remarkable feature was the precipitation of seizures and EEG abnormalities by specific activities. Visual stimuli are known to trigger seizures in juvenile myoclonic epilepsy, about half the number of patients being photosensitive. However, our patient was not photosensitive. The experiment showed that playing the cube, not mere gazing at the colour pattern, was the important factor in producing dysrhythmia. Mental arithmetic, which did not involve any visual or peripheral stimuli, was also a potent trigger. These observations suggest that in this patient, the seizure-provoking stimulus lay in the higher cerebral functions, acting under very specific conditions. Epilepsy evoked by higher mental activities is a rare but recognised phenomenon. Reports are available of patients who develop epileptic seizures, including myoclonic jerks, during arithmetical calculations, decision making, and chess, draughts or card games. The three factors suggested by Forster complex decision...
making, sequential decision making, and stress, seem to be applicable to the situations under which our patient developed his seizures and EEG discharges.

The cube and draughts, two of the most potent seizure-evoking stimuli in our patient, involve processing of spatial information. It is noteworthy that in a previous case also, many of the epileptogenic tasks (arranging blocks to form a design, describing a letter outline, and drawing from memory) involved spatial processes. These observations could mean that the neural substrate for triggering seizures in these cases lies in the parietal lobe, in an area responsible for spatial processing and calculation.

In some centres, the standard EEG recording includes the performance of mental arithmetic. In our EEG laboratory, we now employ the Rubik's cube as one of the provocative tests in the investigation of seizures related to higher mental functions.

I thank Miss RMA Vijitha and Miss SM Weerasuriya, my EEG technicians for their assistance.

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References


Letters

Gilles de la Tourette's syndrome in monozygotic twins

Sir: Gilles de la Tourette's syndrome is a rare disorder with onset between 2 and 15 years of age. There is a characteristic waxing and waning of both vocal and motor tics which spread in a cranio-caudal direction. Coprolalia, echolalia, palilalia, echopraxia and behavioral, that is, mostly obsessive-compulsive abnormalities may additionally be present. Annual incidence rate has been calculated as 4-6 cases per 1 million inhabitants.

From the earliest description, familial aggregation has been noted. Genetic evidence now suggests an autosomal dominant trait with a preponderance of males. A continuum of phenotypes between chronic multiple tics of childhood, Gilles de la Tourette's syndrome and obsessive-compulsive disorder has been described. In a questionnaire-based study concordance rates for monozygotic twins with Gilles de la Tourette's syndrome and/or tics have been estimated to be 77%, and those for dizygotics 23%, which indicates a variable clinical expression of the trait.

The following report may help to elucidate further the phenotypic variations of Gilles de la Tourette's syndrome in monozygotic twins.

The twins, A and B, were of non-Jewish origin. There was no consanguinity. Appearance (hair, ear lobe, iris, general gestalt) indicated monozygosity which was substantiated by common blood group antigens in both parents and twins (A; CcDe; M +; P1; Lea+) and by HLA labelling in both twins (A2; A11; B27; BW58; CW2; CW3). Both had an unremarkable gestation and birth in July 1967. Birth weight was 3150 g in A and 3100 g in B. Details of the placenta were not obtainable. Both were reared together in the parent's home. In preschool age both were hyperactive and destructive. Developmental milestones were normal. From the age of 6 years both showed compulsive behaviour: for example, before sleep they had repetitively to control the opening of the bedroom door at a width of three fingers.

A first experienced "hair-out-of-eyes" tics at the age of 13 years. Later eye-blinking, vocal tics as snorting and sniffing, and compulsive swearing were occasionally observed.

B had an emergency tracheotomy because of pseudocroup in his first year. His first movements emerged already at the age of 12 also as "hair-out-of-eyes" tics which at times made him running in front of the mirror up to fifty times a day. Eye-blinking, facial grimacing as well as marked squeaking and screaming vocal tics were observed some months later. At the age of 16 throat-clearing, grunting, gurgling, and humming were noticed. Vocal tics sometimes interfered with normal speech towards a barking or sighing expression. States of restlessness occurred for 10 to 15 minutes while he was running through the home compulsively swearing in obscenities (shit, trash) and offending his relatives. From August 1985 shoulder jerks and piano movements, both more marked on the left were noted. Echolalia, mental and overt palilalia, mental and toilet coprolalia also occurred.

On clinical examination B was far more severely affected by the tics and showed a

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M*, monozygotic; E, eye; H, head; N, neck; S, shoulder; A, arm; L, leg; OC, obsessive-compulsive; np, not presented.
*Second twin not examined.
†No details stated.
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