Episodic dyscontrol syndrome

Amy McTague, Richard Appleton

Department of Neurology, Alder Hey Children's NHS Foundation Trust, Roald Dahl EEG Unit, Liverpool, UK

Correspondence to
Dr Amy McTague, Department of Neurology, Alder Hey Children's NHS Foundation Trust, Roald Dahl EEG Unit, Liverpool L12 2AP, UK; amy.mctague@alderhey.nhs.uk

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ABSTRACT

Episodic dyscontrol syndrome (EDS) or intermittent explosive disorder (IED) is a clearly identified category in the *Diagnostic and Statistical Manual of Mental Disorders (DSM IV)*. It affects children and adults. Children are often considered to have epilepsy or a mental health problem. The episodes comprise of recurrent attacks of uncontrollable rage, usually after minimal provocation, and may last up to an hour. Following an episode, children are frequently exhausted, may sleep and will usually have no recall. These features often raise the possibility of epilepsy, and children may be referred for an electroencephalogram (EEG). An abnormal EEG may subsequently lead to a diagnosis of epilepsy and the prescription of an anticonvulsant. Five patients with EDS are described, all referred to a single neurology clinic with a possible diagnosis of epilepsy, one of whom was being treated with an anticonvulsant. Four had an EEG, which was normal in two patients and showed "abnormalities" in two, leading to an incorrect diagnosis of temporal lobe epilepsy in one patient. All five children responded to psychological (behavioural) intervention. Recognition of the clinical features of IED/EDS should preclude the need for EEG and other investigations in these patients and prompt referral to psychological services.

Outbursts of rage or anger are common in children and they are often referred to exclude epilepsy or autistic spectrum disorder (ASD). It is important to appreciate that, as well as neurological or psychiatric disorders, a number of other conditions may present with rage, including episodic dyscontrol syndrome (EDS),[1 2] also called intermittent explosive disorder (IED).[3 4] Paediatricians may be unaware of this condition and request an electroencephalogram (EEG) to confirm or "exclude" epilepsy.

The purpose of this study is to describe a series of children with EDS or IED and outline the potential danger of misdiagnosis.

PATIENTS

Children were identified retrospectively from all new patient referrals to paediatric epilepsy or neurology clinics over a 12-month period. All patients were seen by a single consultant in paediatric neurology (RA). A retrospective case note review was undertaken in those children with a diagnosis of EDS or IED.

RESULTS

Of 149 new patients seen in the clinics, five (three boys and two girls, aged 7.5–10.3 years) were considered to have a diagnosis of EDS/IED. All five children were referred with a possible diagnosis of epilepsy, one from his General Practitioner and three with "temporal lobe epilepsy". All were born following normal pregnancies, with normal birth weights and uneventful perinatal periods. There was no family history of epilepsy. The mother of one of the female patients had moderate learning difficulties and adult-onset depression, but there was no other relevant family history. One boy was described as an irritable baby who slept poorly during the first 2 years of life. Early development was normal in two patients; one was thought to be a bit "slow" and two required speech and language therapy input. One boy experienced two brief febrile seizures at 15 and 17 months of age; none had experienced any epileptic seizures. None were described as having frequent temper tantrums in infancy or in nursery school. The boys presented between 6 and 7, and the girls between 7 and 8 years of age, with sudden outbursts of rage which were occasionally provoked but most were thought by witnesses to be unprovoked. The episodes lasted from 10 min to over an hour. A typical attack was described as follows: for either a trivial or no obvious reason, he would start to shout and scream, throw objects and purposefully attack others, and specifically teachers. At the height of the attack, he would be flushed and hypersalivate and would often need to be restrained. The episodes would last from 20 min to an hour, after which he would become pale, tearful and exhausted and would sometimes sleep. Following an attack, he had no recollection of what had happened and would be subdued for a number of hours. One male patient was suspended from school because of these episodes. Four of the five patients were referred to the community child health department for formal assessments for attention deficit hyperactivity disorder and two for ASD, all with negative results. Three patients were referred to the child and adolescent mental health service (CAMHS) and none were diagnosed with a mental health disorder. Four patients had an EEG and three a MRI brain scan. The EEG in one patient at 7 years of age demonstrated "bi-temporal slow wave activity"; this patient was diagnosed with temporal lobe epilepsy and commenced on sodium valproate. The EEG in another patient at 7 years of age was reported to be "immature". The EEG was normal in the remaining two patients undertaken at 9 and 10 years. MRI was normal in the three patients. Following review in the neurology clinic, sodium valproate was discontinued and all were referred to CAMHS with a diagnosis of probable EDS/IED. The children and their families were subsequently offered programmes to develop strategies to manage their rage; these strategies were shared with the school. The episodes have reduced in both frequency and severity in four

and have resolved in one. One patient has been placed with foster parents.

DISCUSSION

EDS, otherwise known as IED, is characterised by discrete episodes of aggressive behaviour which are severe, causing damage to people or property, and result from provocation, which may be minimal or even absent.[1–4] The episodes may be preceded by an anticipatory fear or sensory symptoms, including tingling of the body or limbs or hyperacusis,[2] and are often followed by extreme remorse or amnesia or both, and, occasionally, sleep. The episodes occur rapidly, are short-lived and are often directed against a friend or family member. The condition is often accompanied by other mood and behavioural difficulties, including of impulse control and inhibition.[5] Males are predominantly affected and the mean age of onset is 15 years. In younger children, episodes of rage are often regarded simply as temper tantrums, and there is often a clearly identified trigger or provocation. Although the five patients in this series were young, they clearly met the descriptive criteria for IED/EDS.

Numerous organic causes, including epilepsy, and specifically seizures arising from the temporal or, less commonly, the frontal lobes, are frequently considered to be responsible for outbursts of sudden rage or aggression and is often termed "ictal aggression".[6–8] However, the prevalence of how often EDS is misdiagnosed as epilepsy is unknown. One multicentre study of 5400 patients of varying ages reported that only 19 patients (0.4%) showed aggressive behaviour during seizures.[9] Rage as a manifestation of a partial (focal) seizure shows characteristic features. These include behaviour that is stereotyped, primitive, random, resistive and not usually directed against specific individuals.[7 9] Ictal confusion, resembling aggressive behaviour, may also be seen in the immediate postictal period following a focal or generalised tonic-clonic seizure. It is very rare for children to manifest ictal rage as the only type of epileptic seizure and most individuals with ictal rage will demonstrate more easily recognised epileptic seizures. Consequently, outbursts of sudden rage or aggression as an ictal phenomenon are rare and epilepsy must be diagnosed with extreme caution in these children.

Patients referred with outbursts of rage frequently undergo an EEG to either "confirm" or "exclude" epilepsy. Although during ictal (epileptic) rage there will usually be an electrical correlation on the EEG, this is not invariable, particularly if the seizures originate in the frontal lobes. In addition, movement artefact during the attack may mask any genuine ictal changes. The inter-ictal EEG in EDS/IED has been reported to show a range of abnormalities. In a study of 23 patients with IED,[10] the EEG was normal in 16 and showed diffuse or focal slowing in seven. Another study showed spiking or non-specific "abnormal" activity in the temporal region in 14 of 22 patients.[11] However, such findings cannot be used to make or confirm a diagnosis of epilepsy. One of our patients was diagnosed with epilepsy on the basis of bi-temporal slow wave activity, which may be a normal variant at this age. This emphasises the danger of using the EEG to either "exclude" or "confirm" a diagnosis of epilepsy. EEG should also not be used in the diagnostic work-up for EDS/IED.

Treatment may be pharmacological or psychological, or both. There have been few randomised controlled trials of treatment of EDS/IED. Antidepressants and mood-stabilisers including lithium, sodium valproate and carbamazepine have been used in adults, and occasionally in children with oppositional defiant disorder or conduct disorder to reduce aggression. Cognitive behavioural therapy (CBT) is effective in the treatment of anger. A recent trial[12] randomised adults with IED to 12 weeks of individual therapy, group therapy or waiting list (no therapy). Intervention resulted in an improvement in anger and aggression levels, with no difference between group and individual CBT.

Adolescents and young adults may experience educational and social consequences but also mental health problems,[4] including parasuicide,[13] if IED/EDS is undiagnosed in early childhood. Consequently, earlier recognition and treatment of IED/EDS could potentially prevent these complications, with obvious benefits for the individual and their community.

Our patients were initially considered to have possible epilepsy and one was treated with an antiepileptic drug on the basis of the episodes and a non-specifically abnormal EEG. All five patients were subsequently diagnosed with IED/EDS and have responded to psychological input. It is essential to be aware of this condition as a possible diagnosis in a child or young person presenting with outbursts of anger and aggression and offer appropriate intervention and support. An accurate and detailed history is crucial to establish the diagnosis and the EEG should not be used to exclude, or make a diagnosis of epilepsy.

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