

Epilepsy in adolescence

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Introduction

Although the incidence of epilepsy is high in adolescence and the prevalence of psychiatric disorder is also higher than in childhood or adulthood, specific services for adolescents with epilepsy are rarely provided and this subject receives surprisingly little attention in many books on epilepsy. Adolescence is a time of great change; growth into adulthood and issues such as preparation for university or employment, driving, drinking, social/sexual relationships, marriage/conception, as well as a general increase of responsibility need to be considered. Epilepsy impinges on all these areas to a significant if not major degree. In addition, adolescents tend to be very body-conscious and do not like to be different from their peer group. The stigma associated with a condition which implies loss of control and requires the regular taking of medication is liable to have a very negative effect on the adolescent unless the situation is managed well¹. Denial of the epilepsy may result in some teenagers taking risks, such as refusing to accept medication or other precautions.

The subject of epilepsy in adolescence has been covered in a number of reviews²⁻⁶.

Management dilemmas

There are some specific management dilemmas in adolescents with epilepsy. Although the focus of both the initial interview and follow-up discussions should be on the adolescent, because the history depends so much on the account of someone who has actually seen the seizures it is generally also necessary to interview the parents. This situation needs to be explained to the adolescent.

Sodium valproate remains one of the most effective antiepileptic drugs for juvenile myoclonic epilepsy (JME). However, it may be associated with weight gain – a particularly unfortunate adverse effect in body-conscious female patients, who may refuse to continue taking the drug. There has been considerable debate in the literature about the apparent association of polycystic ovary syndrome with this drug (see Chapter 44 on epilepsy in women). In addition, the increased risk of neural tube defects, valproate syndrome and cognitive deficits in children born to mothers taking valproate are issues of concern.

The dilemma of declaring epilepsy on job/college applications may need to be discussed. Although it is important to be honest with a prospective employer, the declaration of epilepsy may prevent short-listing. One option is to leave the appropriate place on the application form blank and, after the candidate has been interviewed and the job has been offered, declare the epilepsy in a positive way. This allows the applicant to explain how the epilepsy should not interfere significantly with their ability to carry out the duties required, and to indicate what measures would be needed if a seizure were to occur at work.

The broad area of 'independence versus safety' is difficult for an individual who is trying to establish independence and a smooth transition to adulthood but may need to rely on others to some extent to maintain safety. The specific issue of drowning in the bath must always be discussed in this context. The issue of independence versus safety also impinges on a number of other areas.

Adolescents do not like being told what to do. The doctor should try to avoid giving advice but should instead encourage questioning and provide information, emphasising that the individual is in control of his or her own life. The following are suggested guidelines:

- If possible, talk to the adolescent first, ignoring the parents initially.
- Ask the adolescent to introduce the parents to you.
- Explain to the adolescent what will happen during the consultation.
- Present talking to the parents as 'a necessary evil' and explain why this is necessary to the adolescent.
- Write to the adolescent rather than the parents.
- Ask the adolescent's permission to send copies of letters to the parents.

In addition the following practice points should apply:

- Check the diagnosis.
- Characterise the syndrome.
- Provide accurate prognostic information.
- Treat with appropriate medication.
- Provide information on the following:
 - high risk of the unsupervised bath
 - effect of irregular sleep
 - alcohol
 - driving
 - sport
 - employment
 - contraception
 - genetic implications
 - advantages/adverse effects of specific antiepileptic drugs (AEDs).
- Listen, discuss, inform; avoid giving advice.

Diagnosis

There are several epilepsy syndromes which should not be missed. The following may present in adolescence:

- Juvenile myoclonic epilepsy (JME)
- Juvenile absence epilepsy (JAE)
- Epilepsy with generalised tonic-clonic seizures on awakening
- Benign partial seizures in adolescence
- Photosensitive epilepsy
- Reading epilepsy
- Subacute sclerosing panencephalitis
- Epilepsy from cortical brain tumours.

In addition, seizures from substance abuse or from neuronal antibodies should be considered.

Juvenile myoclonic epilepsy (JME)

JME is an idiopathic generalised epilepsy syndrome with age-related onset, commonly between the ages of 12 and 18 years. The sex distribution is equal. Bilateral, single or multiple irregular myoclonic jerks occur, mainly in the upper limbs. Most of the patients who present for treatment also have tonic-clonic seizures and many have absence seizures. The tonic-clonic seizures predominantly occur soon after awakening.

Patients often present with a history of one or more episodes of tonic-clonic seizures on awakening. The doctor should always ask specifically about morning myoclonic jerks, which, if not severe, may be viewed as slowness or clumsiness. Specific enquiry should also be made about 'blank spells'. Patients often do not declare myoclonic jerks or absence seizures because they do not realise that these are epileptic seizures but if this information is not available a diagnosis of JME is likely to be missed. It is important to diagnose this condition because most cases respond very well to sodium valproate. This often needs to be continued long term even if the patient is seizure free for years, since the chance of relapse is high if sodium valproate is stopped. The addition of lamotrigine may be effective in those patients who do not respond adequately to monotherapy with sodium valproate. Levetiracetam has been shown to be effective in treating the myoclonic seizures in JME and has a licence for this indication.

Juvenile absence epilepsy (JAE)

The onset of JAE is typically between the ages of 10 and 17 years, with males and females equally affected. Subjects are usually neurologically normal and a family history of epilepsy is common. The photosensitivity rate is high. Over 80% also have generalised tonic-clonic seizures. The absence seizures usually respond well to treatment with anti-absence medication such as sodium valproate, ethosuximide or lamotrigine.

Epilepsy with generalised tonic-clonic seizures on awakening

The peak onset of this syndrome is around puberty. Seizures occur exclusively or predominantly soon after awakening from sleep at any time of the day, with a second seizure peak during evening relaxation. Seizures may be precipitated by sleep deficit, excessive alcohol or sudden arousal.

There is some overlap between these three syndromes. This has generated much discussion on the way in which these three generalised epilepsies should be classified^{7,8}.

Benign partial seizures in adolescence

This syndrome needs to be distinguished from the very different syndrome of benign partial epilepsy of childhood. Age of onset is between 10 and 20 years, with a peak around 13–14 years. It is more common in boys. There is usually no family history and no cognitive or neurological impairment. The subject has simple or complex partial seizures, which can be secondarily generalised. There may be a cluster of 2–5 seizures in a 36–48 hour period. The patient may have only one episode of either a single seizure or a single cluster of seizures. The EEG is typically normal or shows only mild abnormality, in contrast to the syndrome of benign partial epilepsy of childhood, in which centrotemporal (rolandic) spikes occur. Because benign partial seizures in adolescence often present with only one seizure or a cluster of seizures, treatment should be avoided unless there is a recurrence or unless there are particular reasons for treating.

Photosensitive epilepsies

These are more common in adolescence. They are most often detected around 12–14 years of age, although careful history-taking may elicit an earlier onset. Two-thirds of subjects are female. The photosensitive epilepsies do not constitute a single syndrome. It is always important to define the syndrome in which the photosensitive epilepsy occurs, such as JME or JAE, so that specific information on treatment and prognosis can be given.

Reading epilepsy

This is a rare, benign form of epilepsy with a mean age of onset of 17–18 years. It is more common in males. There is a strong genetic predisposition. Diagnosis is confirmed by the very characteristic motor/sensory aura: after reading for a period, abnormal sensations or movements occur (with full consciousness), involving the tongue, throat, jaw, lips and face. If the patient does not stop reading, this aura may progress to a tonic-clonic seizure. If the subject stops reading when the aura occurs, tonic-clonic seizures can often be avoided and treatment with AEDs may not be necessary. If treatment is given then sodium valproate appears to be the drug of choice. The inter-ictal EEG is usually normal.

Subacute sclerosing panencephalitis (SSPE)

This condition, which typically follows measles infection very early in life (under two years of age), usually presents in either in late childhood or in the teenage years with relentless deterioration and eventual death. Initially there may be subtle loss of intellectual ability but myoclonic jerks or more complex abnormal movements soon become evident and the ensuing dementia is all too obvious. The EEG pattern is characteristic, with a discharge in all the leads when each jerk occurs. Measles antibody is raised in blood and is high in cerebrospinal fluid (CSF).

Epilepsy from cortical brain tumours

Cortical brain tumours can occur at any age. Because of this, serious consideration should be given to neuroimaging of adolescents who present with partial seizures. The exception would be those who have characteristic benign partial seizures with a single seizure or cluster of seizures, no abnormal neurological signs and no recurrence.

Investigation

The investigations of epilepsy in adolescence are similar to those at other ages. Basic blood tests for full blood count, creatinine and electrolytes, calcium, and liver function tests should be performed. An EEG with photic stimulation should be obtained. Neuroimaging should be considered but will not be necessary in those conditions which are obviously benign, as described above. In selected cases a urine toxicology screen for substance misuse or testing for neuronal antibodies may be appropriate. The latter may be particularly indicated if the presentation is unusual, especially if there are psychiatric symptoms or other signs of limbic encephalitis¹⁰ (which may or may not be detectable on MRI scanning).

Treatment

It is very important not to group the epilepsies of adolescence together as a single entity. For example, benign partial seizures in adolescence should not be treated whereas treatment of JME with sodium valproate is strongly recommended and often needs to be continued long term.

The mainstay of treatment is with AEDs. First-line drugs, such as carbamazepine and sodium valproate, should generally be used. Sodium valproate is usually effective for JME and for absence seizures. However, valproate is also associated with a risk of neural tube defects and impaired verbal IQ in offspring of mothers who take this drug during pregnancy. It is also

associated with weight gain, endocrine problems and fertility problems. Lamotrigine and levetiracetam are being used increasingly as a first-line drugs. They have a wide spectrum of action and are well tolerated. They are not associated with the weight gain and endocrine problems reported with sodium valproate. However, oestrogen decreases lamotrigine blood levels and this interaction may cause problems with oral contraceptives and during pregnancy. For those adolescents with seizures of partial onset who either cannot tolerate the adverse effects of AEDs or refuse to take them, self-control of seizures can be offered. This method may be effective in suppressing at least a proportion of partial seizures, especially those which are heralded by a clear aura.

In treatment-resistant seizures the possibility of non-epileptic attacks must always be considered and should be managed appropriately with a positive, non-punitive attitude. The concept of 'locus of control' is important. An approach which is often helpful is to say: 'Wouldn't it be wonderful if you were in control of the attacks instead of the attacks being in control of you?' The adolescent should be encouraged to find a way of controlling the attacks. He or she should be reviewed after a specified period of time, for example three weeks. If there is any reduction in the frequency of the attacks he/she should be praised for having done so well and for having begun to gain control themselves. Sometimes a change of life situation may be necessary.

The possibility of seizures precipitated by substance abuse must also be considered, although screening of patients in accident and emergency departments has shown that this is a relatively uncommon cause of presentation with seizures. A number of substances may be associated with the precipitation of seizures in people who do not necessarily have epilepsy. If substances such as alcohol or benzodiazepines are used in large intermittent doses ('bingeing'), then withdrawal effects may precipitate seizures. Seizures as a result of cocaine toxicity have been reported in a number of publications⁹. Ecstasy may also precipitate seizures. If substance abuse is suspected then a urine specimen should be sent for toxicology testing. Hair testing may also be useful in this context. Testing of hair is not of value in the acute situation but can be helpful in determining whether substances have been abused in the recent past and may offer some temporal indication of when the substance misuse took place. Treatment of the underlying substance abuse, rather than the prescription of an AED, is appropriate in these cases.

If the cause of the seizures is neuronal antibodies, for example anti-NMDA receptor antibodies or voltage-gated potassium channel complex antibodies, prompt treatment with immunotherapy can be curative and can also treat, prevent or minimise additional complications.

Surgery may be indicated in a number of circumstances. The most obvious of these is a tumour presenting de novo in adolescence. Some teenagers may have had a history of complex partial seizures for many years and MRI scanning may reveal mesial temporal sclerosis, a dysembryoplastic neuroepithelioma or a hamartoma. It could be argued that these patients should have had surgery earlier. If surgery is necessary, it is probably better to carry this out sooner rather than later. The longer the seizure disorder affects education, development and the social situation, the more difficult it will be to overcome the adverse effects of the epilepsy, even if the seizures themselves are controlled.

Conclusions

Adolescence is an exciting but uncertain period. If epilepsy presents for the first time in adolescence, this adds greatly to complexities of this period. Well-established epilepsy may vary over the course of adolescence, increasing the uncertainty when so many other changes

are taking place. When managing epilepsy in adolescence it is important to consider specific syndromes and causes because these may require very different styles of treatment or management. It is also important to consider the impact of epilepsy on the life of the adolescent, and to minimise the isolation and stigmatisation that the teenager may feel at a time when being part of an approving peer group is so important. These factors, together with the issues such as alcohol, driving, sport, contraception, genetic implications and ‘safety versus independence’, imply that the management of epilepsy in adolescence requires skill and sensitivity.

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