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PERSONAL PRACTICE

Teenagers with epilepsy

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Recent attention has focused on the importance, but inadequacy, of adolescent medicine,[1] and the paucity of medical services for this specific population.[2] Adolescence is, in itself, a difficult and traumatic time; when complicated by epilepsy it poses a great challenge not only to the young people themselves, but also to their carers and physicians.[3 4] This paper provides a guide to the management of the teenager who has epilepsy.

Establishing a correct diagnosis

The adolescent period is an important time to review the diagnosis of both epilepsy (differentiating it from other paroxysmal but non-epileptic disorders/conditions) and the epilepsy syndrome, and to consider any underlying cause. Common misdiagnoses at this age include vasovagal attacks, migraine with aura ("classic" migraine), non-epileptic (pseudo-epileptic) attacks, and substance abuse (including recreational drugs). Cocaine, heroin, and 3–4 methylenedioxymethamphetamine ("ecstasy") are known to cause convulsions; cannabis is less likely to have this effect.[5 6] The false diagnosis rates of epilepsy and the epilepsy syndrome could be at least 10% and 6%, respectively.[3] Juvenile myoclonic epilepsy is the most commonly unrecognised and misdiagnosed epilepsy syndrome; a history of myoclonic seizures, which are the defining seizure type in this syndrome, needs to be sought specifically because teenagers either fail to recognise them or consider that the early morning jerks are a normal part of waking up.[7] Brain tumours and temporal lobe epilepsy (caused by hippocampal atrophy/mesial temporal sclerosis) may present at this time. Finally, the onset of seizures in adolescence may, rarely, herald the onset of a neurodegenerative disorder including subacute sclerosing panencephalitis, Unverricht-Lundborg or Lafora body disease (both progressive myoclonic epilepsies), or juvenile Huntington's disease.

Repeat electroencephalogram (EEG) monitoring and neuroimaging with magnetic resonance imaging may be indicated, particularly if seizures have an onset at this time, if seizures change in frequency or character, or if seizure control remains poor.

In girls, catamenial epilepsy may develop in adolescence and might manifest as either an increase in seizure frequency or, less commonly, as seizures that occur only at or around the time of menstruation.[8 9]

Establishing the correct diagnoses of the seizure type(s), the epilepsy syndrome, additional neurological impairments, and any underlying cause is particularly crucial in the teenage years, in view of the imminent entry into adult life. Incorrect diagnoses, at any level of the diagnostic process, may have important and potentially serious implications for employment, driving, and psychosocial health.

The teenager with additional neurological impairments

Many teenagers with epilepsy dating from early childhood will have additional and occasionally major neurological impairments. These may include motor disorders (such as hemiplegia or quadriplegia), learning difficulties, and psychiatric/behavioural problems (such as autism and attention deficit disorder). These impairments are commonly associated with the more malignant epilepsies of childhood.[10] Some patients will require lifelong care and support from many disciplines and others will be able to develop a degree of control over their lives, although with continuing support from their parents and carers.

Choosing the most appropriate antiepileptic drug

The correct identification of the seizure type(s) and epilepsy syndrome is important in helping to prescribe the most appropriate antiepileptic drug (AED), predicting the likely response (in terms of seizure control), and predicting the risk of seizure recurrence if the drug is discontinued. Carbamazepine and vigabatrin exacerbate myoclonic and typical absence seizures,[11 12] including precipitating absence status, and should be avoided in syndromes characterised by these seizure types (for example, juvenile myoclonic epilepsy and childhood or juvenile onset absence epilepsy). Sodium valproate and lamotrigine have a relatively broad spectrum of action against most seizure types, although lamotrigine may not be as effective as sodium valproate in suppressing myoclonic seizures. Gabapentin and topiramate are of benefit in treating partial and secondarily generalised tonic clonic seizures, although their role in treating other seizure types and epilepsy syndromes is not yet clear. True catamenial epilepsy can be treated by a

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number of regimens. For teenagers already taking an AED, a temporary increase in the dose of this AED or the additional, intermittent use of clobazam or acetazolamide for the three days before menstruation, as well as throughout menstruation, may be useful; for teenagers who only experience seizures at menstruation, intermittent clobazam or acetazolamide, or perimenstrual progestogens, may be prescribed.

The prescription of an AED must also take account of the side effect profile, because of both patient safety and compliance with medication. Although sodium valproate is effective in most seizure types and epilepsy syndromes of adolescence, it may be associated with a number of side effects that limit its usefulness and "acceptance" by this age group, particularly girls. These include increased appetite and weight gain, transient hair loss, menstrual irregularities (including primary/secondary amenorrhoea and the polycystic ovary syndrome, which may, rarely, lead to infertility),[13] and a distal tremor. The continued use of sodium valproate in adolescent girls should be considered on a risk to benefit basis.

The identification of a potentially surgically treatable lesion is clearly important because any surgery might result in a "cure" for the teenager's epilepsy and, as a consequence, it might prevent or at least reduce the social, educational, and behavioural sequelae of continuing seizures.

Specific issues

Teenagers with epilepsy are frequently caught between paediatric and adult services with neither service being able to understand or satisfy their specific needs and concerns.[4]

Pregnancy and contraception are important issues—for both sexes. In certain situations, particularly if the teenager is accompanied by her boyfriend rather than her parents, it may be prudent for clinic medical or nursing staff to raise the issues of contraception and pregnancy directly. The choice of contraceptive and, in particular, the oral contraceptive should be discussed, including the fact that some AEDs may reduce its effectiveness, thereby necessitating a higher strength of oestrogen to try to "ensure" safe contraception.[14] For young women on enzyme inducing AEDs (carbamazepine, ethosuximide, phenytoin, and topiramate) wishing to take a combined oral contraceptive, the starting dose should be 50 μg of ethinyl oestradiol, but doses of 75 or 100 μg might be required if breakthrough bleeding occurs. Preconception planning must be emphasised, including optimising the number, dosage, and blood levels of AED(s), recommending daily folate supplementation,[15] ensuring early antenatal booking, and monitoring of the fetus throughout pregnancy, preferably in a fetal centre. Ideally, any change in medication should be completed before conception, with monotherapy as the objective. Folic acid supplements (a minimum of 0.4 or a maximum of 5 mg/day) should be taken by any young woman with epilepsy who is sexually active, irrespective of whether a pregnancy is being considered, because at least 30% of teenage pregnancies are unplanned. Although folic acid supplementation has not been shown to reduce the risk or incidence of neural tube defects in children born to mothers who take sodium valproate, it is reasonable to extrapolate from data that have shown that folic acid has a protective role in the prevention of neural tube defects.[15 16] There is an increased risk of teratogenicity in women with epilepsy. The aetiology is complex and encompasses demographic and genetic factors as well as environmental factors, including AEDs and seizures.[17] As yet, there is no conclusive evidence for a difference in teratogenetic risk between most of the AEDs. Sodium valproate taken during the first trimester is associated with a 1–3% risk of neural tube defects, some 10–20 times higher than the background incidence of 0.2–0.5%; urogenital and cardiovascular malformations may also occur.[18] More minor defects, including facial abnormalities and delayed development, might also complicate the use of valproate or phenytoin taken throughout pregnancy. Higher doses of valproate (> 1000 mg/day) are thought to carry a higher risk, possibly because of high peak concentrations.[19] Carbamazepine may also be associated with neural tube defects and microcephaly. The new, established AEDs (gabapentin, lamotrigine, and vigabatrin) appear to be safer but human data are currently very limited.

Teenagers with epilepsy may apply for a driving licence and drive providing that they have not experienced a seizure of any type for one year, or have had only nocturnal seizures for three years.[20] It is also important to remind them that if they are already driving and an AED is to be withdrawn, they should stop driving at the time that the drug is withdrawn and for a period of six months thereafter.

Alcohol is another important issue. Total abstinence is unnecessary but more than two units of alcohol a day is likely to increase the risk of seizures in patients with epilepsy by a number of mechanisms including a lowered seizure threshold, an increased metabolism of the AEDs or a disruption of "normal" sleep patterns.[21 22] Teenagers should also be informed that a relative lack of sleep, for whatever reason, may also lower the threshold for further seizures.

The choice of employment and the practicalities of how to apply for jobs are frequently of concern to the teenager with epilepsy. It is important to stress that most careers or jobs are available and can be undertaken by people with epilepsy, but it is equally important to outline those jobs that are not currently available—specifically the armed and uniformed services and drivers of public transport. This will provide the teenager with accurate, realistic information and prevent, or at least reduce, any future disappointment. When applying for jobs, a specific recommendation is to suggest that the health section in the application form is left blank, but that epilepsy must be subsequently disclosed if the teenager is offered the job. This approach is an attempt to prevent the not uncommon discrimination against

people with epilepsy at the initial stages of job selection. Potential employers should also be provided with the name and address of the relevant hospital consultant.

The withdrawal of AEDs

The withdrawal of AEDs should always be considered in the teenager whose seizures appear to be well controlled. Although adolescent onset epilepsy appears to carry the highest risk of relapse,[23] serious consideration should be given to discontinuing treatment in this age group, specifically to determine whether teenagers can stop taking treatment before they are living independently, driving, or working. Not all teenagers will need to remain on treatment for the rest of their life. It is important that the adolescent is given accurate information and facts about the risks of seizure relapse if the anticonvulsant is withdrawn. This again emphasises the need to have diagnosed the epilepsy syndrome accurately, because different syndromes may be associated with greatly different relapse rates—for example, juvenile myoclonic epilepsy is associated with a high relapse rate (70–75%), whereas the benign partial epilepsies in adolescence tend to have very low relapse rates after withdrawal of treatment. It must also be explained that any treatment should be withdrawn gradually, over at least six to eight weeks.[24]

The giving of information

Teenagers and their parents must be given adequate and accurate facts and information including the type of epilepsy syndrome, specific issues, and any AEDs that are prescribed. The rationale behind using an AED should be explained, in addition to the potential consequences if it is not taken regularly, or stopped suddenly, and any likely side effects, including possible interactions with the other drugs (specifically, the oral contraceptive, alcohol, and recreational drugs). Based on this information, the teenager and his/her parents should come to their own decision regarding whether treatment should be taken, and which drug should be used. Although it is often tempting to offer the teenager advice or specific "recommendations", this may be perceived or misinterpreted as being directive or giving an order; advice or suggestions should therefore be offered only if specifically requested.

Doctors are often seen as figures of authority. In view of this and the fact that epilepsy is not just about having seizures or taking medication, other disciplines should be involved in talking to and "counselling" teenagers. Such disciplines could include psychology or nursing and specifically, nurse specialists. Nurse specialists have an important role in this area and may be considered by the teenager to be less authoritative and therefore more approachable; nurses are also more able to undertake home visits, the benefits of which are obvious.[25]

The process by which children take over responsibility for their medical care should ideally be begun well before the teenage years. They should always be included in the discussions and consulted in a way that is appropriate for their maturity. This approach should facilitate a more gradual passage from childhood into adolescence and, subsequently, into adulthood. All health professionals involved with teenagers must appreciate that adolescence is a time of self identification, a wish to become independent, and empowerment. Epilepsy can be a disempowering condition and can militate against achieving independence, with important social, educational, and behavioural/emotional consequences. These issues must be sensitively approached to allow the teenager to pass successfully from childhood into maturity and to enable the teenager to feel in "control" of their epilepsy as well as their life.

Whenever possible, verbal information should always be supported with written or audiovisual information. Teenagers should also be given the name, address, and telephone number of an appropriate voluntary epilepsy organisation so that they can make contact themselves, in their own time, and obtain the most relevant information for themselves.

The families and carers of the teenager

The families and carers of teenagers often need considerable understanding, support, and encouragement in allowing young people to develop their own personality and independence. Inevitably, there will be areas of conflict, with many parents tending to be overprotective and restrict their child's demand for freedom. These concerns and the families' needs must be considered with tact and support, so that both parties can be encouraged to take a sensible and realistic approach to the risks of everyday life.[26]

Follow up

Most teenagers or adolescents find it uncomfortable or inappropriate to continue attending a paediatric clinic. Teenagers who are likely to remain on antiepileptic medication for a number of years (irrespective of seizure control), should continue to have access to a specialist epilepsy service, although their day to day management should be undertaken by their general practitioner. This "specialist" service could be sited within a specific teenager/adolescent epilepsy clinic (supervised jointly by a paediatric and adult neurologist[3]) or in an adult epilepsy clinic. The timing of referral to a teenager or adult epilepsy service will largely depend on the individual but should be considered from the age of 14 or 15 years. Importantly, a coordinated and multidisciplinary approach should be adopted, whichever clinic is felt to be most appropriate. For the teenager who is multiply disabled, it is important that the team includes individuals with the skills of developmental neuropsychiatry and of counselling people with learning difficulties. Failure to provide the teenager with continuing specialist epilepsy services may have potential medical and psychosocial consequences. Importantly, teenagers must always be given the opportunity to talk to the medical

or nursing staff alone, and without their parents.

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