TAKING OVER EPILEPSY FROM THE PAEDIATRIC NEUROLOGIST

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The problems of coping with adolescence are greatly increased by an additional chronic disorder. This is particularly so for those with epilepsy, where there is social stigma and the need for potentially sedative and damaging medication. Epilepsy is the most common neurological condition among patients transferred from paediatric to adult care. Many of the problems encountered in the teenager with epilepsy are shared by those with other chronic disabilities.^{1 2}

This review considers the paediatric environment that the adolescent is leaving, the rationale for a teenage clinic, and the facets of this which make it a desirable facility.

THE TEENAGER WITH EPILEPSY

Even when relatively mild, the impact of epilepsy on the teenager and on other members of the family may be profound. Epilepsy has important differences from other chronic disorders, particularly that the disability is intermittent, and so can be hidden. The perceived need to conceal epilepsy, and concern about seizures occurring in public, can induce great anxiety. The consequences of missing epilepsy, or treating it inappropriately, can be disastrous in terms of impaired education, employment, and driving prospects, unnecessary medication, and diminished self esteem. Perhaps these factors are of greater importance for teenagers than for those in other age groups:

- ▶ Developing independence is impeded, education is at a critical stage, employment and driving prospects are just being realised, and issues of relationships, contraception, pregnancy, and parenthood are becoming increasingly relevant.
- ▶ Deviations from peer group norms hold great importance for teenagers: epilepsy can be disastrous for self esteem and self identity.
- ▶ Social handicaps acquired in childhood and adolescence can remain with the individual even after seizures have been adequately controlled.

Complete control achieved before adult social and vocational responsibilities can help to avoid these problems. A relatively aggressive approach to treatment, including early consideration of epilepsy surgery, is therefore justified.

THE TEENAGER CLINIC

Taking over a patient's care offers opportunities to reassess the diagnosis by reviewing the history, and to ensure appropriate investigations are to hand. This is best done in the setting of a specialist teenager epilepsy clinic. Developing teenagers often find paediatric clinics embarrassing and inappropriate. A transitional arrangement offers the benefit of consulting both an adult and a paediatric neurologist, supported by an epilepsy specialist nurse.³ The general principles of a teenager epilepsy consultation are listed in box 1.

Two main patient groups are referred to this type of clinic:

- ▶ Patients presenting with blackouts beginning as teenagers (referred from general or hospital practice) where the main initial issue is diagnosis.
- ▶ Patients with ongoing and often complex epilepsy from childhood (referred from general paediatrics or paediatric neurology) where the main issue is ongoing management of the epilepsy.

The following should be addressed at first contact:

- ▶ Is the diagnosis of epilepsy correct?
- ▶ If the diagnosis is epilepsy, which type is it?
- ▶ Is the current epilepsy management appropriate?
- ▶ Is there appropriate knowledge and understanding of the condition, its prognosis, and its lifestyle implications? In particular, will the seizures interfere with completion of education, training for a career/job, or qualification for a driving licence, and are there special social circumstances that need to be addressed?

IS THE DIAGNOSIS OF EPILEPSY CORRECT?

A correct diagnosis is essential for the appropriate management of any patient with blackouts. Despite major advances in the technology for investigating blackouts, the most important investigation remains the taking and re-taking of the clinical history.

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Box 1: Principles of consulting with teenagers with epilepsy

- ▶ See the teenager in a clinic setting with other teenagers or adults
- ▶ Focus the consultation on the teenager rather than the parents—for example, invite the teenager to introduce their parents or carer
- ▶ Discuss with the teenager adult topics such as alcohol, driving, pregnancy, and contraception
- ▶ Speak to the teenager alone during the consultation; an opportunity arises if the physical examination is conducted in another room
- ▶ Give the opportunity to speak to an epilepsy specialist nurse; like many adults, teenagers often open up to a nurse more than to a doctor
- ▶ Offer written material on relevant aspects of epilepsy. Sending copy letters to patients empowers them and acts as continuing education and encouragement
- ▶ Encourage carers to allow the teenager an appropriate amount of responsibility—for example, for his or her own tablets

Blackouts in teenagers may be caused by attacks that are not epilepsy (usually syncope, psychogenic non-epileptic attack disorder, or migraine), by epilepsy persisting since childhood, or by epilepsy presenting for the first time in the teenage years.

IF THE DIAGNOSIS IS EPILEPSY, WHICH TYPE IS IT?

Several epilepsies present first during adolescence. These include idiopathic generalised epilepsies (such as juvenile myoclonic epilepsy, and, less commonly, juvenile absence epilepsy and generalised tonic-clonic seizures on awakening), as well as idiopathic partial epilepsies such as late onset benign occipital epilepsy.

There are three broad groups of teenagers with epilepsy:

- (1) Those whose epilepsy could be temporary.
- (2) Those whose epilepsy is likely to be life long, and in whom there are no significant intellectual or physical problems.
- (3) Those whose epilepsy is associated with additional physical and/or learning problems.

The adult neurologist is most likely to become involved with the second of these groups.

Epilepsy which could be temporary

The likelihood of these teenagers being passed on to the adult neurologist will depend on the age at which transfer takes place. There are two main groups where remission of epilepsy is likely before, or at, puberty.

- ▶ Children with classical, typical, childhood absence epilepsy, with no other seizure type, are unlikely to continue to have epilepsy or any other neurological problem.
- ▶ Those with benign partial epilepsies of childhood, particularly benign partial epilepsy with centro-temporal spikes (BECT), are also likely to remit. When a typical history of simple partial seizures involving the face and arm, arising from sleep, together with characteristic EEG changes allows a diagnosis of BECT, medication can safely be discontinued. Almost invariably these teenagers will be of normal intellectual ability, but a small percentage have learning difficulties that may be specific—for example, language and/or reading difficulties. Some young adults who have had BECT might realistically aspire to obtaining a class 2 driving licence or joining the armed forces. Those with

benign occipital epilepsies may not remit,⁴ but continued treatment with carbamazepine is likely to control their seizures.

Epilepsy without significant intellectual or physical problems

Most of this group have idiopathic generalised epilepsies. The main problems lie with the diagnoses of the more atypical forms of absence epilepsy and with acknowledging the importance of photosensitivity.

- ▶ Juvenile absence epilepsy, and the syndromes of myoclonic absences, eyelid myoclonia with absences, and absences with perioral myoclonia, do not remit in adolescence. Since the EEG patterns are not readily distinguishable from those of typical childhood absence epilepsy, these diagnoses will be missed unless a careful history, or a video recording, of the clinical features is obtained. Continued untreated absences may be dangerous for the independent teenager, and for other members of the public, should a driving licence be obtained.
- ▶ Juvenile myoclonic epilepsy can go unrecognised for long periods before a generalised tonic-clonic seizure occurs. This diagnosis is important, since lifestyle measures, such as avoidance of sleep deprivation and overindulgence with alcohol, can make significant contributions to seizure control; it is associated with photosensitivity in a high proportion of cases.
- ▶ Photosensitivity, although tending to be less prominent with increasing age, does not remit.

Epilepsy with physical and/or learning problems

It is probable that, in the regular paediatric clinics, attention to general needs, as well as to epilepsy, will have been coordinated. Involvement of carers is likely to continue to be necessary, but perhaps in a less strictly parental sense. Many teenagers and their carers have difficulties with adjusting to these aspects of transfer to an adult service.

IS THE CURRENT EPILEPSY MANAGEMENT APPROPRIATE?

The principles of epilepsy management in teenagers are similar to those for adults, though the consequences of continued seizures or of cognitive side effects of medication are potentially more serious.

Antiepileptic drug treatment

The type of epilepsy and the patient's sex are the main factors influencing the first choice of antiepileptic drug (AED) treatment. The main principle is to prescribe the lowest effective dose of a medication appropriate to the type of epilepsy. Preferably, the medication should be one with few side effects, and required to be taken only once or twice daily (thereby avoiding taking tablets to school or work). In order to prevent breakthrough seizures, the prescribed dose might need to increase over time to match the rapid physical changes of adolescence.

Medication side effects

In childhood and adolescence, even mild cognitive dysfunction—for example, from inappropriate AEDs—may permanently harm educational and employment prospects. Cosmetic effects of phenytoin, its interactions, and the need for blood monitoring make it a poor choice for first line treatment in young people, particularly women. Weight gain is sometimes a prominent side effect of valproate and this may limit its usefulness. Young women taking AEDs must be

Box 2: Factors influencing antiepileptic drug compliance in teenagers

- ▶ Compliance at any age may be impeded by:
 - denial of the epilepsy
 - concern over side effects, including possible teratogenicity
 - complacency about good seizure control, paradoxically resulting in poor compliance and seizure recurrence
- ▶ Compliance in teenagers is further influenced by:
 - peer pressure not to be different: with each dose, tablets remind them that they are different
 - rebellion against parental involvement in the management of their epilepsy

warned of potential interactions with the contraceptive pill. More importantly, they must appreciate the possible teratogenicity of their medications (this is discussed in detail below).

Teenagers with epilepsy are likely to need to take AEDs for many years to come. Long term problems with older AEDs such as phenytoin (peripheral neuropathy, ataxia) derive from 60 years prescribing experience. The long term side effects of the newer AEDs (if any) are largely unknown. However, the delays in identifying certain major risks from newer drugs—for example, teratogenicity from valproate or visual field constriction from vigabatrin—give no reassurance on the long term safety of the range of new AEDs, and emphasise the need for constant vigilance.

Medication compliance

Factors influencing AED compliance are shown in box 2.

Epilepsy surgery

Adolescence is perhaps the ideal time to consider epilepsy surgery, especially for medication resistant temporal lobe epilepsy. Surgical outcomes are generally better at a younger age. Furthermore, teenagers, to a much greater extent than children, can be involved in the decision and contribute to the management plan.

Partners in care

Teenagers with epilepsy can be encouraged to participate in and even lead discussions and decisions about their clinical management. Patients with epilepsy (as with other chronic conditions) may be considered “experts” in their own right as they often have acquired the life skills to cope with a chronic condition, and have the potential to become confident partners with professionals in their care.⁵

IS THERE APPROPRIATE KNOWLEDGE AND UNDERSTANDING OF EPILEPSY AND ITS IMPLICATIONS?

Teenagers require clear and relevant information about epilepsy, its management, prognosis, and its many lifestyle consequences.

Prognosis

The prognosis of the epilepsy should be discussed. Paediatricians tend to be unrealistically optimistic about some of the idiopathic generalised epilepsies, and do not always recognise the possibilities of benign outcomes for some of their patients who have focal epilepsies.

It is particularly important to recognise that issues covered in previous consultations with the parents must be re-explored with the teenager. This applies especially to genetic issues, where the diagnosis could influence future family planning decisions.

Restrictions and expectations

Unnecessary or inappropriate restrictions may be imposed by concerned parents, suggested by peers, or initiated by the patient.

- ▶ *Restrictions*—Limitation of certain leisure pursuits is often appropriate, but undue restrictions may have major psychosocial consequences. A sensible balance must be achieved with the teenager's agreement. In general, restrictions should be kept to a minimum.
- ▶ *Expectations*—Parents may have lowered expectations and aspirations for a child or teenager with a chronic disorder; this can become a damaging self fulfilling prophecy, leading to poor self esteem and performance.

Peer pressure

The teenage years bring an intense peer pressure not to be different. For the teenager with epilepsy, this has important implications:

- ▶ lack of openness and unwillingness to discuss problems and fears
- ▶ poor compliance with medication since the tablets are a reminder to the teenager that they are different
- ▶ alcohol and recreational drugs may be experimented with under peer pressure, with consequences for epilepsy control.

Lifestyle

Epilepsy has lifestyle implications of particular concern to the teenager. Seizures and their treatment can adversely impact on educational and employment success, driving, sport and leisure activities, use of alcohol, contraception, pregnancy, and parenthood. Safety advice about cooking, swimming, bathing, open fires, sports, and childcare is essential. However, peer pressure to participate and to appear normal often lead to failure to comply with this advice.

The general principles upon which lifestyle advice is based are:

- ▶ acknowledgment that living with epilepsy is about living with certain risks
- ▶ informing patients of the risks presented by epilepsy while encouraging teenagers to make their own decisions about risk avoidance
- ▶ acceptance of the legal ban on driving with its consequences for certain career choices.

Driving

Young people with epilepsy must be informed of the driving laws well before the age at which they may drive. Adolescents may hold misplaced beliefs about the legal driving restrictions or not understand the impact of their epilepsy on their legal ability to drive.

Employment

Career ambitions must take account of regulations limiting entry into certain jobs—for example, the armed services. In practice, the main barriers to obtaining work arise from misinformation from family, friends, and medical personnel, and, more importantly, from employer discrimination against persons with epilepsy. Where particular difficulties arise, referral of teenagers for career counselling at an early stage is ideal.

"It's remarkable, Mr. Fisher. You have the clothes of a man half your age!"

Reproduced with permission from *The New Yorker Book of Doctor Cartoons*, 1988. Aurum Press Ltd.

Figure 1 The principles of epilepsy care in teenagers are applicable also to other age groups.

Sport

For most teenagers with epilepsy, full and normal participation in sport should be encouraged. The risk of having a seizure (which may be high) must be distinguished from the risk of injury (which may be low). Certain sports carry particular risks from seizures—for example, swimming, cycling, riding—and the level of supervision required should reflect the frequency and severity of seizures. In sports involving additional risk to others, participation would be unwise without special arrangements—for example, scuba diving, rock climbing.

Alcohol and illicit drugs

These are used or experimented with by the majority of teenagers. Indeed, consumption of some alcohol may be important for social bonding in teenagers. Although alcohol is itself anti-epileptic, it may impair seizure control because of interactions with hepatic metabolised medication, sleep disruption, forgetting of medication, and the risk of withdrawal seizures. A sensible compromise is to advise patients with epilepsy to keep their alcohol consumption to no more than 4 units in a 24 hour period. The risks of recreational drugs to epilepsy are not known, although there are strong pointers that amphetamines and cocaine are epileptogenic.

Photosensitivity

Photosensitive patients (on EEG) should be advised caution at discos and use of computer screens, especially if not taking an AED. It is worth discussing this risk even with those who are not photosensitive, since many people with epilepsy erroneously believe they should avoid these situations.

Sleep pattern

Missing sleep may provoke seizures in idiopathic generalised epilepsy, the most common epilepsy in teenagers.

Young women with epilepsy

Special issues for young women are contraception, pregnancy planning, teratogenicity, and motherhood.⁶ Although teenagers and their parents are often surprised to hear such issues raised at this stage, it is actually an ideal time to optimise treatment, with future pregnancy in mind, before the responsibilities of driving, work, and family exert pressure to leave medication unchanged. A fact sheet for patients giving useful information on this is available from *Epilepsy Action*.⁷

- ▶ **Hormonal contraception**—Certain contraceptives are rendered less effective by medication that induces hepatic glucuronide conjugating enzymes. Thus, women taking carbamazepine, phenytoin, topiramate or oxcarbazepine, who wish to take an oral contraceptive, would require a preparation containing at least 50 µg ethinylloestradiol, and even then should consider using additional barrier contraception. Levonorgestrel implants are occasionally ineffective in women receiving these AEDs, and medroxyprogesterone injections must be given every 10 rather than 12 weeks. Although lamotrigine does not promote hormonal contraceptive failure, lamotrigine concentrations may be lowered by the oral contraceptive, allowing possible breakthrough seizures or toxic effects on contraception withdrawal.⁸
- ▶ **Pregnancy planning**—It is essential that every young woman is aware of the potential harm from medication to her unborn baby. This is especially important in adolescence where at least 30% of pregnancies are unplanned. Ideally, the need for AED should be reconsidered before any planned pregnancy, and the regimen optimised. Folate 5 mg daily may in theory help to prevent neural tube defects associated with valproate treatment, and is recommended for all women taking AEDs who are not taking definite contraceptive measures. Nausea and vomiting in early pregnancy may upset medication blood concentrations. Many women unilaterally stop their prescribed medication before or during pregnancy, without necessarily informing their physician. The indications for assessing blood AED concentrations during pregnancy remain a matter of debate.
- ▶ **Teratogenicity**—All AEDs are potentially teratogenic. Prospective observational data from the UK Epilepsy and Pregnancy Register show an alarming trend towards valproate being more associated with major congenital malformations than either carbamazepine or lamotrigine.⁹ There are also suggestions of increased neurodevelopmental delay among children exposed in utero to valproate.¹⁰ Consequently, sodium valproate should no longer be regarded as a first choice AED in young women. A case could be made to target those on valproate for review, aiming to shape informed decisions about their long term treatment.
- ▶ **Parenthood**—Loss of sleep and increased stress in the postnatal period may increase the seizure tendency, presenting additional risk to mother and baby. Sensible precautions for mothers with frequent seizures include feeding and changing the baby on the floor surrounded by cushions, using a baby bath on the floor with only very shallow water, and carrying the baby strapped into a carrycot, especially on stairs.

Emphasising independence

Independence should be encouraged in the majority of teenagers with epilepsy. However, the frequent association of learning disability may mean that true independence can be difficult or impossible to achieve. A useful marker of independence is the taking of responsibility for administering medications. The teenager with disability is more likely to be treated by family and elders as a child than as an adult. Such

attitudes strongly influence the response of the teenager to their condition.

Parents play a major role in helping the teenager to cope, and almost invariably accompany them to initial consultations. While wishing to encourage teenager independence, the continuing involvement of parents must be respected. Indeed, parental involvement may well be what teenagers have chosen.

Teenage boys and girls appear to cope with chronic disability such as epilepsy in different ways.¹¹ Boys are more likely than girls to try to hide their disabilities, or to appear as being in control and unaffected by illness, particularly when in public. Girls more readily admit to a chronic disability, independently manage their disease, and feel that parents, teachers, and health professionals expect this of them. Despite this, girls are more likely to be secretly non-compliant in their treatment regimens and to feel guilty because of this.

FOLLOW UP

Patients with either active epilepsy or significant medication side effects need regular review, and continued efforts to alleviate the symptoms. However, regular review of teenagers whose epilepsy is stable on medication can also be justified. Rapid growth and changes in lifestyle and responsibilities during the teenage years mean the disorder and its treatment require regular re-evaluation in any new context which arises.

CONCLUSION

The challenge of adolescence is compounded by disabilities such as epilepsy. The problems faced by teenagers with epilepsy are not unique to them but apply to patients with epilepsy of all ages (fig 1). However, epilepsy in teenagers can harm personal and social development, and directly impact

upon everyday lifestyle issues such as education, employment, driving ability, alcohol use, contraception, and pregnancy. The physician must encourage an adult approach to teenage epilepsy management, encouraging responsibility and partnership in the management of their condition, and thereby provide the best opportunity for them to cope with their disability and to optimise treatment.

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