Some Aspects of Epilepsy and Convulsions in Childhood

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SUMMARY

The diagnostic and therapeutic problems of epilepsy and convulsions in childhood are reviewed. Their non-specific aetiology is stressed, particularly with neonatal convulsions and infantile spasms. The role of febrile convulsions in causing later temporal lobe epilepsy and gross neurological deficit, underlines the importance of preventing and terminating these common attacks.

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In this review of some of the problems of epilepsy and convulsions in children, it seems wise to start by drawing a distinction between the child with epilepsy, defined as a tendency to recurrent attacks, on the one hand, and the child who, in certain situations, and only in such situations, has one or more seizures. Fits occurring with transient biochemical disorders in the neonate or in older children with fever, or from photic stimulation from television, are examples of the latter. The ease with which convulsions may be provoked in the young individual is well known, and Lennox's paraphrase of Orwell 'all men are epileptic, but some are more epileptic than others', expresses this truth, and the fact that, but for certain inbuilt safeguards and inhibitory mechanisms, seizures would be much commoner than they are. The tendency to recurrent attacks, which constitutes epilepsy, is believed to affect 6 - 8% of school-going children in the UK, and the figures are probably not much lower, and in some cases higher, in other countries.

NEONATAL CONVULSIONS

In the neonatal period, seizures occur in 3 main situations, viz. in relation to perinatal problems such as prolonged and difficult labour with neonatal asphyxia and injury to, or haemorrhage into, the brain, or kernicterus; as a result of infections, including meningitis, but also with infection outside the nervous system; and in association with certain transient biochemical disorders. These include hypoglycaemia, hypocalcaemia, hypomagnesaemia and the rare condition of pyridoxine dependency, in which there is a genetically determined excessive requirement for this vitamin and in which fits may occur in the neonatal period (and even *in utero*) or later. The condition is easily treated by parenteral pyridoxine, which should be given as a therapeutic test in neonatal convulsions. It is sometimes not realised how high a dose may be needed to control the seizures, and inadequate doses may be given. In my experience as much as 1 g/day may be needed.

INFANTILE SPASMS

Infantile spasms, known also by various synonyms, are a most interesting form of seizure, having their peak onset at the age of 5 or 6 months, and rarely under 3 months or over 2 years of age. The subject has been well reviewed by Jeavons and Bower.[1] The first and classical description of the attacks was written in 1841 by West,[2] a practitioner in Kent, in a letter to the *Lancet* describing the case of his own son. The later 19th century literature provides occasional references to the condition, and the term 'epileptic apprentices' was used to refer to the tendency of patients to develop later major convulsions. It was only in 1952 that Gibbs and Gibbs[3] described the syndrome in which the characteristic seizures, for which salaam attacks is a very descriptive synonym, are associated in most cases with mental retardation and a typical electro-encephalographic pattern called hypsarrhythmia. Although easily recognised when seen, the attacks may be confused with attacks of colic or attempts to sit up.

About half the cases in most series are idiopathic or cryptogenic, but this proportion is decreasing as investigation allows recognition of underlying diseases. Of the symptomatic cases, birth problems and intra-uterine infections (toxoplasmosis, cytomegalovirus, syphilis, etc.) account for the majority, and these cases often show other evidence of cerebral dysfunction owing to these insults in the form of mental retardation, microcephaly or evolving cerebral palsy preceding the onset of the spasms. Inborn metabolic errors, such as phenylketonuria, account for a small proportion and, rarely, more obviously progressive degenerative neurometabolic diseases. The association between the onset of infantile spasms and immunisation procedures, particularly the triple diphtheria-tetanus-pertussis vaccination, is still not fully understood, but in certain cases the temporal relationship between the two events is so striking that it is difficult not to suspect a causal relation.

An important aetiological group is provided by tuberous sclerosis, recognisable in early life, before the appearance of the classical adenoma sebaceum, by detection of pale, non-pigmented naevi, often leaf-shaped, which are most

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easily recognised in contrast to normally pigmented surrounding skin in summer-time. Skin lesions should be looked for in siblings and parents in view of the dominant mode of inheritance of the disease, which has a very wide clinical spectrum so that skin stigmata may be the only features in relatives. Intracranial calcification should be looked for by skull X-ray examinations, but may not appear until some years have passed.

The treatment of infantile spasms, whatever their aetiology, is notoriously difficult. Conventional anticonvulsants are normally ineffective, although some good results have been reported with diazepam and nitrazepam. The best results have usually been obtained with adrenocorticotrophic hormones or adrenocortical steroids. Adrenocorticotrophic hormone is best used in large doses, such as 20 units twice daily, and may need to be continued for a length of time. Its use is often followed by a marked reduction or abolition of the spasms, and often also by improvement in the EEG. Unfortunately, although the child's development may show a temporary improvement, the ultimate outlook for intellect is very bad, and most series agree in reporting that 90% of patients are intellectually subnormal.

FEBRILE CONVULSIONS

Febrile convulsions have been known for many years. In certain families some children, when aged between 1 and 6 years, show a tendency for fits to occur with common febrile illnesses owing to infection outside the nervous system, and there may be a history of a similar tendency in a parent during childhood. This tendency ceases at the age of about 6 years in most of these cases but in a small proportion, perhaps 5%, fits may continue to occur in later life in the absence of fever.

Two important features of febrile convulsions are that they are common and are often prolonged, lasting half an hour or more, thus probably constituting the commonest cause of status epilepticus in childhood. For this reason alone they should be treated with respect. Many paediatricians are familiar with the situation in which a child, who has had one or more fits with fever without sequelae, develops a similar prolonged fit, and never recovers, either dying in status or, more often, being left grossly handicapped with dementia and spasticity. Although encephalitis is often queried in such cases, the evidence for it, in my view, is scanty, and it seems much more likely that it is the prolonged seizure itself, with associated hypoxia, hypoglycaemia and electrolyte disturbances, which is responsible for the neurological picture. This view is supported by neuropathological evidence in some cases, and this aspect has been well reviewed by Ounsted *et al.*[4] in their retrospective clinical study of the aetiology of temporal lobe epilepsy.

In one-third of the hundred patients investigated a past history of fits with fever was the only aetiological factor which could be implicated. Further evidence for the role of febrile seizures in the aetiology of temporal lobe epilepsy comes from studies of temporal lobes removed in the surgical treatment of this condition. Falconer,[5] in a recent series, found that the commonest pathological diagnosis in the temporal lobes removed was mesial temporal sclerosis, and that these patients have an early onset of epilepsy, and a high incidence both of positive family history of epilepsy and of prolonged febrile convulsions in earlier life. The importance of febrile convulsions has been stressed from another angle by Taylor[6] who derived evidence from a series of 158 cases of temporal lobe epilepsy with age of onset under 10 years for differential rates of cerebral maturation between the sexes and between hemispheres.

The practical importance of these studies is to show the possibility of preventing epilepsy and other handicaps by the urgent control of status epilepticus with fever and the prevention, where possible, of febrile seizures. Status is always an emergency. The first-aid treatment by the family doctor is often by means of intramuscular paraldehyde. Diazepam intravenously has proved effective but may not produce a lasting effect, so that continuous infusion may be necessary, and care is needed to avoid respiratory depression or cardiac arrest, especially when barbiturates have previously been used. The importance of maintaining a clear airway and oxygenation, reduction of hyperpyrexia, and correction of electrolyte imbalance and hypoglycaemia cannot be overstressed. The problem of prevention of febrile convulsions, and when to give anticonvulsants and whether continuously or intermittently, is a difficult one, and the decision must be an individual one.

TEMPORAL LOBE EPILEPSY

Temporal lobe epilepsy is commoner in childhood than is usually realised, and is often mistaken, when the attacks are mild, for petit mal absences. A careful history and encouraging the parents to demonstrate the attacks, amplified, when co-operation allows, by provoking an attack by asking the child to overbreathe, will usually resolve the doubt. Automatic movements, bizarre behaviour, lip smacking, chewing and swallowing movements, and autonomic features such as flushing, pallor, tachycardia and borborygmi may all occur in children, as in adults with temporal lobe epilepsy, and should be inquired about. The complex hallucinations and distortions of perception experienced by adults may also occur in children, but the younger patients are usually unable to describe them. Sometimes an older child will be unwilling to admit his symptoms, since he is afraid that they indicate madness. Discussion and reassurance are essential, and the patient's need to discuss his symptoms and what they mean to him must never be forgotten.

PETIT MAL ABSENCES, MYOCLONIC AND AKINETIC ATTACKS

Although the so-called petit mal triad is still spoken of, it seems preferable to distinguish between true petit mal absences with the associated 3 c/s spike and wave EEG pattern, on the one hand, and myoclonic and akinetic attacks on the other. They differ not only clinically but in their EEG correlates and drug responsiveness, the

former normally responding well to the succinimides, so that a therapeutic trial of ethosuximide may be diagnostic. When these 3 forms of 'minor' seizure occur infrequently, they are of nuisance value only, but if prolonged and continuous, they may be grossly disabling. Such is the case in a few children with intractable epilepsy who show a fluctuating condition of frequent myoclonic jerking and akinetic attacks which has been called minor epileptic status,[7] and found to respond poorly to conventional anticonvulsants and better to adrenocorticotrophic hormone or steroids, which may need to be continued for long periods.

INVESTIGATION

The investigation of epilepsy in childhood cannot be considered in any detail in this article. It is influenced, however, by certain principles and differences which separate it from epilepsy of later onset. Thus, childhood epilepsy is much more likely to be due to perinatal problems and much less likely to be due to cerebral tumour, than in adults. Biochemical disorders and particularly inborn errors of metabolism must be considered, especially when retardation is associated. Skull X-ray films often give useful information, with calcification suggesting intra-uterine infection by toxoplasma or cytomegalovirus, tuberous sclerosis or the Sturge-Weber syndrome. The EEG, although often helpful, is seldom diagnostic, and should be considered only as part of a complex of clinical and investigational data. Special neuroradiological studies of epilepsy in childhood, although commonly undertaken, are seldom helpful, being much more likely to demonstrate 'cerebral atrophy' (which is non-specific and of limited prognostic significance) than the cerebral tumour which the clinician is often afraid of overlooking.

MANAGEMENT

The management of epilepsy in childhood (apart from the important question of prevention) can be considered under 2 headings: drug treatment and general management. Anticonvulsant drugs are generally well tolerated by children, provided they are used in the correct dose, based on the patient's mass or surface area. This is especially important with phenytoin, which is frequently used in excessive doses, thus causing severe ataxia. The correct dose is about 5 mg/kg body mass/day, and it should not exceed 7 mg/kg. Hirsuties and gum hypertrophy are common side-effects, but good dental hygiene helps to limit the latter, and cosmetic gum surgery can give excellent results, so that the drug need not be discontinued. Phenobarbitone, although often effective, is notorious in childhood for aggravating or provoking behaviour disorder and hyperkinesis, and may render the child unmanageable. Primidone, in a dose of 20 - 25 mg/kg/day, is often well tolerated if the dose is increased very gradually. Diazepam, nitrazepam and many other drugs are also available, which may prove helpful especially when used in combination with one of the other anticonvulsants. The regimen of treatment should be as simple as possible and combinations of many drugs should be avoided. Combined tablets have little place in childhood epilepsy, preventing the adjustment of the dose which is needed with increasing age and other circumstances. Timing of dosage is important and most children need 2 or 3 doses per day, but rigid dosing by the clock is unnecessary. Nocturnal or early-morning attacks may often be prevented by a large dose on retiring for the night. Certain interactions between drugs must be borne in mind, as well as effects on folic acid and calcium metabolism reported in recent years. Reynolds[8] has produced a useful review of iatrogenic disorders in epilepsy. The tendency, noted in folate-deficient adults, for folic-acid supplements to provoke status epilepticus has not generally been found in children.

The duration of treatment cannot be decided in advance, and the decision when to discontinue anticonvulsants is a difficult one. A period of 2 years without fits is often arbitrarily taken as required before stopping drugs. The possibility of fits recurring cannot be excluded and parents and children should be told so that they may not be too greatly upset by a recurrence. It often happens that fits recur when the patient has outgrown the paediatric age group and comes under the care of a physician. For this reason the recurrence rate in epileptic patients whose treatment is discontinued in childhood may not be known to those responsible for the decision to stop treatment. This subject is well reviewed in a recent article by Holowach *et al.*[9] who followed up 148 children who had been seizure-free on anticonvulsants for 4 years, and found that an early age of onset of epilepsy and prompt seizure control were the factors associated with the lowest recurrence rate. The EEG was of limited value in predicting recurrence.

The general management of the child with epilepsy includes concern for all the social, educational and other problems which play a role. It is very much more than the manipulation of drugs, and involves not merely a child, but his whole family and social environment. As with all chronic handicaps, the aim must be to allow as full and normal a life as possible and to promote the fullest development of the child's potential. Restrictions of activities and unhealthy attitudes of parents, teachers and, be it said, some doctors, may be far more crippling than the condition itself, and the word epilepsy is for many people a source of fear and anxiety totally disproportionate to the real problems. It is in the education of all towards healthier and more enlightened attitudes that some of the most important work of SANEL and associated organisations consists.

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