Epilepsies of Childhood

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Editor's Foreword

'Age influences disease' and a chronological approach has clarified our understanding of many childhood disturbances. In the epilepsies it is surely essential, for maturation (that goes with age) provides the critical variable in all seizure disorders, as Ounsted pointed out. Why do fits occur rarely in the premature infant, most commonly in the few years following later infancy, and then decline in frequency in the years afterwards ? We have learned a great deal from many disciplines about how the contributory causes vary with the patient's age. No less important, we are learning how and why the clinical presentations take the different forms we may see in the panorama of growth. Based on the newer understanding, therapy – both preventive and remedial – can be applied more effectively to the fits themselves and to the patient who suffers them.

This book is the more logical and persuasive because of Dr O'Donohoe's chronological presentation, and he makes it glow with his rich and extensive clinical experience. At the same time experimental and research contributions, culled from many sources, are woven into the texture of his argument and inform the intensely practical advice he offers about both diagnosis and treatment.

Dr O'Donohoe has the gift of conveying the sense of complex scientific concepts in lucid words that clinicians will appreciate. With his delightfully personal writing, what could have been a dull collection of dry facts, and details of drugs, comes to life. The result is a book combining down-to-earth experience and perspective with literary charm. When clinicians refer to it, both for up-to-date ideas and for details of treatment, many will surely go on reading for the sheer enjoyment which he shares with them.

John Apley

Preface

Convulsive disorders have their highest incidence in childhood and seizures are among the most frequent symptoms presenting to the paediatrician and the paediatric neurologist. They cause parents great anxiety and doctors are often perplexed and confused by the problems they create. Public attitudes about epilepsy are still, unfortunately, rooted in prejudice and superstition.

There have been remarkable advances in the understanding and treatment of epilepsy in recent times, although the communication of new knowledge of the subject has been made difficult by an abundance of classifications and by misleading and outdated terminology. However, the *International Classification of the Epilepsies,* first introduced in 1969, is being accepted gradually and is providing an international language of epilepsy for the first time.

I have attempted in this book to describe the epilepsies of childhood in a more or less chronological sequence, while using the terminology of the *International Classification* and emphasizing the important influences of age, growth and developmental status on the type of epilepsy which develops.

Although the book is written primarily for general paediatricians at all stages of their careers, I hope that others, including specialist and community paediatricians, specialists in mental handicap, child psychiatrists and family doctors may find it useful and interesting.

My own interest in epilepsy derives from the fortunate experience of working with the late Dr Paul Sandifer at the Hospital for Sick Children, Great Ormond Street, in the late 1950s. I owe a special debt to the late Dr Ronald MacKeith, one of the greatest international paediatricians of his generation, who ensured that I joined the main-

stream of British and European paediatric neurology. Dr Christopher Ounsted and Dr David Taylor of the Park Hospital for Children, Oxford, have influenced my thinking about childhood epilepsy profoundly and their work is referred to frequently in this book.

I am greatly indebted to my excellent EEG technician, Miss Geraldine Monaghan, who has taught me much about the EEG and epilepsy, and to Miss Catherine Condron who typed my manuscript. The Research Centre at Our Lady's Hospital for Sick Children, Dublin, helped with a generous grant.

Above all, I must thank Dr John Apley for suggesting the book and for his constant advice and encouragement during the writing of it.

Niall V. O'Donohoe

Contents

CHAPTER 1

Epilepsy: History and Statistics

Epilepsy is not easily defined. The words 'epilepsy' and 'epileptic' are of Greek origin and have the same root as the verb meaning 'to seize' or 'to attack'. The word epilepsy means simply 'to be seized' in a passive sense. The idea of a disease seizing a man goes back to the old magic concept that all diseases were 'attacks' or seizures by gods or demons. Since epilepsy was the demoniac disease par excellence, the term gradually acquired a more particular meaning and came to signify an epileptic seizure. Epilepsy acquired its name because it attacked or seized both the senses and the mind. This concept of epilepsy is of ancient origin and was already in use, as were the terms 'epilepsy' and 'epileptic', in Hippocratic times (Temkin, 1971).

However, although 'epilepsy' and 'epileptic' were used in the earliest times as designations of both epileptic attacks and epileptic persons, the term epilepsy did not mean the underlying disease which the physicians preferred to refer to as 'the sacred disease' (morbus divinus). This name arose from the belief, common from antiquity to the relatively recent past, that diseases were phenomena more or less dependent on the supernatural and were considered to be a divine retribution for wickedness or a consequence of possession by spirits. Epilepsy, more than any other condition, was susceptible to explanation in these terms. Temkin (1971) has written: 'In the struggle between the magic and the scientific conception, the latter has gradually emerged victorious in the western world. But the fight has been long and eventful, and in it epilepsy held one of the key positions.'

The battle was first joined around 400 BC when Hippocrates attacked the supernatural explanation of epilepsy in the book on 'The Sacred Disease' (Chadwick and Mann, 1950). The alleged divine character, the

author or authors argued, was merely a shelter for ignorance and fraudulent malpractice. Epilepsy, it was claimed, was not more divine than any other disease, but, like all diseases, it was hereditary with its cause residing in the brain – a brain overflowing with a superfluity of phelgm, one of the four humours. Epilepsy, therefore, should be treated not by magic but by diet and drugs. Unfortunately the mythology surrounding epilepsy persisted, including a supposed link between epilepsy and lunacy and the idea that epilepsy could be caused by the influence of the cycle of the moon. The various ideas connecting epilepsy, madness, possession and similar states with the moon and other stars spawned a rich astrological literature through the centuries and even up to present day.

Having the sacred disease did not do the unfortunate sufferer much good. The epileptic was regarded as unclean; whoever touched him might become prey to the demon. It was thought that spitting would keep the demon away and throw back contagion, and thus one could escape infection. The magic concept, according to which epilepsy was a contagious disease, was one of the factors that made the epileptic's life a misery and gave him a social stigma. It was a disgraceful disease and the unfortunate person who felt an attack coming on rushed home or to a deserted place where he covered his head. To the ancients the epileptic was an object of horror and disgust, and throughout almost all of history those afflicted have been viewed with anxiety and fear. No other illness has set individuals apart so far, so often and so long, and these attitudes persist to the present day despite the quite frequent association of greatness and genius with the condition (Socrates, Julius Caesar, Napoleon, Dostoevski, Dante and Handel, to name a few).

From the time of hippocrates physicians wrote about 'convulsions', particularly with reference to children, without clearly defining the meaning of the term and its relationship to epilepsy. There was an awareness in medical writing about children that convulsions in early childhood might have a different prognostic significance from epileptic seizures in older persons, but for a long time terminology remained confused. Attitudes to children with epilepsy were unenlightened and often cruel until well into the nineteenth century, and they frequently suffered the general fate of epileptics in being confined with insane persons.

The enlightenment of the eighteenth century and advances in neurology in the nineteenth century led to improved knowledge and understanding of epilepsy and to the final abandonment of the idea of demoniacal possession as a cause. The National Hospital for the Paralysed and Epileptic in London was opened in 1860 and during the early years of that decade the work of the great John Hughlings Jackson began in that hospital. In 1873 this produced a broad definition of epilepsy which is still perfectly valid: 'Epilepsy is the name for occasional, sudden,

excessive, rapid and local discharges of grey matter.' According to his definition there was not just one form of the disease, but many epilepsies: one of these was a type of unilateral epilepsy with a characteristic march of clinical events which bears his name. Hughlings Jackson's former assistant W. R. Gowers published his important book *Epilepsy and Other Chronic Convulsive Diseases* in 1881, which incorporated many of Hughlings Jackson's ideas and attempted to differentiate epilepsy from conditions such as hysteria and migraine.

The epoch-making discovery of the human electroencephalogram (EEG) and the first publication of his observations on it by Hans Berger in 1929 (Gloor, 1974) provided the tool which facilitated the separation of epilepsy from other conditions and offered visual proof of Hughlings Jackson's theories. The subsequent explosion of knowledge about the EEG in epilepsy over the next two decades culminated in the publication of the classic *Atlas of Electroencephalography* by F. A. and E. L. Gibbs (1952). Other important milestones in the scientific understanding of epilepsy were Penfield and Jasper's (1954) *Epilepsy and the Functional Anatomy of the Human Brain* which dealt in detail with the neuroanatomy and neurosurgery of epilepsy, and William Lennox's great book *Epilepsy and Related Disorders*, which was published in 1960 at the end of his long life devoted to the study of epilepsy and the care of its victims.

Lennox (1960) wrote that epilepsy was a disturbance of the normal rhythms of the brain. 'The rhythm of the body when orderly spells health. Dysrhythmia is a disease. Of all the systems of the body, the central nervous system is most nearly cyclic in function, and recurrent irregularity of its rhythm most profoundly disturbs the functions of both body and mind.' Lennox regarded epilepsy as an anarchy of cell function just as cancer is an anarchy of cell growth.

The manifestations of epilepsy are legion and no single symptom is essential. Lennox (1960) quoted Boerhaave (1974) as follows: 'For there is no one gesture, inflexion, or posture of the body known, which it has not shewn at some time, and it emulates all the motions of running, walking, turning, bending forwards, lying down, standing upright, or keeping the body in a very stiff and almost insuperable action.' The manifestations of epilepsy may appear as disturbances of consciousness or be evinced by sensory, visceral or motor signs, or present as perversions of ideation, emotion or mood. Some patients may experience all of these symptoms, others only one or two, and the symptomatology and intensity of the disturbances may vary from time to time. They may also experience what Lennox called the horror of epilepsy, quoting Margiad Evans (1953) as follows: 'Ever since a second convulsion I have been incredulous of all things firm and material. The light has held patches of invisible darkness. Time has become as rotten as worm-eaten

wood, the earth under me is full of trap-doors and the sense of being, which is life and all that surrounds and creates it, a thing taken and given irresponsibly and without warning as children snatch at a toy.'

It is important to realize that all forms of epilepsy, as Hughlings Jackson wrote, arise from recurring excessive neuronal discharges occurring somewhere in the brain. Sometimes the site of origin can be easily identified while at other times no epileptic focus can be found. However, whatever their source the discharges frequently spread to other parts of the brain and even to the central nervous system as a whole. It must also be remembered that, as a rule, epilepsy is a chronic recurring disorder, and that many upsets in homeostasis originating outside the central nervous system may provoke disturbances of the brain culminating in epileptic phenomena identical with those caused by epilepsy itself. Anyone may have a fit given the right circumstances. These sporadic disturbances must be distinguished from epilepsy which is a recurring condition. Furthermore, the diagnosis must be made positively, based on careful history and clinical observations, and not just by exclusion. The label of epilepsy is, unfortunately, still too pejorative for mistakes to be made. In making a diagnosis, it is better to err on the side of 'not epilepsy' and subsequently to correct one's mistake than to apply a mistaken diagnosis of epilepsy which may be difficult to rectify later.

EPIDEMIOLOGY: PREVALENCE AND INCIDENCE

The study of the epidemiology of epilepsy is beset with problems since epilepsy is often a hidden disease. This makes the accumulation of accurate statistics on the prevalence and incidence of seizures difficult and unreliable. Prevalence refers to the ratio of those people affected by a particular disorder to those not affected at a given time. Incidence refers to the number of new cases of a disorder within a given population in a given time period and may also be expressed as a percentage.

The survey of Pond, Bidwell and Stein (1960) was comprehensive and gave an overall prevalence rate of 6.2/1000 population with an inception rate of new cases of 0.7/1000/year. These authors included every type of epileptic fit. Kurtzke *et al.* (1973) stated that the best evidence available indicated the prevalence of convulsive disorders as about 4–6/1000 population at the time of publishing their book. They further emphasized that all prevalence rates must be considered as minimal estimates. Taking a reasonable estimate of prevalence as 5/1000 would mean that there were about one million people with epilepsy in the USA. The Epilepsy Foundation of America and the National

Institute of Neurological Diseases and Stroke in the USA have produced higher estimates of up to four million people with some form of epilepsy living in the USA (*Basic Statistics on the Epilepsies*, 1975). Epileptic patients outnumber those with any other serious neurological disease, with the probable exception of cerebrovascular disease. Roger (1970) has written a comprehensive review of the literature on the epidemiology of epilepsy in Europe and North America.

Prevalence in childhood

Convulsions are the commonest problem encountered in paediatric neurology. It has been estimated that 90 per cent of those who develop symptoms of epilepsy do so before the age of 20 years. The inception rates of new cases per year are highest in the youngest age groups, they fall off during childhood, rise again in the age range 10–20 years, and then fall off rapidly among adults (*Epilepsy in Society*, 1971). According to Miller *et al.* (1960) in Newcastle upon Tyne, 7.2 per cent of the children in the 'Thousand Family Survey' had had one convulsion or more by the age of five years, and 20 per cent of these children had died. All but one of the deaths occurred under the age of one year and were ascribed mainly to birth trauma and infections. Millichap (1968) estimated that approximately 3 per cent of the population of the USA under five years of age were affected by febrile convulsions. Various studies have indicated that in Great Britain the prevalence of epilepsy in schoolchildren is in the order of 8/1000 (*People with Epilepsy*, 1969). This means that there must be almost 60 000 schoolchildren in that country who are subject to epilepsy, and the inception rate of new cases is between 2 and 4/1000/year. However, it is generally believed that only about half of the schoolchildren with epilepsy are known to the school health authorities in Great Britain.

Cooper (1965), in a survey of epilepsy seen in general practice, found that among eight-year-old children with epilepsy there was an excess in the group classified as average or below average in performance at school, and others have commented on reading retardation in children with epilepsy. Learning difficulties in children with epilepsy are probably due to several different causes including underlying brain damage, the effects of long-term medication and also to the fragmentation of attention by subclinical discharges. These various aspects of the problem are discussed in detail elsewhere.

It is clear, therefore, that epilepsy is common in childhood and that affected children are vulnerable medically and educationally, in addition to facing considerable social difficulties as a result of their seizures. Epilepsy in childhood, just as in adults, is a family problem which will

have an impact on and modify the lives of all the family members. Looked at from the medical standpoint, the complexity of the problems facing the child with epilepsy and his parents is such as to make it impossible for any one discipline to deal adequately with all of them, and a team approach is essential. The many facets of epilepsy make it difficult to define concisely. David Taylor (1969) has attempted it as follows: 'Epilepsy is a phenomenon, it scarcely warrants being called a symptom and it is not a disease.' However, we need not take too much time trying to define epilepsy, but rather try to understand it; to a considerable extent this is now possible.

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CHAPTER 2

Problems of Classification and Aetiology, Including Genetic Aspects

Classification in general is a method of conveying information. The important components of any classification are accurate nomenclature, arrangement and grouping. The terminology and classification of epilepsy have evolved over many years creating a profusion of interchangeable and confusing descriptive terms. Denis Williams (1976) said that there are as many classifications as there are needs to classify. Marsden (1976) defined the problem of classification as a need to evolve a single code to cover three basically incompatible systems of classification, namely, that of clinical signs and symptoms of the fit, that relating to the anatomical and electrophysiological evidence of the source of the fit and that defining the aetiology of the fit. Any classification employed must be useful and should reflect the needs of the user.

To the paediatrician and paediatric neurologist the chronological aspects of epilepsy through infancy, childhood and adolescence are particularly interesting, and a chronological approach will, to a large extent, be adopted in this book (see *Figures 3.1* and *4.1*). It is often difficult to convince doctors dealing exclusively or predominantly with adults that paediatrics is not just medicine (or neurology) in small people. The factors of age, growth and development exert their influences constantly and certainly not least in the problems of childhood epilepsy. Ounsted (1971) wrote that: 'for all seizure disorders, maturation is the critical variable', and Lennox (1960) said that: 'the type of epilepsy which occurs in a child represents a confluence of age, heredity, and structural brain abnormality'.

The simplest, time-honoured classification of epilepsy is to divide it into idiopathic and symptomatic epilepsy, and this is still in wide clinical use. Another popular and practical classification has been pithily stated by Williams (1968) in a discussion on the management of epilepsy. 'For our present purpose there are two kinds of epilepsy, general and focal. The general epilepsies fall into two groups, petit mal and grand mal. There are only two kinds of petit mal, absences and myoclonic fits without disorders of consciousness, and they both have as their electrical accompaniment a generalised three-a-second spike-wave discharge of characteristic kind. There is only one kind of grand mal, and it in effect constitutes a unit of disturbed consciousness and behaviour. All other forms of epilepsy are focal, whatever their behavioural or experimental accompaniments, and however elaborate or unitary may be their nature.'

An international committee was convened by the *International League Against Epilepsy* to consider the formulation of a comprehensive classification of epilepsy which would include the type of clinical seizure, the type of EEG seizure, the EEG interictal expression, the anatomical substrate and aetiology and the age of the patient. The product of this committee was published in 1969 and is now generally known as the *International Classification* (Gastaut, 1969, 1970). Epileptic seizures were classified as follows:

1. Partial seizures or those beginning locally. These fits were further subdivided into those with elementary symptomatology, generally without impairment of consciousness; those with complex symptomatology, generally with impairment of consciousness; and partial seizures becoming secondarily generalized.
2. Generalized seizures or those which were bilaterally symmetrical and without local onset. These included petit mal absences, bilateral massive myoclonus, infantile spasms, clonic and/or tonic seizures, tonic-clonic seizures (grand mal) and atonic and akinetic seizures.
3. Unilateral seizures.
4. Unclassifiable seizures.

Merlis (1970) modified the classification as follows:

1. Generalized epilepsies
   (*a*) Primary generalized epilepsies including petit mal and grand mal
   (*b*) Secondary generalized epilepsies
   (*c*) Undetermined generalized epilepsies
2. Partial (focal, local) epilepsies which include Jacksonian and temporal lobe seizures
3. Unclassifiable epilepsies.