

Commentary

Are We Failing Those With ‘The Falling Sickness’? Time to modernise the approach to epilepsy care.

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The epilepsies are a common, heterogeneous group of disorders linked by a tendency to recurrent seizures in affected individuals and which have no respect for age, gender, race or social status. A seizure is defined as an intermittent, stereotyped, disturbance of consciousness, behaviour, emotion, motor function, or sensation that on clinical grounds is believed to result from cortical neuronal discharge. Descriptions of seizures can be traced as far back as civilisation itself. In antiquity, seizures were felt to occur as a consequence of supernatural forces. Since the late twentieth century there has been an explosion in the understanding, investigation and management of seizure disorders. Perhaps as a cultural hangover from millennia of myths and the potentially dramatic clinical manifestations of seizures, ‘civilised’ society remains alarmingly adept at stigmatising the person with epilepsy.

THE FALLING SICKNESS AND SACRED DISEASE

Currently the earliest known detailed description of various seizure types are contained within the stone tablets of the Sakikku (meaning ‘All Diseases’), a Babylonian text compiled circa 1000 BC that is held in the British Museum, London. A Sumerian term referring to descriptions of seizures has been translated as ‘The Falling Sickness’. The Babylonians thought that each seizure type was thought to represent possession by a particular demon or departed spirit and hence treatment focussed on spiritually based methods.¹ Hippocrates (circa 400 BC) has been credited with authorship of a text ‘On the Sacred Disease’ which concludes that seizures are purely a physical ailment and a manifestation of an abnormality within the brain rather than some sacred, in other words supernatural, influence.² Despite his insightful conclusion this opposing theory failed to rival the notion of supernaturalism in the thinking of others who came after. Galen’s (129-200 AD) descriptions of epilepsy were more restrictive than much that preceded him but such was his influence on so many aspects of medicine his theories dominated well into the Renaissance. He concluded there were three forms of epilepsy “In all forms it is the brain which is diseased; either the sickness originates in the brain itself,... or it rises in sympathy into the brain from the cardiac orifice of the stomach... Seldom, however, it can have its origin in any part of the body... and then rises to the head in a way which the patient can feel...”³ Greek Galenists employed dietary manipulation, pharmacological and surgical intervention depending on the type of epilepsy present.

THE MIDDLE AGES

Raphael’s (1483-1520) painting the “Transfiguration of Christ”



The Transfiguration by Raphael (1516-1520). A boy with epilepsy is depicted in the foreground. Reproduced with permission of the Vatican Museums.

illustrates passages in the Gospels in which Jesus Christ casts out a devil from a boy with epilepsy. There are several similar references to epilepsy in the Bible, in large part forming the Christian Middle Age view of epilepsy as a demonic disease or result of witchcraft. Galen’s restrictive influence on what

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phenomenologically could be considered *epileptic* waned and other manifestations (partial and complex partial events) were recognised as being epileptic in origin. Unfortunately, better recognition of epilepsy did not necessarily translate into better understanding its true origins. Seizures are documented as one of many possible characteristics of witches in the classical text on witch-hunting, *Malleus Maleficarum*, something that led either directly or indirectly to the deaths of thousands of women, many of whom had epilepsy, during the approximately 200 year lifespan of the inquisition.

Toward the late nineteenth century the work of John Hughlings Jackson, a London neurologist, helped further the understanding of many aspects of neurology, not least epilepsy. His recognition that a seizure is merely a reflection of underlying neuronal dysfunction, rather than a stand alone entity in itself was a great leap forward. Jackson's ideas have been refined into what we accept as the principles of modern day epileptology. This enlightenment led Jackson and his contemporaries to found the Chalfont centre for epilepsy near London, an agricultural colony where those with epilepsy could live and work in a compassionate atmosphere free from the negative prevailing social attitudes. Today, the Chalfont Centre continues to look after residents with refractory epilepsy but it now also acts as a national referral centre for investigation and management allied to a world class research portfolio.

In recent years Government advisors have published key documents providing guidance on management of epilepsy (NICE, SIGN).^{4,5} The National Sentinel Clinical Audit of Epilepsy-related Death, published in 2002, reported on sudden unexpected death in epilepsy (SUDEP) which is thought to account for 500 deaths per annum in the UK.⁶ The factors leading to the excess of sudden death remain a matter of speculation but it is accepted that poor seizure control is implicated.

CLASSIFICATION AND MANAGEMENT

Although imperfect the International League Against Epilepsy (ILAE) classification system has been adopted worldwide and acts as a framework for the organisation and differentiation of seizure types and epilepsy syndromes.^{7,8} The terms 'grand mal' and 'petit mal' though part of the vernacular are quoted all too often in medical communication and have no place in the modern lexicon of epilepsy. Our patients deserve an accurate (as possible) diagnosis which in turn helps direct appropriate management and point to prognosis. Pharmacological therapy that may greatly benefit one epilepsy syndrome may lead to a dramatic worsening of another. Furthermore, mindful that 'not all that shakes is epilepsy' it is imperative that patients and their doctors have ready access to expert services to ensure individuals are not inappropriately labelled as having epilepsy. The last decade has seen the proliferation of specialised epilepsy clinics throughout the UK. Such clinics have been demonstrated, through providing ready access to epilepsy specialists, specialist epilepsy nurses and others working within a multidisciplinary framework, to improve epilepsy diagnosis and management.⁹

INVESTIGATION

Modern neuroimaging modalities with their impressive spatial resolution have been integral in improving our ability

to identify the aetiology of localization-related epilepsies in particular. We can now easily visualise hippocampal sclerosis in some patients with medically refractory seizures, offering a significant (carefully selected) proportion of them a cure with epilepsy surgery, and even in some cases anti-epileptic drug withdrawal. The rapid expansion in knowledge of disorders of cortical development, the second most frequent cause of intractable focal epilepsy, has also only been possible through advances in neuroimaging. The combination of structural and functional imaging (fMRI, PETCT) further promises additional insights into ictal and interictal cerebral functioning.

In contrast to all forms of imaging interictal electroencephalography (EEG) is a well established technology and remains a useful adjunct in the investigation of epilepsies. However, requests along the lines of "EEG please, rule out epilepsy" both overvalues EEG and misrepresents its place in clinical practice. Combining continuous long term EEG with real time video capture has become a vital resource for the investigation of episodic attacks, when the diagnosis is not clear, for the classification of seizures and as part of a pre-surgical evaluation. Continuous EEG monitoring for the management of status epilepticus in an ICU setting can also be useful.

The last fifteen years has seen a proliferation in the number of anti-epileptic drugs available. Some confer definite advantages over the older, established therapies but are more costly. To date little in the way of head to head comparison between these newer agents and the established therapies has been published. The NHS R&D health technology appraisal programme has reviewed the available evidence on the longer term clinical outcomes and cost-effectiveness of standard and new antiepileptic drugs.⁴ Choice of therapy should be based on a number of variables including gender, age, seizure types or syndromes, co-existent medications and patient choice. Monotherapy should be the aim where possible with carefully designed schedules for medication changeover periods.

GENDER ISSUES

The last decade has seen a focus on gender issues in epilepsy and local neurologists in tandem with allied specialists have been contributing to the evidence based management of epilepsy during pregnancy, gaining international recognition for the ongoing work of the UK Epilepsy and Pregnancy Register¹⁰. We now have for the first time accurate data on the teratogenic risk of the most commonly used drugs. As a result of our greater understanding of the complexity of managing women with epilepsy during pregnancy joint specialist epilepsy / obstetric clinics are being set up, with one recently having been established at the Royal Group of Hospitals in Belfast.

FUTURE GOALS

Given an incidence of approximately 50 cases per 100,000 persons per annum and a prevalence of 5-10 cases per 1000 for epilepsy, it can be (conservatively) estimated that for Northern Ireland the number of consultants required to service epilepsy lies in the range 4 to 13. ILAE guidelines contend that all patients with epilepsy should be managed by someone who has an interest in epilepsy. In contrast, at present, there

is a stark mismatch between that which is required and that which is available in Northern Ireland.

Traditionally general physicians have provided a great amount of the care of patients with epilepsy and other neurological conditions, and not only in the acute situation. However, since the introduction of Calman type training, very few trainees in general medicine have been able to work in neurology units, above the senior house officer level. This might be viewed as somewhat of an organisational faux pas given that one in six of all acute medical admissions is for a neurological problem with one in three of these being seizure related.¹¹ Given this shift in the organisation of post-graduate medical training it is therefore essential that neurologists become more involved in the management of all patients with neurological symptoms and conditions.

In order to translate advances in epileptology into improved standards of care locally a broad ranging package of measures including improved education and capital investment is required. The current positioning and time given to the teaching of neuroscience at the local medical school does not reflect the prominence of neurological disorders in practice. Numbers experiencing postgraduate training in neurology must undergo dramatic expansion as should the number of career neurologists. We must improve links to primary care, accessibility to specialist opinion and investigations. We need only look sideways at the achievements of our diabetology colleagues to realise that such change is not simply a pipedream but truly achievable given adequate planning, resources and political will. Not to modernise is to fail our patients.

Conflict of interest: The authors have received funding grants from the pharmaceutical industry for an antiepileptic drug population register.

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Assistant editor Rex Wilson retires

After twenty years as Assistant Editor, Rex Wilson has corrected his last paper for the Ulster Medical Journal. He first published a paper in the journal in 1946 when he was a 'penicillin officer' at the Royal Victoria Hospital, and went on to have a career in General Practice. We wish him many more years of 'retirement' following his recent 90th birthday. Rex has an excellent grasp of grammar and rarely missed a spelling or syntax error. His skills will be missed in this era of writers more used to text messaging. He was recently presented with a book 'Meetings with Remarkable Trees' (by Thomas Pakenham) as an appropriate reminder of all the papers that he has handled, and a certificate from the Ulster Medical Society and the journal, by the current editor Prof. Patrick Morrison and former Editor Prof. David Hadden.

Patrick Morrison

