

Epilepsy

# Idiopathic generalised epilepsies: not only for the paediatrician

J W Sander

Adult onset IGE is more common than generally realised

The commonest epileptic syndrome is the so-called idiopathic generalised epilepsy (IGE), which accounts for at least a third of all cases of epilepsy in the community.<sup>1</sup> This proportion is even higher in the paediatric age range. The syndrome is characterised by the presence of generalised tonic clonic, myoclonic, and typical absence seizures on their own or in different combinations. The onset is usually before the age of 16. IGE has a typical electroencephalographic (EEG) pattern with paroxysms of generalised spike and wave and polyspike discharges, which is the hallmark of the syndrome. Age of onset and main seizure type are used to classify IGE further into four main subsyndromes: IGE with tonic-clonic seizures only, childhood absence epilepsy, juvenile absence epilepsy, and juvenile myoclonic epilepsy. As the majority of patients have the onset of IGE in childhood or adolescence, the international classification currently in use only recognises subsyndromes with onset in childhood. Although it is well known in epilepsy specialist centres that IGE may present in adult life, this is often not well recognised by neurologists who do not have a specialist interest in epilepsy. Therefore,

the paper by Marini *et al* (this issue, page 192–196)<sup>2</sup> is of interest as they describe 34 consecutive patients with IGE beginning after the second decade of life, including several people over the age of 50 presenting with “de novo” IGE. The authors quite rightly suggest that adult onset IGE is more common than generally realised and this is an important takeaway message. The second message is the importance of a directed diagnostic EEG strategy in all patients presenting with a first unprovoked epileptic seizure independent of age. This goes against the suggestion that after the age of 25 years, EEG is less important as an investigative tool in epilepsy.<sup>3</sup> The authors’ practice is to record an early postictal EEG, followed by a sleep deprivation EEG in negative cases and they have previously shown that this leads to a higher diagnostic yield in first seizure cases in all ages.<sup>4</sup> A second spin off seems to be a relatively frequent identification of adult-onset IGE that might otherwise have been regarded as being partial epilepsy of uncertain origin. The paper also highlights the growing importance of genetics in epilepsy, particularly in IGE.<sup>5</sup> Early onset IGE has a genetic aetiology with complex inheritance. Family and twin

studies suggest a common genetic origin for all subtypes of IGE and this seems also to apply to the late onset form. Currently, intensive work is ongoing to clarify fully the genetic blueprints of IGE, and the Melbourne group, which authored this article, are major players in this area. For the general neurologist the recognition of adult-onset IGE has implications for accurate diagnosis and treatment. Adults with a family history of IGE and in whom other causes have been ruled out are prime candidates for adult-onset IGE. The diagnosis of adult-onset IGE can avoid unnecessary investigations, direct appropriate treatment, and allow an optimistic prognosis, and this should not be forgotten.

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Dementia

# Dipoles and dementia

C M Epstein

Can the link between MEG dipoles and cortical activity lead to increased applications for the unique capabilities of MEG?

For almost two decades magnetoencephalography (MEG) has been the stepchild of functional neuroimaging. Despite its evolution from single sensors to arrays of more than 100, with unmatched temporal resolution, the legitimacy of MEG as a clinical and

research tool has been repeatedly challenged. On the one hand, neurophysiologists persistently—and appropriately—question the advantage of MEG over scalp-recorded electroencephalograms and evoked potentials when the latter are analysed by sophisticated algo-

rithms. On the other hand, advocates of functional magnetic resonance imaging (MRI) and positron emission tomography have disparaged MEG because of the assumptions required to obtain unique solutions from magnetic field data. The paper by Maestú *et al* (this issue, page 208–212)<sup>1</sup> provides an example of work that is establishing the capabilities of MEG.

Maestú *et al* used MEG to compare patients with Alzheimer’s disease and elderly subjects. They had previously found that reduced MEG activation was present in left temporal and parietal areas during a probe letter working memory task; this reduction correlated with results from neuropsychological assessments and activities of daily living. In the present study, they report that

magnetic field measures correlate significantly with the relative volume of lateral and mesial temporal regions. Consistent with the long latency of cognitive evoked potentials, the MEG features of interest occurred more than 400 ms after stimulus onset. After a Bonferroni correction, the only correlation coefficient considered significant was that between left temporal lobe activity and the relative volume of the left hippocampus. The smaller the number of late MEG activity sources in the left temporal region, the greater the atrophy in the mesial portion of the left temporal lobe. Definable changes in MEG may correspond to the exquisite anatomic resolution of MRI.

Different types of algorithms, embodying different assumptions, have been employed to solve for the four-dimensional origin of electrical activity within the brain. The commonest algorithm models magnetic flux as the product of single equivalent current dipoles.

Despite its simplicity, this approach has given good results when compared with invasive recordings. A straightforward MEG technique is to plow mathematically through a second or so of evoked potentials, solving for dipoles every 4 ms and identifying those that show high goodness of fit and a restricted intracranial volume. Analysis of a single evoked potential can produce hundreds of dipoles scattered across multiple brain regions. This approach provides interesting results, and the assumptions behind it seem reasonable: but it's fair to ask whether all those dipoles represent something "real", or are merely artifacts of elaborate computer processing.

The report of Maestú and colleagues suggests that the plethora of dipoles is a valid reflection of brain activity. Significantly fewer of them are found in the atrophic left temporal lobes of Alzheimer's disease patients compared to elderly subjects. Although we still don't know what the individual dipoles

mean, we have increasing reason to believe that they do reflect neuronal processing. We can hope that a greater understanding of the link between MEG dipoles and cortical activity will lead to increased applications for the unique capabilities of MEG.

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Parkinson's disease

## Multidisciplinary rehabilitation for people with Parkinson's disease

E D Playford

### Worse general and mental health for patients receiving rehabilitation?

There is little evidence available to support or explain the role of rehabilitation in progressive neurological disorders, so the paper by Wade *et al* (this issue, page 158-162) is a welcome contribution.<sup>1</sup> The paper examines whether patients with Parkinson's disease have sustained benefit following a multidisciplinary outpatient rehabilitation programme. The programme comprised individualised multidisciplinary interventions and a group education programme and was delivered to six patients at a time over six weeks. The findings suggest that six months after randomisation those receiving rehabilitation had worse general and mental health and that their carers had a trend to more strain. This contrasts with findings from studies of the rehabilitation of people with multiple sclerosis<sup>2</sup> and brain injury.<sup>3</sup> Why should this conscientiously performed intervention have apparently negative effects?

The authors themselves suggest a number of reasons, including previously

satisfactory management leaving little room for improvement, limited input, little psychological or medical input, need for continued input, and finally, the use of inappropriate measures. These reasons may all provide an explanation for the lack of benefit but do not explain the negative impact. To explain this we should consider further the nature of the intervention and its assessment.

In this study the multidisciplinary therapy input was assessed using two main measures. The impact of the physiotherapy intervention was measured using a sit-stand-walk time and showed a trend towards improvement. The occupational therapy components were not measured directly and the speech and language therapy aspects were measured on a 5 point scale, which may be relatively unresponsive.

The authors also measured health related quality of life. Interestingly this deteriorated in the intervention group. Health related quality of life (HRQoL) may be defined as the patients' appraisal

of their current level of functioning and their satisfaction with it compared with what they perceive to be ideal. In the context of this study it will be influenced by the whole intervention, including the educative aspects of the programme. This will include the day to day education provided by the therapists as part of the treatment sessions, as well as the formal education programme. There is now considerable expertise in the areas of patient education. Teaching packages should enhance self-management and self efficacy.<sup>4</sup> The patients recruited to this study are not described in terms of disease severity and duration but patients early after diagnosis will have very different needs to those who are entering a more complex stage of the disease with an unpredictable response to medication. Mistimed education programmes may be seen as trivial or intrusive and distressing. There is evidence that significant parts of the variance of HRQoL are accounted for by mood<sup>5</sup> but, as pointed out by the authors, doctors and psychologists were not involved in this study.

This is a useful contribution to the literature but the findings emphasise the need for clarity when planning interventions and the measurement of their benefit. The design of rehabilitation trials is a developing science, but two questions need to be kept in mind. Firstly, does the intervention work and, secondly, is the intervention of value to the patient? In this study the intervention may have worked in some areas but not others, but despite this the patients still appeared to value it. Clarifying the

reasons for these findings is important so the design of future trials can be improved.

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**NEUROLOGICAL PICTURE**.....

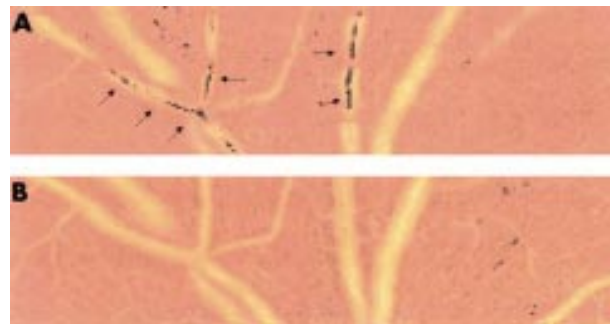
**Vasospastic amaurosis fugax**

A 54 year old man was admitted because of repeating (10–12/day) visual disturbances in the left eye. He reported shrinkage of the visual fields and a shadow-like visual impairment progressing to complete darkness within about 3 min and lasting about 10 min, followed by complete recovery. There was no personal history of hypertension, diabetes, and smoking. General medical examination was normal. The blood pressure was 130/80 mm Hg. Investigations of standard haematological and biochemical parameters revealed elevated low density lipoprotein cholesterol 4.45mmol/L (normal < 3.35 mmol/L) and elevated triglycerid level 2.67 mmol/L (normal range 0.45–1.80 mmol/L). Ultrasound studies of the extracranial vessel showed normal intima-media thickness (0.9 mm) and no plaque formation in the carotid bulb. Complete cardiological examination, including electrocardiography, transthoracic echocardiography, and chest x rays, were unremarkable. Magnetic resonance imaging of the brain was unremarkable, as was conventional angiography.

During examination of the retinal circulation by laser Doppler by scanning laser Doppler flowmetry<sup>1</sup> elaborating a map of perfused retinal vessels and capillaries, the patient suffered a visual disturbance as described above. The retinal perfusion map (fig 1A) revealed impaired perfusion of the retinal temporal artery at the 11 and 12 o'clock positions (arrows), which resolved within 21 min (Fig 1B). A treatment with statin to lower lipids and the antiplatelet agent copidogrel was initiated, but without influence on the events. Suspecting vasospastic amaurosis fugax,<sup>2–4</sup> we supplemented the therapy by calcium-channel blocker cycloandelate (1200 mg daily),<sup>2</sup> which yielded significant improvement of the clinical condition.

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The retinal perfusion map using scanning laser Doppler flowmetry demonstrates revealed impaired perfusion of the retinal temporal artery at the 11 and 12 o'clock positions (arrows in A) with resolution within 21 min (B).

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