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Familial temporal lobe epilepsy in the 19th century

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ABSTRACT

Purpose: To draw attention to a publication showing that familial temporal lobe epilepsy had been described in 1895, considerably earlier than 1994, usually thought to be the date of the original account of the disorder
Methods: Examination of the contents of Sir James Crichton-Browne's Cavendish lecture on 'Dreamy mental states' that was published in the *Lancet* in mid-1895
Results: At a time when the clinical phenomena that later became associated with the idea of temporal lobe epilepsy were beginning to become known, Creighton-Browne described the presence of this disorder in members of four consecutive generations of a British family throughout the course of the 19th Century
Conclusions: There is evidence that the genetic abnormality responsible for familial temporal epilepsy had probably appeared considerably earlier than hitherto thought

1. Introduction

Until quite recently, heredity was usually believed to be unimportant in the genesis of focal epilepsies, including ones of temporal lobe origin. In 1953, Paillas *et al.* [1] had considered the question of inheritance in temporal lobe epilepsy, but seemed unimpressed by the possible relationship. However, they mentioned the existence of two families, each with two members with this type of seizure disorder, but provided no further details. Then, in 1994, Berkovic *et al.* [2] described a family in which four members across two generations suffered temporal lobe epileptic seizures. Further report of familial temporal lobe epilepsies followed relatively quickly, most originating from Europe. The familial disorder began to be subdivided into mesial and lateral (neocortical) types, following the terminology of the then current ILAE classification. The former subtype, as mentioned, was first reported by Berkovic *et al.* [2,3], and the latter by Ottman *et al* in 1995 [4]. The patterns of inheritance of the subtypes differ [5], with the genetic basis of the autosomal dominant lateral subtype being identified in nearly half of the affected families [6]. The genetic basis of the mesial temporal subtype is not yet established. Such familial temporal lobe epilepsies are thought to be rare, though it has been suggested that failure to pursue the matter of family history in affected individuals after temporal lobe epilepsy has been diagnosed may have resulted in some familial instances escaping recognition. This suggestion has been supported by the findings of a recent study which provided evidence consistent with some 20% of non-lesional temporal lobe epilepsies having a genetic basis [7].

While contemporary medical awareness of the existence of genetically determined temporal lobe epilepsy dates only from 1994, there is published evidence that the disorder was present in a British family at least a century earlier. The relevant material is contained in a lecture given by Sir James Crichton-Browne in 1895. The lecture's title 'Dreamy mental states' gives little clue to the presence within its contents of the record of four generations of a family, some of whose members of each generation suffered very probable temporal lobe seizures [8].

2. Materials and methods

This paper summarises the relevant part (some 16%) of the text of Crichton-Browne's account in the *Lancet* [8], which was reprinted some 30 years later, with very slight alterations, in that author's collection '*Stray Leaves from a Physician's Portfolio*' [9]. A pedigree chart based on the data in the paper has been drawn up.

3. Results

On 20th June 1895 (or 30 June – in the reprinted version) Sir James Crichton-Browne (Fig. 1), the British Lord Chancellor's Visitor in Lunacy, delivered the Cavendish lecture entitled 'Dreamy mental states' before the West London Medico-chirurgical Society. The text of the lecture appeared in consecutive issues of the *Lancet* (6 and 13 July 1895). During the lecture Crichton-Browne described

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Fig. 1. Sir James Crichton-Browne (1840–1938). Courtesy of the Wellcome Library, London.

the seizure disorders which occurred in members of four generation of a family (Fig. 2). He seems to have based his account on contacts with members of Generations III and IV.

3.1. Generation I

The affected female was born toward the end of the 18th century. She was said to have been of a peculiar personality and, from girlhood, had 'spells of absent-mindedness'. In late life she developed epileptic seizures that began with an aura in the left hand and arm. She gave birth to 10 children (Generation II).

3.2. Generation II

No details were available concerning four of these children, three others had non-neurological disorders, another died in infantile convulsions, and one female, from whom the relevant members of Generation III were descended, had migraine with a visual aura. Crichton-Browne suggested that this migraine could be considered an 'allotropic form of a dreamy mental state or a sensory epilepsy'.

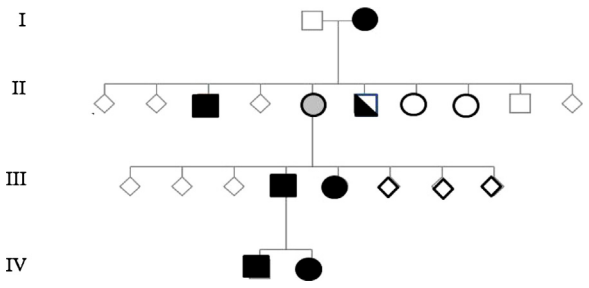

Fig. 2. Pedigree chart constructed from Crichton-Browne's 1895 account. Membership of generations II and III is not shown in order of date of birth, that information being unavailable. No information was available concerning those shown as faint symbols. Affected members are represented by solid black symbols, the half solid male in Generation II having died of infantile convulsions. The solid grey symbol in Generation II represents a migraine sufferer.

The one remaining male family member in Generation II was medically qualified. In youth and early adult life he had experienced brief seizures. In these he would suddenly become spatially disoriented and 'felt that it was a matter of pressing moment that he should recover his true position in space, and for a few seconds was in a tremor of perplexity and dread'. These episodes became decreasingly frequent as he grew older, but around the age of 40 he had his first epileptic fit. Subsequently he had further fits, mainly at night, though the episodes of disorientation did not return.

3.3. Generation III

No details were available for three of the 8 members. Three others died young. A son, around the age of 9, began to experience 'frightened feelings' with sudden loss of sense of personal identity and of his relationship to time and space. He experienced intense terror with violent palpitations, but never lost consciousness. The attacks always occurred when he was alone and mostly when going to bed. In adult life, just as he was falling asleep, the old terror experience and the palpitations would return.

His sister described very similar attacks which began when she was 10 year old. In them she experienced infinite distress, temporary loss of personal identity, embarrassed breathing and palpitations. The attacks lessened as she grew older.

3.4. Generation IV

The affected male member of Generation III had two children. At the age of 10 his son, nearly always when alone, experienced odd and horrid frightening feelings that lasted only a few seconds and were accompanied by a rapid heartbeat. During these episodes he would pinch his body and say to himself 'who am I?' As he was falling asleep he sometimes was disturbed by violent muscular jerks, and once experienced a numb feeling followed by shaking down his left arm. He also had a strange recurrent dream that became painfully familiar to him. At its onset, his body and his leg, if he attempted to move it, seemed enormously large, a 'vast haze' developed with vacancy and great fear and he saw black sticks or stripes. These seemed to close in on him before he woke, terrified. After treatment with bromides his attacks lessened, but continued until his 20th year.

His sister began having similar attacks at 8 years of age, when she was unoccupied. Her attacks caused uneasiness, but not terror. There was no palpitation. Once or twice her left hand was numb during the attacks and she had a transient hemianopia (side not indicated). When falling asleep she had constantly recurring dreams in which what she looked at receded and became abnormally small. She heard low murmuring crooning noises that she found horrid and alarming so that she jumped from bed and rubbed her ears. Sometimes there would be a loud ding in her ears as she fell asleep. Once she saw a spectral face. Her attacks were partly relieved by treatment and ceased by the age of 20.

4. Discussion

4.1. Crichton-Browne's role

James Crichton-Browne [10] had an influential career in British medicine, becoming a notable public figure and, in effect, almost its informal spokesman for neuropsychiatry over half a century. Appointed Medical Superintendent of the West Riding Lunatic