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Epilepsy and associated effects on adaptive behaviour in adults with Down syndrome

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A total of 201 adults with Down syndrome were investigated, of whom 32 (15.9%) had epilepsy. A bimodal age distribution for seizure onset in childhood and later in middle-age was found. Good seizure control was associated with early onset epilepsy. Down syndrome adults with epilepsy scored significantly higher overall on the adaptive behaviour profile but did not have significantly greater maladaptive behaviours.

Key words: epilepsy; Down syndrome; prevalence; adaptive; maladaptive; behaviour.

INTRODUCTION

Until recently, research studies had differed on whether there was no association between epilepsy and Down syndrome (DS)[1–3] or whether a significant association did exist[4–7]. During the past two decades such an association has been established, in particular between seizures and dementia in adults with DS[8–11]. Late-onset seizures are a strong indicator of an underlying dementing process[12].

A decline in adaptive skills with age has been demonstrated in adults with DS[13–14]. However, the underlying causes involved in this decline remain to be resolved but ageing *per se*, hypothyroidism, sensory impairment[15] and depression[16] have been demonstrated to be important factors. Whether epilepsy and long-term anticonvulsant therapy can, in individuals with learning disability (LD), also result in deterioration in skills remains controversial, although intellectual deterioration[17] and impairment of psychosocial functioning have been reported[18]. For people with LD, some studies have demonstrated an association between epilepsy and an increase in maladaptive behaviours[19,20] whereas other studies have found no such association[18,21].

This study aimed to examine the prevalence and type of epilepsy in adults with DS and to investigate an association between a history of seizures and level of adaptive functioning in this selected population.

PATIENTS AND METHODS

Adults (aged 16 years and over) with DS, involved in a longitudinal study investigating healthcare for adults with DS in the West Midlands were screened for a history of epilepsy. The study cohort was relatively representative of DS adults in the West Midlands; the cohort had a large sample size, subjects had been assessed clinically and cytogenetically for DS, had a wide age distribution, included both hospital residents and individuals living in the community and included subjects from differing districts in the region.

Severity of LD was assessed by review of previously reported intelligence tests' results, previous level of functioning as determined by review of medical notes, from carer interview and from mental state examination of the individual. Severity of LD was classified using ICD-10 criteria[22].

Carers were interviewed to ascertain information regarding frequency and type of epilepsy. Epilepsy was diagnosed if three or more seizures had occurred during a 2-year period, and/or if subjects were on regular anticonvulsant medication. Seizures were classified using

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the International League Against Epilepsy classification of seizure type[23]. Due to early work[24] highlighting poor diagnostic reliability of partial-complex seizures in people with LD by carers, these were not recorded. The age of onset and anticonvulsant medication prescribed was recorded. Medical and psychiatric records, interview with carers, mental state examination of subjects and venesection for haematological, biochemical and thyroid screening was performed to screen for concurrent medical or psychiatric morbidity. The level of adaptive functioning using The American Association of Mental Deficiency Adaptive Behaviour Scale (ABS)[25] was determined by carer interview. This scale is an observer-rated measure of adaptive behaviour with good validity and reliability. The ABS has two parts; Part I evaluates an individual's skills in 10 behaviour domains considered important in daily living, and Part II measures maladaptive behaviour related to personality and behaviour disorders. The domain 'use of medication' was excluded from Part II analysis.

Subjects with epilepsy but no other significant physical or psychiatric disorder were compared to age and severity of LD matched subjects without epilepsy and any other disorder. Exclusion of physical and psychiatric disorders enabled the effects of epilepsy and anticonvulsant medication *per se* on adaptive behaviour to be assessed.

RESULTS

Two hundred and one adults with DS participated in this study. Demographic details of the sample population are given in Table 1. The study population was relatively old for people with DS, with a mean age of 42.22 years; over 110 individuals (55%) aged 40 years and over and 17 (8.4%) aged over 60 years of age. Over three-quarters (78.6%) of the sample were living in the community. The most common form of severity of LD was moderate, in two-thirds of subjects.

Thirty-two (15.9%) subjects had a history epilepsy (Table 2), with the majority having onset of seizure activity after their third decade of life (seven in the fourth decade of life, five in the fifth decade and four in the sixth decade). Tonic–clonic seizures only (40.6%) was the commonest form of seizure disorder. Eighteen (56.3%) had good seizure control (no seizures during the previous 6 months) of which nine were of the 11 subjects with onset in childhood. Fifteen (83.3%) had only type of seizure disorder (13 tonic–clonic, one absence, one myoclonic).

Table 1: Demographic details of Down syndrome study population

Variable	Finding	
Sex distribution	Males	102 (50.7%)
	Females	99 (49.3%)
Mean age	42.22 years (S.D. = 12.51)	
Age range	16–72 years	
Residence	Hospital	43 (21.4%)
	Community units	73 (36.3%)
	Family home	85 (42.3%)
Severity of LD	Mild	38 (18.9%)
	Moderate	134 (66.7%)
	Severe	27 (13.4%)
	Unknown	2

Table 2: Findings for Down syndrome subjects with epilepsy

Variable	Finding (Sample population = 201)	
Frequency	32 (15.9%)	
Mean age	49.56 years (S.D. = 12.3)	
Age range	24–71 years	
Sex distribution	Males	11 (34.4%)
	Females	19 (59.4%)
Age of onset		
< 16 years	11 (34.4%)	
⩾ 16 years	21 (65.6%)	
Type of seizure disorder*	Tonic–clonic only	13 (40.6%)
	Absences only	6 (18.8%)
	Myoclonic only	2 (6.3%)
	Tonic–clonic and myoclonic	8 (25.0%)
	Tonic–clonic and absences	2
	Myoclonic and absences	1

* Partial seizures not assessed in study.

Anticonvulsant medication commonly prescribed included phenytoin, carbamazepine, sodium valproate and to a lesser extent phenobarbitone, diazepam and clonazepam.

Of the 32 subjects with epilepsy, 20 had a concurrent physical or psychiatric disorder (of which 11 had dementia) and were excluded from further adaptive functioning analysis. Of 169 subjects without epilepsy, 47 subjects were free of concurrent morbidity (no significant sensory impairment, no hypothyroidism, no psychiatric illness) and were used as a control group. Demographic details of the two groups are given in Table 3. The difference between the mean ages for the two groups was not statistically significant ($t = 1.42$, $P > 0.05$). There was no significant difference in proportion of male and female subjects in each group or in

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