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Orofacial Dyskinesia in Down's Syndrome

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Of 54 patients aged 30–60 years with a diagnosis of Down's syndrome, 38 had evidence of orofacial dyskinesia, assessed using the AIMS. There was a strong relationship between the presence of such movements and the severity of mental handicap. No relationship was found between abnormal movements and age. None of the patients had previously taken neuroleptic medication.

The prevalence of orofacial dyskinesia in the general population increases with age, with a prevalence of 6–8% in those aged over 65 years (Klawans & Barr, 1982). Schizophrenic patients administered neuroleptic medication have a higher rate (see Waddington, 1987), as do patients with bipolar affective disorder (Dinan & Kohen, 1989). The presence of orofacial dyskinesia is highly correlated with intellectual impairment (Waddington & Youssef, 1986). Schizophrenic patients with abnormal involuntary movements show impairment on a conceptual level analogy test (Struve & Willner, 1983) and on a paired associate learning test (Famuyiwa *et al*, 1979). It has been argued that the movement disorder and intellectual deterioration are manifestations of the same pathophysiological process (Waddington, 1987).

In a recent study of patients with senile dementia of the Alzheimer type we found that 83% had evidence of orofacial dyskinesia (O'Keane & Dinan, 1990). Those individuals with abnormal movements tended to have more paranoid ideation and higher neuroleptic exposure. Alzheimer's disease of the familial type and Down's syndrome have recently been linked with the finding that Alzheimer patients have a gene defect on the proximal part of the long arm of chromosome 21 (St George-Hyslop *et al*, 1987) and most middle-aged Down's syndrome patients have neuropathological changes indistinguishable from Alzheimer's disease (Oliver & Holland, 1986).

The extent of orofacial dyskinesia in Down's syndrome has not previously been reported. Gualtieri *et al* (1986) investigated the presence of such abnormal movements in young mentally retarded individuals. All patients had been exposed to neuroleptics and precise reasons for the mental handicap were not taken into account. Thirty-four per cent of the sample had evidence of persistent orofacial dyskinesia. We decided to investigate the presence of such movements in Down's syndrome patients who had no exposure to neuroleptics.

TABLE I
Characteristics of patients with and without abnormal involuntary movements

	With	*Without*
Age: years		
30–40	14	7
40–50	15	6
50–60	9	3
mean ± s.e.m.	42.5 ± 1.2	44.0 ± 2.1
Men	9	4
Women	29	12
Handicap		
moderate	16	13
severe	22	3

Method

Sixty-two long-stay patients, aged 30–60 years, were randomly drawn from the institutional patients in the catchment area who had a diagnosis of Down's syndrome. Patients with a history of psychotic episodes, epilepsy, neuroleptic exposure, or a score of greater than 4 on the Hachinski index (Hachinski *et al*, 1975) were excluded. The latter criterion ensured that patients with multi-infarct cerebral deterioration were omitted. This resulted in the exclusion of eight patients, leaving a study group of 54 (41 women, 13 men). All patients were assigned to a DSM–III category (American Psychiatric Association, 1980) of mild, moderate, severe, or profound retardation as assessed by their overall adaptive behaviour/social maturity and IQ (WAIS). They were then assessed using the Abnormal Involuntary Movement Scale (AIMS; Guy, 1976). This scale measures the presence and severity of abnormal movements in seven body regions: (a) muscles of facial expression, (b) lips and perioral area, (c) jaw, (d) tongue, (e) arms, (f) legs, and (g) trunk. Each area is rated 0–4 (0, none; 1, minimal; 2, mild; 3, moderate; 4, severe). Assessment of dyskinesia was made blind to assessment of intellectual functioning.

Results

Thirty-eight (29 women, 9 men) of our sample of Down's syndrome patients had evidence of orofacial dyskinesia as judged by a score of 3 or more on at least one body area on

the AIMS. The mean (± s.e.m.) total AIMS score was 12.1 ± 0.93 (men: 12.1 ± 1.8; women: 12.1 ± 1.1). A comparison of patients with and without involuntary movements is made in Table I. More patients in the severely handicapped group had evidence of movement disorder than in the moderately handicapped group ($\chi^2 = 6.9$, d.f. = 1, $P \leqslant 0.05$). When patients are grouped on the basis of age (30–40, 40–50, 50–60 years) there seems to be little difference in the prevalence of dyskinesia in the different groups, with each group having a prevalence of around 70% ($\chi^2 = 2.72$, d.f. = 2, NS). When age groups are compared on the basis of severity of orofacial dyskinesia again no relationship is observed ($\chi^2 = 0.18$, d.f. = 2, NS). Severe dyskinesia is as likely to occur in young as in old patients.

Discussion

The principal finding in this study is the high level of orofacial dyskinesia present in Down's syndrome over the age of 30 years. This is considerably higher than reported for mental handicap as a whole (Kalachnik, 1983). The dyskinesia occurred in the absence of neuroleptic medication. As tardive dyskinesia is a disorder with a multivariate aetiology, where the ageing process and neuroleptics are principally implicated, the use of neuroleptics in Down's syndrome should be undertaken with caution and only for specific indications.

Studies of orofacial dyskinesia in the general population (Klawans & Barr, 1982), in schizophrenia (Waddington, 1987) and in bipolar affective disorder (Dinan & Kohen, 1989) have almost all found an increased prevalence with increasing age. In Down's syndrome we have been unable to demonstrate any relationship between the prevalence or severity of orofacial dyskinesia and age. As the level of intellectual handicap in the three groups was approximately equal, one is left with the conclusion that the ageing process is not a significant factor in the genesis of the condition in such patients. Perhaps differences might emerge using a larger sample. Nonetheless, on the basis of this preliminary investigation, it is tempting to suggest that the neuropathological changes in adult Down's syndrome patients predispose not only to the intellectual handicap but also to the development of abnormal involuntary movements. This is further supported by the fact that patients with most severe forms of Down's syndrome, as assessed by level of intellectual handicap, had the highest prevalence of abnormal involuntary movements.

The study has implications for the pharmacological management of behavioural disturbance and psychotic illness in patients with Down's syndrome. With such a high prevalence of movement disorder in the group further studies in such patients may throw light on the pathophysiology of orofacial dyskinesia not just in Down's syndrome but in other conditions as well.

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