Unlike these cases, the patient reported here had chronic psychosis lasting for several years associated with dependent abuse of mephentermine.

It appears that our patient developed chronic amphetamine psychosis – although not the classical paranoid type – characterised by continuous non-affective auditory hallucinations, thought broadcasting, and psychotic withdrawal, on abusing dexamphetamine for 2 years. Since the publication of a monograph by Connell (1958), amphetamine psychosis is well recognised. Connell described the clinical picture as "primarily a paranoid psychosis with ideas of reference, delusions of persecution, auditory and visual hallucinations in a setting of clear consciousness", and suggested that it might be indistinguishable from acute or chronic paranoid schizophrenia. The psychosis in our patient persisted for 3 years after discontinuation of amphetamine, probably because he started taking mephentermine within a month. Subsequently, the psychosis remitted and the patient was able to complete his medical training. On restarting mephentermine abuse, this time by injection, he again developed psychosis resembling schizophrenia, characterised by ideas of reference, delusion of black magic, visual hallucinations, and psychotic withdrawal. It lasted as long as mephentermine abuse continued (i.e. for 5 years) and responded well to haloperidol.

It is interesting to note that all three cases reported by Angrist *et al* (1970) had received diagnoses of schizophrenia at least once during their several previous admissions. In one case the diagnosis of chronic schizophrenia was confirmed. Hence, in view of the long duration of psychosis in our patient, the question arises as to whether he had primarily a schizophrenic illness. Escobar & Karno (1982) have reported a case of chronic hallucinosis of more than 20 years' duration, misdiagnosed as schizophrenia, associated with chronic use of nasal drops containing sympathomimetics other than mephentermine. Another case reported by Snow *et al* (1980) had addiction to intranasal phenylephrine associated with paranoid psychosis, but it is not clear how long the psychosis persisted. In our patient, psychosis surfaced both times only on chronic abuse of a CNS stimulant, and there was a spontaneous remission lasting for several years. In addition, the psychosis remitted within 2 weeks of starting haloperidol. Thus it is unlikely to be schizophrenia. Being schizoid premorbidly, probably the patient had a vulnerability to develop schizophrenia-like psychosis even with a very weak CNS stimulant like mephentermine. This case illustrates that drugs which are weak CNS stimulants can lead to dependence as well as the development of chronic psychosis in vulnerable individuals.

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Benign Hereditary Chorea

A Case Report

A case of benign hereditary chorea is reported, along with a brief review of the condition. It is a rare and little known autosomal dominant disorder which may be confused with

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Huntington's chorea, which has a more serious prognosis. The case described shows some characteristic features. It is difficult to decide how many of the patient's psychiatric difficulties are a reaction to his disabilities and how many have an organic substrate.

Only in 1966 was benign hereditary chorea (BHC) recognised as a separate entity (Pincus & Chutorian, 1966), and although a large number of cases have been reported since, it remains little known in psychiatric circles. Sufferers are often misdiagnosed (Chun *et al*, 1973; Sleigh & Lindenbaum, 1981).

Affected individuals show choreic or choreo-athetoid movements, which are symmetrical and most prominent in the trunk and head, less so in the limbs. The chorea is continuous but fluctuating, being worse when the patient is tired or stressed, stopping only during sleep. Onset is early in life, almost always in the first decade, often in the first or second year. From childhood the movements tend to maintain the same intensity, sometimes improving towards middle life, perhaps as a result of learned voluntary control (Sleigh & Lindenbaum, 1981). Children affected particularly early in life are often delayed in motor development.

Less often seen are wide-based unsteady gait and intention tremor (Pincus & Chutorian, 1966; Chun *et al*, 1973; Sleigh & Lindenbaum, 1981), and dysrhythmic speech caused by involuntary movements interfering with the smooth expulsion of air (Chun *et al*, 1973).

The title *benign* hereditary chorea was given to emphasise that a progressive dementing process does not occur, but it is increasingly recognised that patients frequently show poorer intellectual function than unaffected siblings, particularly on tests of verbal skills (Leli *et al*, 1984); indeed, Bird *et al* (1976) have stated "the maladroitness and . . . differential between verbal and performance IQ scores in the [described] proband support a mild neurological deficit beyond simple chorea".

Psychiatric problems have very often been recognised in sufferers, although they have usually been described simply and briefly as "behavioural problems" or "anxiety" and attributed to "physical problems" or the consequences of stigma (Bird *et al*, 1976; Sleigh & Lindenbaum, 1981). It has also been suggested that diffuse cerebral involvement in the disease process might be a cause (Bird *et al*, 1976).

The underlying biochemical mechanism is presumed to be relative dopaminergic overactivity due to lesions in the caudate, although the intention tremor may be due to extension into the dentate nucleus/cerebellar system in some cases (Bird *et al*, 1976). Investigations into a number of cases have revealed no consistent biochemical, radiographic, or electroencephalographic abnormality (Sleigh & Lindenbaum, 1981).

Authors to date have not agreed as to the best treatment; theoretically, dopamine antagonists should be useful (De Silva, 1977), and phenothiazines have been successful (Pincus & Chutorian, 1967). However, several cases have seemed refractory to treatment (Haerer *et al*, 1967; Sleigh & Lindenbaum, 1981). One case reported showed a response to steroids, presumably due to modulation of neurotransmission (Robinson & Thornett, 1985).

Inheritance of BHC is by an autosomal dominant gene with partial penetrance, near complete in the male, 0.75 in the female (Harper, 1978). An autosomal recessive subtype may also occur (Nutting *et al*, 1969). The prevalence would seem to be about 1 in 500 000 (Harper, 1978), but Sleigh & Lindenbaum (1981) have pointed out that the condition may be much commoner.

The differential diagnosis is well discussed elsewhere (Bird *et al*, 1976; Harper, 1978; Sleigh & Lindenbaum, 1981), but clearly the distinction from Huntington's chorea (HC) is of special importance. HC very rarely presents in childhood, and when it does, adult relatives with the disease are usually to be found. Juvenile HC tends to present with marked rigidity, and the accompanying chorea is less prominent than in the adult form; seizures are common and the electroencephalogram is abnormal; there is progressive dementia and an early death. Other differential diagnoses include Sydenham's chorea, cerebral palsy, drug effects (particularly dopamine antagonists and oestrogens), Lesch–Nyhan syndrome, and Wilson's disease.

Case description

The patient is a 35-year-old, recently separated, unemployed man who lives alone. He was referred to the psychiatric out-patient clinic with a six-month history of anxiety and depression, and has received intensive treatment in the day hospital.

Over the years he has received much psychiatric treatment, both as an in-patient and out-patient; in the past he has been variously diagnosed as suffering personality disorder, neurosis, alcoholism and benzodiazepine dependence. Previous treatments have been psychological in approach. He tends to be a rather dependent individual, readily settling to institutional life. Outside hospital he has had a number of relationships with women, all of which seem to have foundered because of his excessive reliance on them.

He is the youngest of five sibs and was born uneventfully after a straightforward pregnancy. At the age of three it was noted that his speech was hard to understand, and he was referred to a speech therapist, who was critical of his

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efforts. Abnormal movements of his head and neck were noticed at about the same time, but were considered to be tics.

At school he was a 'nervous child' and he was teased by his peers. In particular he recalls being encouraged to speak so that his disability could be mocked. The 'nervousness' had some compensations: he tended to escape punishment. He initially performed well intellectually, but his performance subsequently declined and he left school at 16 with four O levels. After school he began work as a salesman, which proved impossible, despite sympathetic employers. Subsequently he has worked as a computer technician, although often unemployed.

His mother is a rather controlling, strong-willed, tough woman for whom he has respect as much as affection. His late father was more sensitive, a heavy drinker with whom he often clashed.

His first psychiatric contact was at 19 when, following encouragement from a girlfriend, he visited his GP for help and was referred on because of his heavy drinking, a habit which reduced his considerable social anxiety and also improved his abnormal movements and speech.

The disorder occurs in other members of his family. His late father was diagnosed as suffering from St Vitus's dance and used to drink heavily, at least in part to reduce his abnormal movements. His eldest brother, a successful businessman, has considerable speech difficulties, although the abnormal movements are less of a problem. He currently receives MAOIs for depression. Two of his brother's children have the disorder, and the elder, a 23-year-old woman, has also found alcohol a useful palliative. Another nephew has recently developed tics and habits, but no formal diagnosis has been made.

The condition has remained static over the years, tending, if anything, to improve slightly. At no time has he had gait disturbance.

Before the start of treatment, examination showed a labile, anxious, unhappy man of normal appearance and dress. Rapport was good and his affect warm. He had an odd speaking style. His talk was hesitant but rapid with bursts of speech; often he would begin to say something but abandon the effort shortly afterwards, only to start on a new theme. Occasionally he would stammer or stutter. There were choreiform movements, particularly of the arms, shoulders, trunk and neck, which he occasionally attempted to disguise by pretending voluntary intent. Both the speech disorder and the movements varied considerably depending on the degree of anxiety, worsening if he became flustered. He had a gentle but dry sense of humour in which his deficiencies in both speech and personality were wryly acknowledged.

Routine investigations including full blood count, VDRL, thyroid function, electrolytes and liver function tests are normal; a CAT scan with enhancement shows slight generalised prominence of sulcal markings over both cerebral hemispheres. There is also a suggestion of minor temporal lobe atrophy, with minor bilateral prominence of the Sylvian fissures. His electroencephalogram is normal. Psychometry gives a Wechsler Adult Intelligence Scale full score of 114, a verbal score of 117, and a performance score of 109; the differences are not significant.

Treatment in the day hospital has consisted of an anxiety management programme combining cognitive techniques and relaxation. In addition to this he has been given a behavioural programme in which he has to tackle a progressive series of tasks which he finds difficult; the tasks involve social interactions such as shopping and negotiating with the Department of Health and Social Security.

To date a completely satisfactory drug treatment eludes us. Haloperidol in a dose of 3 mg b.d. substantially improves the chorea so that, when he is calm, abnormal movements are not apparent, but this is at the cost of coarse tremor and distressing akathisia, despite the addition of procyclidine; a slightly lower dose may prove to be more beneficial.

The anxiety management training has been of only limited use; when anxious, a state readily provoked by attempts at social interaction in new situations, the movements return and his speech may deteriorate to the point of incoherence. This experience is so upsetting that it seems that the coping strategies he has been taught are swamped. Despite perseverance his anxiety has not attenuated. The only effective remedy in these circumstances seems to be prophylactic pharmacological anxiolysis; both alcohol and lorazepam work well, and we believe the latter is preferable. Because of the likelihood of dependence developing, he is currently limited to 15 mg weekly.

Discussion

The case reported illustrates a number of features typical of BHC, in particular the movement disorder, its form, age of onset, and variation with affective state. The case also shows associated psychiatric problems which demonstrate the inappropriateness of the title *benign* hereditary chorea, and raises questions as to how much these problems are 'reactive' and how much the result of diffuse cerebral involvement in the disease process. Similar problems have been less clearly described in other cases (Sleigh & Lindenbaum, 1981).

Our patient suffers considerable distress and social disability, and has demonstrated the problems that can arise if alcohol is used as self-medication, especially when it is so effective.

The organicity of his movement disorder seems to have prevented full success of standard behavioural techniques.

It is important to make the correct diagnosis in these patients so that appropriate counselling and therapy can be carried out to maximise social abilities, minimise suffering, and avoid the development of alcohol dependence. It will also allow the best, albeit inadequate, drug treatment.

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Failure to Convulse with Electroconvulsive Therapy

A Case Report

A case is reported of a severely depressed patient in whom treatment with electroconvulsive therapy was abandoned because of repeated failure to induce a convulsion. Reasons for the failure to convulse and alternative treatment approaches are discussed.

There is now substantial evidence that electroconvulsive therapy (ECT) is an effective treatment for severe depression (Johnstone *et al*, 1980; Brandon *et al*, 1984; Gregory *et al*, 1985). This treatment produces a generalised convulsion by passage of electric current through the brain. The convulsion manifests as a brief initial stimulus-related tonic and subsequent longer clonic phase. It is the latter that is associated with therapeutic efficacy (Ottoson, 1962). Failure to recognise non-occurrence of the clonic phase is common (Pippard & Ellam, 1981). Careful monitoring can avoid this pitfall (Fink & Johnson, 1982). We describe a case in which no clonic phase was observed during 15 treatment applications despite clinical and 'cuff' monitoring (Addersley & Hamilton, 1953).

Case history

A 32-year-old right-handed woman of mixed African and Indian extraction, and with a two-year history of recurrent depressive disorder, was admitted to hospital during her third episode. This was characterised by unremitting low mood, early morning wakening, reduced appetite, weight loss of 5 kg and a suicide attempt. Treatment with amitriptyline and chlorpromazine for four weeks effected no improvement. ECT was then prescribed and administered in eight sessions over four weeks with a total of 15 applications (see Table I).

Convulsions were monitored for the first eight applications by clinical observation only. This was supplemented for the remaining seven applications by the 'cuff' method: a sphygmomanometer cuff was placed around the right upper arm and was inflated to above systolic pressure prior to the injection of succinylcholine, thus allowing observation of unmodified clonic contractions. On each occasion that current was applied a brief tonic phase was observed, but this was never followed by a clonic phase indicative of continuing seizure activity.

Scrupulous attention was paid to electrode application: the hair was parted and the skin was cleaned with Savlon. Lint-covered steel electrodes were soaked for 30 seconds in Ectronolyte and applied with firm pressure. For unilateral applications the electrode positions were temporoparietal

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