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Six Cases of Gilles de la Tourette's Syndrome

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Summary. Six cases of Gilles de la Tourette's syndrome are discussed in the light of conflicting views on the aetiology of the condition. It is hypothesized that the onset of coprolalia in patients with persistent childhood tics indicates a disturbance of the normal balance between a need for tension relief by swearing and a capacity to control such vocal activity.

Some conclusions are drawn on the management of the syndrome by the use of butyrophenones, massed practice of tics, and the promotion of personality development. A flexible approach geared to the individual patient's particular needs is recommended.

Introduction

Gilles de la Tourette's syndrome (1885) is characterized by the following essential features: (1) Childhood onset (below the age of 16); (2) Multiple motor tics; (3) Unprovoked loud utterances which may progress to the forced shouting of obscenities (coprolalia).

A review of the condition (Fernando, 1967) was based on 69 cases which showed these features, and referred to 20 others which had been reported as cases of the syndrome. Several further case reports published in the English language literature are listed in Appendices 1 and 2. Although most case reports are from the United States and Europe, the condition is found in all parts of the World (Shapiro *et al*, 1972b). Reports from Germany, Switzerland, Poland, France, Finland and Canada were noted earlier (Fernando, 1967). Cases have also been reported from Ireland (Healey and Fischer, 1965), Sweden (Eriksson and Persson, 1969), Australia (Ellison, 1964; McKinnon, 1967; Boris, 1968), New Zealand (De Groot and Bardwell, 1970), India (Prabhakaran, 1970), Israel (Lerman and Nussbaum, 1974; Gadoth, 1974) and South Africa (Grover *et al*, 1967). Two new cases seen by the author, together with follow-up reports on four other patients are presented here, and some observations are made on the aetiology and management of the condition.

Case Reports

Case J.T.

A 43-year-old married housewife was referred to the author in December 1968. She was the younger of two children and the only girl born to French parents. Her father was a violent man who drank heavily. Her childhood was characterized by constant parental quarrelling. There was no family history of tics but a maternal aunt was probably psychotic. Early development was normal until the onset of blinking and grimacing at the age of 8–9. She was unhappy in her first school, which she started attending at the age of 6. The tics began a few months before she was removed from that school, and about the same time she was frightened by being bitten by a dog. She was very happy at her next school, but the tics continued unabated, although she saw several doctors over the next two years.

She left school at 14, just before the outbreak of war in 1939. Jerks of her limbs began about one year later, and all her symptoms progressively worsened over the next four years, which were characterized for the patient by the social disruption caused by the German occupation of France, parental dissension, and the illness of her mother. Her parents separated in 1944 and her mother died one year later. Her future husband, whom she had originally met in 1940, renewed contact in 1945 and she came to England two years later to join him; but she was found to be suffering from tuberculosis requiring hospital treatment and therefore did not get married until 1951. Muffled cough-like vocal tics began in 1948 and she began to utter non-specific explosive noises 4–5 years later. All her tics worsened progressively over the next 15 years, resulting in a considerable restriction of her social life. Her symptoms were unaffected by three pregnancies, except for transitory improvement after the delivery of her first child in 1951. Treatment with carbon dioxide inhalation in 1949 and with chlordiazepoxide between 1962 and 1968 had no effect.

When seen by the author in December 1968 the patient exhibited occasional blinking and frequent jerking movements of all limbs, which were sometimes co-ordinated into a 'jump'. The movements were usually accompanied by explosive noises, sometimes recognizable as the words 'bugger' and 'maman'. The tics were absent during sleep, reduced in intensity when the patient was relaxed, and exacerbated under social stress. They were voluntarily controllable for three or four minutes and occasionally disappeared for up to five minutes while the patient's attention was absorbed in a television programme or deep conversation. There was no evidence of echolalia.

The patient was diagnosed as suffering from Gilles de la Tourette's syndrome. There were no obsessional symptoms or signs of depression or other psychiatric illness. Her intelligence quotient was 92 (WAIS). Her responses to the Rorschach test were 'controlled, defensive and conventional'. Electroencephalography showed mild non-specific abnormalities with no evidence of lateralization or localized abnormality. Two sessions of direct suggestion under hypnosis produced no lasting effect. Treatment with haloperidol and orphenadrine hydrochloride produced dramatic improvement when the dosage of the former was increased to 7·5 mg daily. However, the patient refused to continue medication because (according to her husband) she felt that it 'changed her personality to a point which frightened her'. Inquiry in July 1970 revealed that her symptoms and her attitude towards medication persisted. Several attempts to locate her in 1974 and 1975 have been unsuccessful.

Case M.M.

A 31-year-old single man consulted the author in May 1970. He was an only child born to middle-class American parents. There was no family history of tics or mental illness. Early development was normal and his childhood happy until he started blinking and making 'ah' noises at the age of 8. The symptoms started soon after the death of his grandfather and his own tonsillectomy. He was seen by a paediatrician, but no treatment was offered. The tics worsened progressively, and he developed coprolalia at the age of about 21. In spite of restrictions due to his symptoms the patient did well at school and later wrote poetry, acted on the stage and read on the radio. He claimed that all his tics, including vocalizations, were completely absent while he was on the stage or absorbed in reading over the radio.

He was treated with hypnosis at the age of 18, without any lasting benefit. He received psychoanalytic psychotherapy between the ages of 21 and 26—again without any apparent benefit. In May 1965 (aged 26) he started haloperidol medication with dramatic improvement of all his tics. He was then able to obtain work on the open market for the first time in his life. However, he found that he had lost his 'creativity' and he stopped acting and writing poetry. In October 1968 he developed a tolerance to haloperidol. He experimented with the dosage to find that he derived greatest benefit from the drug by alternating between 2–3 months off it (with severe symptoms) and 2–3 months on it (with partial relief of tics) at a dose of 6–15 mg daily.

When seen by the author in 1970, the patient showed jerks of all his limbs, with an occasional explosive utterance of a muffled obscenity, although he was taking haloperidol. It was confirmed that an EEG and other investigations done abroad had revealed no evidence of an organic neurological lesion. Haloperidol was discontinued and the patient was started on triperidol with very good effect. He was stabilized on a maintenance dose of 6–8 mg daily until after six months he suddenly developed a tolerance to this drug. However, he was then able to switch back to haloperidol with good effect. Recent inquiry (May 1975) reveals that his symptoms have been well controlled on haloperidol for over four years.

Case M.P. (Fernando, 1967).

This woman is now aged 31, single and living with her parents while working in regular employment. She has been symptom-free for over 14 years without any medication (May 1975). She attributes her past illness to the disturbed childhood relationship with her father and the (later) mental strain of being 'observed by people'— a reference to her frequent contact with doctors.

Case C.H. (Fernando, 1967)

This man is now aged 41, divorced from his wife on the ground of (her) adultery and courting a widow of about his own age. He continues to maintain a good work record. When he started on haloperidol in June 1966 he required a daily dose of 7·5 mg for maximal symptom control. Two months later his 'requirement' was 3·0 mg and two and a half years later (May 1969) he had managed to reduce the dose to a mere 1·5 mg. His motor tics are completely controlled on this dosage, while the vocal tics are sufficiently modified to cause no social embarrassment. His improvement has been maintained for nearly nine years (May 1975).

Case C.S. (Fernando, 1968)

This woman is now a 30-year-old married woman with two children. She sought psychiatric treatment for anxiety and tics in 1965/6 and again in May 1967, without any benefit. Reports from her general practitioner indicate that she recovered from her

tics around 1968 without receiving any specific medication. Although she suffers from headaches, tiredness and tension, she remains free of tics (May 1975).

Case P.P. (Fernando, 1968)

This patient is now a 34-year-old married woman with five children. She improved considerably between the ages of 19 and 21. When seen in November 1970 she was troubled by a solitary tic of her trunk and an occasional non-specific 'hic'-like vocal tic. She found her tics tolerable and declined to take any specific medication for them. Her condition had remained unchanged for nearly 13 years when inquiry was last made (November 1974).

Discussion

A syndrome in which childhood tics continue into adult life, becoming associated with the explosive shouting of obscenities, is likely to have a complex causation. Corbett *et al* (1969) suggested that it is simply a severe form of tics, but their study of 180 childhood tiqueurs revealed no evidence relating to the aetiology of tics. Morphew and Sim (1969) analysed 43 case reports of Gilles de la Tourette's syndrome to find support for a 'functional rather than an organic basis for the condition'. However, a direct study of 34 patients suffering from the condition (Shapiro *et al*, 1972b) showed no common psychopathology. On the other hand, Sweet *et al* (1973) found a sufficient number of minor abnormalities in the majority of 22 cases studied by them to suggest a neurological basis to the disorder. Tics developed soon after head injury in a patient described by Sanders (1973) and in another reported by Eriksson and Persson (1969). None of our six patients had abnormalities of the central nervous system, but there was firm evidence of disturbed family relationships during childhood in three cases (J.T., M.P. and C.S.). Moldofsky *et al* (1974) found that ten out of fifteen patients had a family history of tics, but Shapiro *et al* (1972a) found such a family history in only four of their thirty-four patients. Sanders (1973) reported a father and son suffering from the condition; and Friel found three cases occurring in one family—two sisters and the son of one of them. There is an isolated report of a chromosome abnormality, namely an XYY karyotype, associated with the condition (Merskey, 1974). None of our six patients had a family history of tics; chromosome analysis was not carried out on them. Morphew and Sim (1969) considered that psychological stresses were often associated with the onset of tics and such precipitants were reported in one third of cases reviewed earlier (Fernando, 1967). Tics seem to have followed stressful events in four of our six patients.

Moldofsky *et al* (1974) examined clinical, biological and psychological variables in fifteen patients to postulate that cases fall into two types: a genetically determined metabolic disorder or an acquired condition relating to physical and psychological traumata. Although our six cases are the latter type, the claim made by these authors that haloperidol is less beneficial in this than it is in the genetic variety was not substantiated. Butyrophenones (haloperidol or triperidol) gave symptomatic relief to all three patients who took these drugs. In fact, tics of all kinds are probably suppressed by haloperidol (Connell *et al*, 1967), and its value in the treatment of Gilles de la Tourette's syndrome is well established (Fernando, 1968). It is possible, however, that brain damage may militate against its efficacy: of the four patients who failed to benefit from the drug in a series reported by Moldofsky *et al* (1974), two had been leucotomized before the drug was used.

Abuzzahab and Ehlen (1971) described two patients whose symptoms became tolerant to haloperidol. One of our patients (M.M.) developed such a tolerance but re-established sensitivity to the drug after switching to triperidol for six months. Moldofsky *et al* (1974) reported the 'loss of vivacity and spontaneity' as a side effect of haloperidol. Two of our patients reported subtle changes of personality coincident with relief of symptoms by haloperidol. One man (M.M.) lost his creative ability, and a woman (J.T.) was so frightened by her subjective feelings that she preferred her symptoms to the overall effect of the drug. It is possible that long-term medication with haloperidol may have serious effects on personality which are not easily apparent.

Clark (1966) has offered an explanation for Gilles de la Tourette's syndrome in terms of learning theory: motor tics are learned because they reduce high autonomic drive, but being

activators of cortical arousal mechanisms they produce cortical inhibition levels far above those which allow adequate motor and verbal control. Treatment based on learning theory (Yates, 1958) which involves the massed practice of tics was successfully used in the case of an 11-year-old boy (Walton, 1961) and in two adult cases (Clark, 1966), but was ineffective (Feldman and Werry, 1966) or only partially helpful (Sand and Carlson, 1973) in two other cases. Reinforcement techniques based on operant conditioning were successful in alleviating tics in a 12-year-old boy suffering from Gilles de la Tourette's syndrome (Rosen and Wessner, 1973), but Jeste *et al* (1973) report that behaviour therapy in the form of negative practice was ineffective in a 13-year-old patient presenting with this disorder. No form of behaviour therapy was used in any of our cases, though Clark (1966) claims that massed practice may be the treatment of choice in Gilles de la Tourette's syndrome.

Three of the six patients reported in this paper have never received haloperidol or behaviour therapy. In fact, they all appear to have improved spontaneously or as a result of changes in life situation. They have remained well for 7 to 14 years, leading normal lives without any specific medication. Each of these three women had a classical history of the condition resulting in severe disruption of adolescence and in two cases childhood. One (M.P.), who was given a poor prognosis and considered untreatable in her teens, recovered completely on leaving her parents. Her comments to the author indicate that getting away from doctors may have been as therapeutic to her as leaving home. The other two women seem to have improved as they matured and became established as housewives and mothers. Goforth (1974) too has reported a female patient whose improvement was associated with personality maturation attributable to psychotherapy. The two male patients reported in this paper continue to take haloperidol. However, one (C.H.) is on a minimal dose, which may be having a placebo effect only, while the other (M.M.) volunteers his subjective impression that 'an automatic remission in the symptoms has occurred'. Thus a good outcome is evident in all five patients who have been followed-up well into adulthood. The likelihood that prolonged spontaneous remission has taken place is high in three of them and reasonably high in the other two.

Conclusions

The aetiology of Gilles de la Tourette's syndrome remains uncertain. Tics occurring in children may be understood in terms of organic, developmental, behavioural and emotional models (Corbett *et al*, 1969). Swearing is a normal occurrence related to socio-cultural factors and psychological need—mainly tension relief (Montague, 1967). Its appearance as an involuntary vocal activity may reflect an increased need for such activity or a reduced capacity to control it. The latter may be related to hereditary and/or organic factors, while the former is likely to indicate emotional disturbance. It is hypothesized that coprolalia emerges when there is an alteration in the balance between control and need; remission of symptoms would therefore occur when the balance is restored either by an increase of control or by a reduction of need.

The effectiveness of butyrophenones in suppressing tics and of massed practice in alleviating or eradicating them must be seen in the context of the likelihood of spontaneous remission of the syndrome which seems to be associated with personality maturation. It is therefore suggested that there are three main aspects to the management of this syndrome, namely:

(1) The use of haloperidol or triperidol in the short term, alternating the drugs if tolerance develops to either one.
(2) The use of massed practice of tics as an alternative to medication.
(3) The promotion of personality maturation with an emphasis on encouraging independent behaviour and discouraging dependence.

The degree of importance to be given to one or other of these aspects of management must depend on the individual case: the taking of drugs or undergoing behaviour therapy may interfere with the third aspect of management by discouraging self-reliance, while some

patients, particularly adolescents, may find it impossible to achieve independence until some degree of symptom control has been established. It is therefore important that the management of the condition should be sufficiently flexible to be geared to the individual patient's needs at any particular time.

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APPENDIX 1

Reports of cases showing essential features of the syndrome

Author	Year	Sex and age at reporting of patients
Ellison	1964	One male aged 67
Healy and Fischer	1965	One male aged 9
Clark	1966	Two males aged 17 and 22
Feldman and Werry	1966	One male aged 13
McKinnon	1967	One male aged 31
Connell *et al*	1967	Two males aged 12 and 15
Grover *et al*	1967	One male aged 15/16
Lucas	1967	One male aged 9 and one female aged 9
Fernando	1968	Two females aged 23 and 26
Boris	1968	One male aged 14
Bruch and Thum	1968	One male aged 12
Eriksson and Persson	1969	Two males aged 15 and 22
Morphew and Sim	1969	One male aged 14, two males aged 34–35 and three females aged 31–36
Prabhakaran	1970	One male aged 15
De Groot and Bardwell	1970	One male aged 17
Abuzzahab and Ehlen	1971	Two males aged 14 and 15, two males aged 30 and 41 and three females aged 17, 25 and 43
Shapiro *et al*	1972	Twenty-seven males and seven females (individual ages not specified)
Clements	1972	One male aged 16
Sweet *et al*	1973	Ten males aged 8–16, three males aged 19–20, four males aged 34–61, one female aged 10 and four females aged 26–31
Sanders	1973	Two males aged 23 and 54
Rosen and Wesner	1973	One male aged 12
Jeste *et al*	1973	One male aged 13
Friel	1973	One male aged 11 and two females aged 28 and 34
Sand and Carlson	1973	One male aged 9
Moldofsky *et al*	1974	Two males aged 11–14, three males aged 17–18, nine males aged 21–40 and one female aged 24
Merskey	1974	One male aged 19
Goforth	1974	One female aged 42

APPENDIX 2

Reports of cases excluded from Appendix 1

Author	Year	Sex of patients	Reason for exclusion
Savin	1961	Three females and two males	Inadequate information on three and late onset in two
Chakraborty	1962	One male	Absence of tics
Balducci and Frascella	1962	One (sex unspecified)	Inadequate information
Stevens and Blachley	1966	One male	Absence of vocal tics
Clark	1966	One female	Inadequate information
Connell *et al*	1967	Two males	Absence of vocal tics
Messiha *et al*	1971	One male	Absence of vocal tics

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