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Gilles De La Tourette syndrome : a review of the recent literature

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GILLES DE LA TOURETTE SYNDROME:

A REVIEW OF THE RECENT LITERATURE

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INTRODUCTION

It shall be the purpose of this paper to review the recent medical and psychological literature regarding the Gilles de la Tourette syndrome. This will be in essence a review of 17 new sources since the publication of a review of this syndrome by Kelman in 1965. (This review by Kelman includes 40 sources and was a summary of her dissertation in psychology.)

There are 17 new sources in this review, including 27 new cases, making a total of 71 reported cases in the medical literature.

GENERAL DESCRIPTION

According to Kelman there were 44 cases of Gilles de la Tourette's Syndrome reported in the literature from 1906 to 1964.[10] Since that article I have found 27 reported additional cases. However, this is by no means the true incidence as many cases go unreported. Many psychiatrists when asked have firsthand knowledge of a case or two.

Gilles de la Tourette's Disease presents as motor incoordination in the form of a tic. The incoordination most frequently begins in the face or upper limbs with the lower limbs affected later. Next there is a period in which the patient utters inarticulate sounds. Finally, and the pathognomonic part of the disease, is the echolalia and coprolalia associated with the motor incoordination. Included in the differential diagnoses are Wilson's Disease, Sydenham's Chorea, Encephalitis Lethargica and other neurological entities which at times may include tics and coprolalia.

According to Kelman the disease affects primarily males in the ratio of 3:1.[10] The usual age of onset is under ten years of age but there are some cases of onset in the thirties. In Kelman's review, she noted that about

half the cases had vocal outbursts without coprolalia. Kelman reported a mean IQ of 106 with a range of 89-142. There are no consistent EEG findings in any series.

ETIOLOGY

The exact etiology of Gilles de la Tourette's Disease is unknown. There are about as many theories as there are writers on the subject.

The behaviorists view the symptoms as being a learned response to the stimulus of a high autonomic drive. At the same time that the drive is reduced via the symptom formation the cortical arousal mechanisms are activated to a higher state of excitement. This level is too high to allow adequate motor and verbal control even if the patient wanted to stop the symptom manifestation.[3]

Chapel views the symptoms as a result of repressed anger toward one of the parents. Once the tic was learned the symptom could be used for secondary gain in interpersonal relations thus reinforcing the symptom. Though the tic was at first anxiety reducing, it often by its nature of coprolalia would lead to social isolation and an increase in anxiety leading to more ticing and thus becoming self-perpetuating.[2]

Chapel also made an interesting observation that the cases he was familiar with came from families in which there was a great deal of conflict. The patient seemed to be in the middle of the conflict. One of the parents was rigid, demanding and punitive while the other was overindulgent and overprotective. The marriage was usually failing, and one of the parents rejected the patient and used him as a scapegoat. This idea was cited in a similar manner by Faux when he stated that his patient's father was forced to choose between his son and his second wife. The patient was hospitalized in order to save the marriage.[7]

Another explanation of the symptomatology of Gilles de la Tourette's disease is that given by Downing. The symptoms are viewed as a form of behavior which allows the patient to "express rebelliousness, hostility and sexuality in the family setting in such a way as to avoid the demeaned self-concept seen in her sibs".[5] It was noted that this particular family tended toward rigid control and "correct behavior" thus producing ambivalence toward authority. The other siblings were noted to have strong elements of the obsessive compulsive, the hysteric and the schizophrenic. Downing postulated that if the patient's symptomatology

were completely removed a schizophrenic psychosis might occur. This is an interesting postulation in that many books and articles, particularly those earlier ones reviewed by Kelman, stated that the end fate of many Gilles de la Tourette patients was hospitalization for chronic schizophrenia.[10]

Kurland reported on two female patients, ages 19 and 15, who had punitive, rigid, overwhelming mothers and distant fathers. They both showed overwhelming ambivalence and inability to directly express hostility toward the mother. He viewed the coprolalia as a symptom formulated to punish the mother and gratify a sexual wish. The total syndrome is seen as "an exaggerated response to feelings of hostility and ambivalence concerning the maternal figure as well as to the growing and unmanageable sexual drives and urges of the adolescent".[11]

Robinson believes the syndrome to be due to a pregenital conversion representing an anal sadistic regression.[14]

In addition to these functional theories of symptomatology there are the organic theories. The best article on the EEG findings is the one by Field. In this review of seven cases he relates a familial history of mental disease in four families, a movement disorder in three, and

a history of seizures in one. Though the EEG findings are inconsistent with each other, Field states that the tracings are definitely different from those of patients with uncontested psychogenic movement disorders.[8]

In the Kelman review, EEG results on 19 patients were given with the findings that 11 were within normal limits. The other eight were, as Field found, inconsistent with each other but somewhat abnormal.

J. R. Stevens, in reviewing a case history of a thirteen year old girl and the effect on her behavior of haloperidol (Haldol), postulated that the syndrome is like certain other movement disorders and thus possibly due to a metabolic disturbance causing central nervous system enzyme or neurotransmitter dysfunction.[16]

In his case review of a graded response to haloperidol, McKinnon noted that his patient had EEG changes and described them: "Very frequent frontally preponderent polyspikes occur associated with myoclonic jerks. Delta waves at myoclonic paroxysms. The jerking paroxysms are accompanied by unequivocal evidence of centrocephalic spike discharges."[13]

As for autopsy findings, Field states that there have only been three autopsies on known cases and the only finding in these was an increase in the number of small cells in

the corpus striatum.[8] It has been suggested in the Kelman review (Brain, 1960) that this area might be involved in tic symptomatology.[10]

In addition to the possibility of the etiology being all organic or all functional, it has also been postulated that the syndrome is the result of the interaction of functional and organic factors. In view of the success of both psychotherapeutic and pharmacological therapies, this hypothesis certainly seems plausible.

TREATMENT

Kelman's article presented a tabular form of all the treatments used in her period of literature review dating from 1906-1964. The following is Table 2 on page 223 of her review.

TABLE 2

DETAILS OF TREATMENT AND OUTCOME

TREATMENT	OUTCOME			TOTAL
	Improvement	No change	Worse	
Psychotherapy	6	4	1	11
Isolation	-	1	2	3
hypnosis	1	3	-	4
told not to suppress it	1	-	1	2
narcoanalysis	1	2	1	4
insulin	-	1	-	1
CO_2	2	1	-	3
ECT	1	2	-	3
chlordiazepoxide and isocarboxazid	1	-	-	1
amphetamines	1	3	2	6
chlorpromazine	5	3	-	8
triflupromazine	2	-	-	2
Trifluperizine	2	-	-	2
thiordizine	1	-	-	1
haloperidol	5	-	-	5
TOTALS	29	20	7	56*

*Some patients had more than one form of treatment.

Only the major forms of treatment are included in the previous Kelman table. In addition "many cases had been given drugs of various sorts at one time or another, sometimes with a temporary remission, but in most cases having no effect at all. The drugs mentioned are tranquillizers--reserpine, meprobamate, barbiturates (unspecified) and bromides; anti-Parkinsonian drugs--procyclidine (kemadrin), trihexyphenidyl (artene), as well as muscle relaxants (unspecified) and mephenesin".[10]

Among the methods of treatment in the more recent literature is the behavioral treatment which aims to cut down on the incidence of ticing by deliberate practice of it.[3] This method was successful in two out of three cases in which Clark tried it. There were no new symptoms mentioned that developed with extinction of the tic.

In another behavioral approach, J. R. Stevens tried aversive conditioning in which the patient received "self-delivered, brief, mildly painful electric shocks each time she twitched, the tics disappeared, only to return with increased vigor as soon as the shock apparatus was detached from her person".[16]

It is interesting, in going over some of the early literature on tics, that the behaviorist approach was mentioned.

Fleichner in 1911 advocated that to treat a tic the child should be: (1) taken out of school and sent to the country; (2) given a diet that is easily assimilated; (3) given rest before and after meals; (4) given a light evening meal and sent to bed within two hours; (5) given sodium or stantium bromide for two weeks; and (6) given educational treatment aimed at concentrating attention or in consciously repeating the tic.[9]

There are two patients who have been treated with frontal leucotomies. The one reported on by H. Stevens was done transorbitally on July 28, 1955. It was performed on a thirty-seven year old man who developed symptoms at age eight. He is now controlled by 100 mg. daily of chlorpromazine.[15]

The other case was treated by a bimedial frontal leucotomy. This involved a twenty-two year old male who developed symptoms at age nine. This particular patient had severe feelings of panic for which the leucotomy was done hoping thereby to reduce the tic. This particular patient also had a history of a difficult birth after prolonged labor and grandmal seizures at ages three and five. His head was also noted to be deformed. He now has a minimal tic and it is controlled with dilantin and chlorpromazine.[1]

The usage of metharbital and hydroxyzine hydrochloride was only noted in one case and the patient appeared to get worse.[4] Dilantin was noted once before in this paper to have been used for treatment in conjunction with a leucotomy patient of Bakers.[1] It was also stated in Kelman to have been used twice before with no effect.[10] Diazepam was used once by Field and he obtained some remission of symptoms.[8] J. R. Stevens used dextroamphetamine to no avail.[16]

Chlorpromazine was used after both leucotomies with good control, but the only new case treated with this drug without surgery was one cited by J. R. Stevens in which there was no reported improvement.

Hypnosis was noted to be used on four cases reported by Erickson. I report on these only for the sake of completeness. Two of them had an onset in their mid thirties thus making me a little suspicious of the diagnosis. Both of these cases were reported as recovered though the causation was not determined. The other two cases he reports were in adolescents but, because of parental opposition, they were not able to complete therapy and are thus assumed not improved.[6] The other case of hypnosis being used was by J. R. Stevens who again reports no improvement with this form of treatment.[16]

One of the more successful methods of treatment is psychotherapy. Faux reported on a case treated with largely group therapy who has done well after release from the hospital. "The patient's most insightful and motivating experience seemed to occur in the variety of group experiences he had plus the fact that he became a meaningful part of a new therapeutic environment where he could flounder and come to grips with some basic problems in relating to people. Peer expectation, confrontation and approval devices plus a social matrix of high-level psychiatric aides and staff seemed the most important factors in his recovery."[7]

Kurland reports that his two cases were much improved by long term psychotherapy.[11] Lucas reported two cases to be improved while in a children's psychiatric treatment center but they were simultaneously receiving haloperidol. This improvement, therefore, probably cannot be attributed entirely to psychotherapy.[12] J. R. Stevens reports no success in treating his patient with psychotherapy.[16]

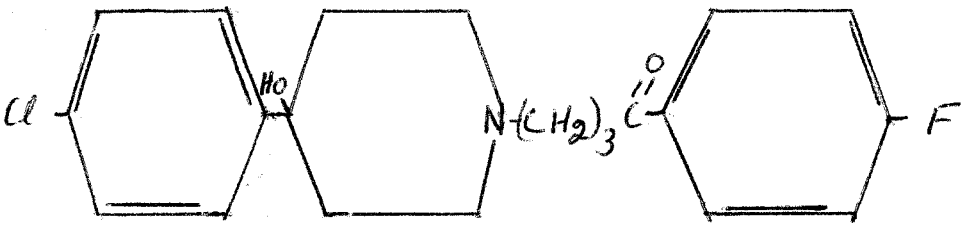
Probably the most important advance in treatment methods for Gilles de la Tourette syndrome is the introduction of haloperidol, a butyrophenone. Chapel states

that "treatment with butyrophenone is the only known method which has proven consistently effective in the control of the symptoms of this disorder".[2] J. R. Stevens cited a case of a thirteen year old girl refractory to all other methods of treatment. She was controlled at a dosage of 12 mg daily which totally abolished the tic while at the same time improving her general behavior and personality. He found that by reducing the dosage or substituting other agents the tic returned after several days. While on this dosage of 12 mg daily there were no neurodysleptic side-effects.[16]

Lucas reported that in two cases which were treated for over a year there appeared to be excellent response. He states: "The markedly palliative effect of haloperidol in these patients with tic syndrome is in accord with the results of other investigators and resembles our previous experience with phenothiazine drugs."[12]

Definite electroencephalographic changes were reported on a case of Gilles de la Tourette syndrome by McKinnon. He states: "He showed a response to haloperidol which was interesting for the apparently graded response of the motor and vocal tics to the dosage level of the drug. Its use made possible a marked improvement in the patients family and social adjustment."[13]

From the drug insert it appears that haloperidol is used in the treatment of acute and chronic psychoses as well as the Gilles de la Tourette syndrome. It is the first of a new series of major tranquilizers called the butyphenones. Its structure is as follows:



Stated in the drug insert is: "The safety of this drug in pregnancy has not been established.....In this connection it is to be noted that a case of phocomelia in an infant whose mother received haloperidol, along with a number of other medications, during the first trimester of pregnancy, has been reported (a causal relationship has not been established in this case)."

It should also be noted that extrapyramidal reactions occur with this drug but are usually reversible. These reactions have been reported to occur with relatively low doses though they are usually dose related. Liver, hematologic and psychophysiological reactions are noted to occur but have a relatively low incidence. The manufacturer also

notes that the drug should not be prescribed for children under twelve because safe conditions for use have not been established.

An attempt will be made to summarize my review in Table A in the same form as used by Kelman and then I will combine my results with hers for an overall tabulation of treatment and results in Table B.

TABLE A

TREATMENT AND OUTCOME

TREATMENT	OUTCOME			TOTAL
	Improvement	No Change	Worse	
Behavioral therapy	2	2	-	4
Chlorpromazine	2	1	-	3
Dextro-amphetamine	-	1	-	1
Diazepam	1	-	-	1
Dilantin	2	2*	-	4
Haloperidol	4	-	-	4
Hydroxizine Hydrochloride	-	-	1	1
Hypnosis	2	3	-	5
Lobotomy	2	-	-	2
Metharbital	-	-	1	1
Psychotherapy	5	1	-	6
TOTALS	20	10	2	32**

*This is from Kelman's review but not included in her table.

**The total exceeds the 27 patients involved due to the fact that many had more than one form of treatment. (In addition some of the 27 new cases are not included due to the fact that no mention of treatment was included in the original article.)

TABLE B

COMBINED TABLE

TREATMENT	OUTCOME			TOTAL
	Improvement	No change	Worse	
Amphetamines	1	3	2	6
Behavioral therapy	2	2	-	4
Chlordiszepoxide and isocarboxazid	1	-	-	1
Chlorpromazine	7	4	-	11
CO_2	2	1	-	3
Dextroamphetamine	-	1	-	1
Diazepam	1	-	-	1
Dilantin	2	2	-	4
ECT	1	2	-	3
Haloperidol	9	-	-	9
Hydroxizine hydrochloride	-	-	1	1
Hypnosis	3	6	-	9
Insulin	-	1	-	1
Isolation	-	1	2	3
Lobotomy	2	-	-	2
Metharbital	-	-	1	1
Narcoanalysis	1	2	1	4
Psychotherapy	11	5	1	17
Told "not to suppress it"	1	-	1	2
Trifluoperazine	2	-	-	2
Triflupromazine	2	-	-	2
Thioridazine	1	-	-	1
TOTALS	49	30	9	88

CONCLUSIONS

1. There are too few cases from which to draw statistical inferences about etiology, treatment, or prognosis.
2. From the foregoing material, it appears that the etiology of Gilles de la Tourette's Syndrome is unknown.
3. Clinically the best known drug therapy is haloperidol.
4. Psychotherapy (behavioral and dynamic) is an effective mode of treatment clinically.
5. Clinically the syndrome can be altered by drugs and psychotherapy and the outcome is not as bleak as originally described

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