Three Cases of Gilles de la Tourette s Syndrome. Treatment with Chlorimipramine: A Preliminary Report

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INTRODUCTION

After a description made by Gilles de la Tourette (1885), student of Charcot, of a syndrome characterized by compulsive uncoordinated movements with cefalo-caudal progression, symptoms of repetitions, echolalia, echopraxia, coprolalia, arithmomania, and vocal non-articulated noises, his name was given to this rare and yet not completely defined neuropsychiatric entity. Obsessions, compulsions, aggressive behavior and self mutilation were also described as complementary symptoms. The age of onset is usually about 10 years old (Fisarova); being 8 years old in 74 percent of their cases and no older than 13 for Anderson and Abuzzahab.

Based on the presence of the vast majority of these symptoms we made the diagnosis, although none of our cases present coprolalia, the most amazing symptom described by de la Tourette. The fact is that is not a pathognomonic one, as only a half progress to it at a late stage of the illness (Corbett) and all of our cases were early depicted, treated, and by the time of the presentation of this paper, not older than 16 years.

We report here three cases successfully treated with chlorimipramine.

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CASE 1: D.M.

Sex: Male.

Birthdate: May 1,1961.

First Consultation Date: July 1972. 11 years and 2 months old.

Symptomatology: Insidious, mild facial tics during childhood; at the consultation moment full symptomatology. Always having a tendency to facial tics, blinking, grimacing, during childhood. Progression to shoulder, arm and neck tics (jerking), thorax, legs (kicking), massive jerkings. Vocal symptoms: cough, throat clearing, stammering, sniffling. Echopathia (repetition of other people's tics). Obsessive-compulsive pattern: obsessive thoughts, rituals (undressing several times a day), urge to perform tics. Early childhood phobias: need of light for sleeping, nictophobia. Tendency to irritability and aggressiveness. Bad temper.

Parents ask for differential diagnosis with chorea.

Evolutive History:

Main Findings: Normal pregnancy. Premature birth: 8 months. Cianosis. Oxygen tent incubator. Artificial lactation. Cry baby, day and night. Phobic obsessive since early childhood (never uses other's glasses, etc), very clean and meticulous and punctual.

Sleeping Pattern:

Rare somniloquia. Psychomotor evolution and independence habits within normal limits.

Scholarship:

Resistance at the beginning. Very good pupil and good social integration.

Health Problem Antecedents: Fever convulsions: 8 months. Amigdalectomy: 4 years old. Measles. Mumps, no complications. Allergies. Hives.

Family Members' Relationship: Well integrated family, affective and communicative. Has an elder brother. Mother: tendency to be hyperprotective. Familial perfectionism.

Psychotraumatic Episodes: Amputation of his mother's leg. Marked aggravation of symptoms.

Hereditary Pattern: Tendency to perfectionism in both parents.

Psychological Evaluation: 11 years and 2 months old.

IQ: Wise-Verbal CI: 110 Execution CI: 92 Total CI: 101 Normal Average Range. Bender test: immaturity visomotor gestaltic function.

EEC: not significant.

Differential Diagnosis: Sydenham's chorea.

Treatment: Psychoanalytic psychotherapy and family orientation from September 1972 up to September 1975. Irregular evolution with mild incomplete remissions. June 1976: Chlorimipramine 0.0125 g three times daily. A week and a half later total remission of symptomatology (especially tics). It is called "the magic pill" by the family. Progressive diminution of doses in a six month treatment. No relapses up to date (March 1978).

CASE 2:A.O.

Sex: Male.

Birthdate: April 3,1962.

First Consultation Date: December, 1974.

Age of onset 11 years old. Facial tics started earlier.

Symptomatology: Facial tics, grimacing, eye blinking, neck jerks, inarticulated sounds: ouch, tsc, pft; lipsmacking, sucking, licking. Repeat own words (palilalia). Compulsions. Echomimics. Echopathy. Arithmomania, delire du touche. Echokinesis. Hyperkinesis. Irritability. Aggressiveness toward members of his family and other boys. Impulsivity. Compulsive joker. Poor adaption at school. Doesn't respond to discipline or follow directions. Compulsions to pinch, touch, bother school mates. Lability of attention, makes stupid questions. No friends, unpopular. Bad temper. Fires setter, cruelty with animals, destructive throwing and breaking toys and furniture. Overly dependent upon mother. Night terrors. Nictophobia. Claustrophobia. Acarbphobia. Difficulty in falling asleep. Hard to get up in the morning. Prone to accidents. Inconstancy. Ritualistic: says good-night as much as twenty times a night, and goes to urinate, pushes his bed close to the wall, etc. Obsessive thoughts: intrusion of "dirty" and obscene thoughts (internal coprolalia), sexual sadism content, magic interrelation of facts. Double checking. Urge to perform and thoughts of performing his compulsive acts. **Evolutive History:**

Main Findings: Pregnancy: 2 months hemorrhages coincident with the mother's father's death. Clinician's suggestion of abortion. The Galli-Mainini reaction death foetus response with the concomitant anxiety of the mother. Fear of deformities.

Birth: Normal. Cry baby, for hours over day and night without stopping. Nervousness. Artificial lactation. Good appetite. Rejects solid food. Teething ring up to 3 years old. Tendency to ambidextrism. Sphincter control - $3^{1}/2$ years.

Sleep Pattern: Continued sleeping difficulties, crying, teeth grinding, startling. Need to sleep with somebody. Hyper-dependency to be fed or dressed.

Psychomotor Evolution: within normal limits.

Scholarship: Social adaptation problems.

Health Problem Antecedents: Measles, mumps, chicken pox, scarlet fever, rubeola, anginas with high temperatures (all without complications).

Family Members' Relationship: Well integrated family. Hyperanxious mother. Two sisters.

Psychotraumatic Episodes: 1973: death of a loved aunt. Mystic questions.

Hereditary Pattern: Brother of his father, hypochondriac; cousin, schizophrenic.

Psychological Evaluation: IQ: above the normal range. E EG: December 1974. Bioelectric disorganization at basal state. Respiratory activation, generalized paroxysmal

activity of 2.3 cycles per seg.
Differential Diagnosis: Severe obsessive - compulsive neurosis.
Treatment: Psychoanalytic psychotherapy and family orientation since April 1975 up to June 1976. Supported with the following medications: N-Benzylchloropropionamide (secular). Hydantoinates. Phenobarbital. Vitamin dietary supplementation. June 1976: Started with 25 mg daily of chlorimipramine. Gradually increased to 75 mg daily...until March 1977, when symptomatic remission was completed. Up to date (March 1978) continue with this dosage, relapses occurring when he abandons treatment.

CASE 3: D.F.

Sex: Female.

Birthdate: May 1,1967.

First Consultation Date: March 1975.

Age of onset: 7 - 8 years old. **Symptomatology:** At 4 years old starts with abdominal pains that alleviate when a phonetic tic (mooing, cattle bellowing sound) is performed. December 1974: this tic diminishes and a head movement starts, rapidly (two months time) spreads to neck, shoulders, upper and lower limbs. A pediatrician is consulted for a chorea differential diagnosis. Other symptoms are explosive and impulsive behavior, aggressiveness, startle reactions, esophageal spasms, throat clearing and noises, inarticulated sounds, muscle pains. Seen by cardiologists (rheumatic fever discarded). Obsessive thoughts. Echopathy. Tendency to copy other people's tics. Phobias: nictophobia, abandonment (doesn't go alone to the bathroom), asks her parents to sleep with her. Compulsive eater (mainly sweets). Permanent affective insatisfaction. Episodic nocturnal enuresis. Poor social integration. Urge to perform and consciousness of performing tics.

Evolutive History:
Main Findings: Pregnancy normal. Birth induced. Buttock presentation. Normal baby. Lactation: maternal 15 days, artificial since then, good appetite. Tendency to am-bidextrism. Mild dullness in attaining her independence habits.

Sleep Pattern: Somniloquia, sudden sitting on bed. Sporadic jerks.

Psychomotor Evolution: within normal limits.
Scholarship: No learning difficulties.
Health Problem Antecedents: 2 years old: repetitions of convulsions. *VA* year old: head traumatism. Strabism develops. 4 years old: strabism operation. 8 years old: another head traumatism.
Family Members' Relationship: Well integrated family. Two elder sisters.
Psychotraumatic Episodes: 4 years old, strabism operation coincident with first tic manifestation.
Hereditary Pattern: Great grandfather: echopraxia. Aunt: obsessive compulsive behavior.
Psychological Evaluation: IQ: Raven test. Percentile 90, Range 11, above the normal range. Basic personality: obsessivecompul-sive. E E G: 8 months: dysrythmia (family reference). 1974: generalized bioelectrical disorganization.
Laboratory Findings: Cholesterol 245 (NV:150-240) Urinary 17 cetosteroids 1.90 mg/24 hs. Diuresis 870 ml/24 hs (NV: 6.0-11.80/24 hs). Method: Drebter mod. Urinary 17 hydroxicorticoids 3 mg (NV: 3-10/24 hs). Method: Porter and Silber. Plasmatic somathotrophin: 2.9mUL/ml (NV 0-20 mUL/ml).
Vainillin Mandelic acid: 5.7 mg/24hs. (NV: 2.57-7.0 mg/24 hs).
5 hydroxy indolacetic acid: 9.6 mg/24 hs. (NV: 5.15 mg/24 hs). 5 hydroxy triptamine: 0,23 m c g/De (U.N. 0,

5 hydroxy triptamine: 0,23 mcg/De (U.N. 0,15-0.35 mcg/De).
Differential Diagnosis: Hysteria. Chorea. Rheumatic fever.
Treatment: 8 months. Phenitoin (Epamin). March 1974: Carbamezapina (Tegretol). N-benzilcholorpropionamide(seclar). Glipharelax. Vitamin supplementation. December 1977: Chlorimipramine 0.0125 g three times daily. partial symptomatology remission.

February 1978: Chlorimipramine 25 mg three times daily. 15 days later very remarkable improvements up to date, March 1978.

The etiology of this syndrome remains obscure, and several authors tend to explain it according to their theoretical background.

Some give a Psychodynamic interpretation (Mahler, Hollander, MacDonald, Aarons, Ferenczi, etc.) while the observations of others (Yaryura-Tobias, Fisanova, Koprouska-Mileska, Stevens, Blachly, Lucas & Rodin, Seignot and Shapiro, Senler, etc.) give preponderance to organicity. For a third group the cause remains unknown or may be a combination of both (Fernando Milman).

We are inclined, by the observation of our modest series, to support the organic background as mainly causative, being psycho-traumatic episodes precipitant factors. Arguments in favour of our position are:

- Age of onsetaverage 10 years old.
- The presence of perinatal pathology (3 cases).
- Cry baby and irritability (2 cases).
- E E G pathological (2 cases) and presence of convulsions (1 case).
- Tendency to ambidextrism (2 cases).
- Pathological sleep pattern (3 cases).
- Worsened under stress situations or psychotraumatic episodes (3 cases).
- Well integrated families (3 cases).
- Failure of psychotherapy as corrective treatment (3 cases).
- Success of psychopharmacological treatment.

It is of special interest for us to point out the obsessive-compulsive feature of this syndrome. (Outlined in Gilles de la Tourette's description). Classically included as psycho-dynamic in origin, the obsessive-compulsive disorders.

Where recently presented as having an organic etiology at the biochemical level, being a serotonergic disturbance the causative factor (Yaryura-Tobias) mentioned by this author, Evarts demonstrated that motor and thalamic regions discharge prior to movements, giving an explanation to the fact that all of our patients are conscious of their tics, and have urge to perform them in at least 50 percent of their occurrences.

Phobias are usually the early infant manifestation of a future obsessive-compulsive disorder. This was certain for our three cases, thus phobias and facial tics tendency in a young child may be signaled as a very premature sign of a possible Gilles de la Tourette Syndrome development.

Following this new approach, we tried Chlorimipramine, a serotonine uptake inhibitor, obtaining in our cases excellent results with a remarkable symptomatology remission in all of them. No side effects were referred by patients at the utilized doses.

Although spontaneous remissions are frequent, we find a cause-effect relation need to reach the correct doses (cases 2 and 3), relapses when medication is interrupted (case 2), total remission of symptoms in ten days at 14 years old (case 1), while spontaneous remissions tend to occur in late adolescence.

We will continue to follow up these patients and time will have the last word about those questions where further work is necessary. Meanwhile, we suggest Chlorimipramine as a first line attack on Gilles de la Tourette's Syndrome, with or without psychotherapy as a complementation.

We have no experience with Haloperidol signaled by many authors as the elective treatment for this illness (Seignot, Senler, Shapiro, etc.).

SUMMARY

Three cases of Gilles de la Tourette's Syndrome are presented. The premature phobic and consequent later obsessive-compulsive development are pointed out as an important aspect of this entity.

Excellent therapeutic results with Chlorimipramine (a serotonine uptake blocker) are reported.

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