Neurological features of Gilles de la Tourette’s syndrome

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SUMMARY Clinical neurological examinations of 22 patients with Gilles de la Tourette’s syndrome and written reports of examinations of seven other patients are reported. Half the personally examined patients had minor motor asymmetries in addition to the typical motor and vocal tics found in all the patients. Thirty-six per cent of patients were left handed or ambidextrous. Electroencephalograms performed on 17 of the 22 patients showed non-specific abnormalities in 12 of them. These findings suggest that a neurological disorder underlies Tourette’s syndrome, but they do not clarify its nature.

Tics may be defined as brief purposeless movements of a body part which occur at random intervals and cannot be suppressed for long periods of time. Wilson (1927) believed they were psychogenic in origin, while others have emphasized developmental (Balthasar, 1957) or inflammatory disorders (Creak and Guttmann, 1935) of the basal ganglia as possible causes.

Gilles de la Tourette (1885) described nine patients with persistent multiple tics beginning in childhood. Five of these patients also had coprolalia. Tourette linked tics with other rare motor and speech disorders, such as latah, myriachit, and ‘jumping Frenchmen’, and emphasized the chronic stable course such patients followed in adulthood.

Subsequent authors have differentiated ‘maladie des tics’ from other involuntary motor disorders (Kelman, 1965; Chapel, 1970). Both psychiatric and neurological disorders have been invoked as causes of Tourette’s syndrome without conclusive evidence (Kelman, 1965; Challas, Chapel, and Jenkins, 1967; Corbett, Mathews, and Cornell, 1969; Morphew and Sim, 1969).

Tourette’s syndrome responds best to treatment with haloperidol (Chapel et al., 1964; Shapiro and Shapiro, 1968), a butyrophenone compound which blocks dopamine receptors (Yeh, McNay, and Goldberg, 1969). It has been suggested that a dopamine excess may underlie Tourette’s syndrome (Snyder, Taylor, Coyle, and Meyerhoff, 1970), but the actual anatomical or pathophysiological abnormality which might cause Tourette’s syndrome and be controlled by haloperidol is still unknown.

Clinical neurological techniques have been used by several authors in an effort to demonstrate a neurological deficit. Fernando (1967) reviewed the world’s literature and noted no specific findings, but found reports of abnormal electroencephalograms (EEG) in 25%. Corbin, Goldstein, and Klass (1966) reviewed seven patients at the Mayo Clinic and again found no pertinent changes on examination but found six patients with rolandic sharp waves on EEG, four while awake and two while asleep. Lucas (1970) described the EEGs of 12 patients, of which seven were abnormal. Five showed diffuse mild to moderate slowing, one had posterior delta activity, and one showed bilateral spike and wave complexes.

Since subtle neurological abnormalities and clinical correlations may escape notice in a few patients, or in a group of patients examined by various physicians, we thought it worthwhile to report the findings in a large series of patients with Tourette’s syndrome.
### Table 1
**Summary of 22 Tourette Patients Examined**

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (yr) at: exam onset</th>
<th>Sex</th>
<th>Handedness*</th>
<th>Prior illness</th>
<th>Family history</th>
<th>Medication</th>
<th>Initial tic</th>
<th>Tics observed</th>
<th>Vocal tic†</th>
<th>Other motor findings</th>
<th>EEG</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>8 5 M R</td>
<td></td>
<td></td>
<td></td>
<td>Unknown adopted St. Vitus' dance—maternal aunt and grandmother</td>
<td>—</td>
<td>Blink</td>
<td>—</td>
<td>Grunts, hisses (coprolalia)</td>
<td>Hyper-active</td>
<td>Not done</td>
</tr>
<tr>
<td>2</td>
<td>8 5 M L</td>
<td></td>
<td></td>
<td></td>
<td>—</td>
<td>Blink</td>
<td>Shoulders and thighs; fist to chest</td>
<td>Grunts, coughs (words)</td>
<td>Hyper-active</td>
<td>Background disorganization and bilateral occipital sharp waves</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>8 4 M A</td>
<td></td>
<td></td>
<td>Induced labour</td>
<td></td>
<td>Haloperidol</td>
<td>Arm</td>
<td>Thigh flexion; mild bilateral intention tremor; shoulder shrugs; chorea of hands</td>
<td>Grunts, words</td>
<td>Hyper-active</td>
<td>Bilateral occipital sharp waves</td>
</tr>
<tr>
<td>4</td>
<td>10 4 F R</td>
<td></td>
<td></td>
<td></td>
<td>—</td>
<td>Head bob</td>
<td>—</td>
<td>—</td>
<td>Grunts</td>
<td>Background disorganization and bilateral sharp waves and spikes posteriorly</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>11 10 M R</td>
<td></td>
<td></td>
<td></td>
<td>—</td>
<td>Head</td>
<td>Rotary head movement; milkmaid's grip</td>
<td>Clicks</td>
<td>† Background disorganization and bilateral sharp waves</td>
<td></td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>14 9 M L</td>
<td></td>
<td></td>
<td></td>
<td>—</td>
<td>Face</td>
<td>Face; pectorals, quadriceps; milkmaid's grip</td>
<td>Grunts</td>
<td>Left parieto-temporo-occipital slow waves and occasional left posterior sharp waves</td>
<td></td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>14 3 M R</td>
<td></td>
<td></td>
<td>Clumsy child; mother pre-eclamptic</td>
<td>—</td>
<td>Blink</td>
<td>—</td>
<td>Grinace; negative head shake; shoulders, thighs; mild chorea</td>
<td>Grunts, squeals, coughs, spitting</td>
<td>‡ Mild slowing and background disorganization</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>14 3 M R</td>
<td></td>
<td></td>
<td></td>
<td>Haloperidol</td>
<td>Right hand</td>
<td>Blinks; grimace</td>
<td>—</td>
<td>Grunts; sniffs</td>
<td>Background disorganization with bilateral slow and sharp waves</td>
<td></td>
</tr>
</tbody>
</table>

*Handedness is indicated as L (left) or R (right).
<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>Sex</th>
<th>Diagnosis</th>
<th>Associated Findings</th>
<th>Medications</th>
<th>Symptoms</th>
<th>Additional Observations</th>
</tr>
</thead>
<tbody>
<tr>
<td>9</td>
<td>15</td>
<td>M</td>
<td>Mother pre-eclamptic</td>
<td>—</td>
<td>—</td>
<td>Blink</td>
<td>Shoulders and thighs; chin to right; athetosis of arms</td>
</tr>
<tr>
<td>10</td>
<td>15</td>
<td>M</td>
<td>Mononucleosis</td>
<td>—</td>
<td>Haloperidol</td>
<td>Bark and head turn</td>
<td>Cough, bark (coprolalia)</td>
</tr>
<tr>
<td>11</td>
<td>16</td>
<td>M</td>
<td>Suicide and depression—maternal uncles and mother</td>
<td>haloperidol (stopped 3 wk before examination)</td>
<td>Face</td>
<td>R. shoulder and arm elevation. Torticollis; inversion of feet</td>
<td>Spitting and coughs (coprolalia, echolalia)</td>
</tr>
<tr>
<td>12</td>
<td>19</td>
<td>M</td>
<td>Insulin shock and prochlorperazine</td>
<td>Facial tics—mother</td>
<td>—</td>
<td>Head movements, face</td>
<td>Shrugs; flexion of arms into belly</td>
</tr>
<tr>
<td>13</td>
<td>20</td>
<td>M</td>
<td>Clumsy child</td>
<td>—</td>
<td>Haloperidol</td>
<td>Blink</td>
<td>Face, shoulders, head, ballism</td>
</tr>
<tr>
<td>14</td>
<td>20</td>
<td>M</td>
<td>Oxygen and blood after birth</td>
<td>—</td>
<td>Haloperidol</td>
<td>Face</td>
<td>Shoulders and right hand</td>
</tr>
<tr>
<td>15</td>
<td>26</td>
<td>F</td>
<td>Bilateral cryothalamotomy</td>
<td>—</td>
<td>Haloperidol</td>
<td>Blink</td>
<td>Squints Mild choreoathetosis, thumb tremor</td>
</tr>
<tr>
<td>16</td>
<td>26</td>
<td>F</td>
<td>Bilateral cryothalamotomy</td>
<td>—</td>
<td>Amantadine</td>
<td>Face</td>
<td>Athetosis on R.</td>
</tr>
<tr>
<td>17</td>
<td>30</td>
<td>F</td>
<td>Haloperidol</td>
<td>—</td>
<td>Arm</td>
<td>Shouts</td>
<td>Shoulders and neck; ballism, echopraxia</td>
</tr>
<tr>
<td>18</td>
<td>31</td>
<td>F</td>
<td>Haloperidol</td>
<td>—</td>
<td>Shrug</td>
<td>Shouts</td>
<td>echolalia (coprolalia)</td>
</tr>
<tr>
<td>19</td>
<td>34</td>
<td>M</td>
<td>'Sleeping sickness', trans-sexualism</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>Smacks and spitting</td>
</tr>
<tr>
<td>20</td>
<td>41</td>
<td>M</td>
<td>Hiatal hernia</td>
<td>Writhing of hands—son</td>
<td>—</td>
<td>Blink</td>
<td>Face</td>
</tr>
<tr>
<td>21</td>
<td>55</td>
<td>M</td>
<td>Hiatal hernia</td>
<td>Glutethimide propantheline</td>
<td>—</td>
<td>Blink</td>
<td>Jumps, windmill sweeps of arms</td>
</tr>
<tr>
<td>22</td>
<td>62</td>
<td>M</td>
<td>Cerebral arthitis</td>
<td>Epilepsy-grandfather</td>
<td>—</td>
<td>Blink</td>
<td>Shoulder elevations; chorea; postural lapses</td>
</tr>
</tbody>
</table>

A = Ambidextrous. † Words in parentheses imply 'history of'. ‡ See Table 2.
METHODS

Twenty-two patients from a series of 34 patients with Gilles de la Tourette’s syndrome treated at the Payne Whitney Clinic of the New York Hospital–Cornell Medical Center were examined neurologically. In addition, written reports of neurological examinations of seven other patients in the series were reviewed. At the time of examination, 10 patients were taking haloperidol, 2–8 mg/day and two were taking benztropine. One other patient had taken haloperidol and benztropine for three years, but was examined after three weeks without medication. A complete medical history was taken with emphasis on early life injury or illness, developmental abnormalities, intercurrent illnesses, and medication. This information was obtained from patients and their relatives and supplemented by previous medical records. A complete neurological examination with special emphasis on motor phenomena was performed, along with a general medical examination. Electroencephalograms were performed on 17 of the 22 patients using unipolar and bipolar montages with 22 scalp leads. All patients were hyperventilated during the recording. Five had both awake and sleep EEGs and four had photic stimulation during EEG recording. Electromyographic tracings from surface electrodes over muscles involved in tics were obtained concurrently with the EEG in one patient. Paper speed was increased from 3 to 6 cm/sec during EEG–EMG recording to facilitate detection of the start of activity.

HISTORY

Data for all examined patients are summarized in Table 1.

Gestation and birth were normal in all but three of the personally examined patients. One man had required oxygen therapy and blood transfusion after birth, presumably because of neonatal blood incompatibility. Two boys’ mothers had been preclamptic while pregnant with them, but their birth and perinatal periods were normal. Labour, lasting 10 hours, was induced in a third boy’s primiparous mother at full gestation with no complication. Birth history was abnormal in three other patients among the total of 34, with two instances of prolonged labour and one of pneumonitis. One reviewed patient is a dizygotic twin whose sibling is not affected by Tourette’s syndrome. Birth weight of examined patients averaged 3.3 kg (7.2 lb.) and ranged from 2.2 to 5.0 kg (4.8 to 11.1 lb.). Maternal age at birth averaged 27.8 years (19–36). Twelve of the 22 examined patients were first children, seven were second children, and three were third or lower in order. Comparable figures for the 34 patients are 17, 11, and six.

Developmental milestones were reached normally by all examined patients under the age of 18 years. Four older patients reported delayed sitting, walking, or talking; the reliability of their retrospective histories is questionable. Another three patients among the series of 34 had delayed milestones. Six patients among the 34 were late in toilet training and two of the boys who were examined were said to have been ‘clumsy’ in early childhood.

The patients were examined at an average age of 23 years (range 8–62). Ten of the 22 examined patients and six of the seven reviewed cases were aged 15 years or less. Seventeen of the 22 examined and six of the seven reviewed patients were males, a predominance also found in other series (Fernando, 1967).

Symptoms began at an average age of 7 years in both examined and reviewed cases. Initial symptoms in examined patients consisted of frequent inappropriate blinking or squinting in 10 patients, facial tics in four, arm and shoulder movements in four, head bobbing in three, and barking in one. Progression of tics from face to proximal limb muscles and the appearance of vocal exclamations within a few years were typical.

Concomitant illnesses noted at the time of examination were cervical spondylosis, hiatal hernia, and transexualism. A woman had undergone bilateral cryothalamotomies with partial relief of Tourette’s symptoms (Cooper, 1963), and a boy had received insulin shock treatment and prochlorphazine for tics during his teens. One boy had infectious mononucleosis three months before the onset of his tics and a man gave an anecdotal history of ‘sleeping sickness’ at 11 months of age.

Handedness was inquired about routinely in the interview. Five of the 22 personally examined patients were strongly left-handed and three others considered themselves ambidextrous. Four others among the series of 34 patients were left-handed, giving a total of 12 patients among 34 (35%) with Gilles de la Tourette’s syndrome who were ambidextrous or left-handed.

Family history disclosed two parents of patients who had occasional facial tics, usually during periods of stress. A grandfather was said to have had ‘epilepsy’, not further characterized. A patient’s son developed writhing hand movements lacking the characteristics of a tic at age 20 years. Two maternal uncles of a patient committed suicide and his mother was periodically depressed. ‘St. Vitus’s dance’, evidently consisting of chronic isolated chorea, led to year long hospitalization of a patient’s maternal grandmother at the age of 16 years and his maternal aunt at the age of 9 years. Neither woman had either a permanent movement disorder or valvular heart
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Disease. The absence of tics in a twin of one patient is mentioned above.

Twenty-four of the 34 patients had at least partial Ashkenazi Jewish ancestry. Antecedents of five others were Italian, three Anglo-Saxon, and one each German and East European.

NEUROLOGICAL EXAMINATION Aside from tic phenomena, the standard neurological examination showed no striking changes. Cranial nerve functions were entirely normal. The two oldest patients, 55 and 62 years old, had mild distal symmetrical lower extremity sensory loss.

Mild motor asymmetries were noticeable in 11 of the patients who were personally examined (Table 2). In 10 patients these were unilateral impairment of rapid alternating movements, pronation or drift of an outstretched extremity, increase in tone or reflexes or decreased associative movements on one side, or decrease in tone and check on one side with ataxia. These asymmetries occurred in patients whose

was found to have a left Babinski sign and mild clumsiness on the left. Some of his tics were atypical because of a dystonic component, though profuse coprolalia was documented. He developed episodes of 'rushing thoughts', paranoia, adhesive head movements to either side, and frequent tics after three years of haloperidol therapy. During an episode observed in the hospital the patient was lethargic but oriented and capable of simple calculations. His vital signs, postural responses, and blood sugar were normal. Such episodes usually lasted hours, though a short aura of 'far away feeling' sometimes occurred without the episode itself. Skull radiography and brain scan were normal. The EEG between episodes showed bilateral frontotemporal sharp waves. A pneumoencephalogram was normal, though the CSF protein was 67 mg/100 ml. The exact nature of this boy's episodes is unclear and he is being followed without specific treatment for them.

Two boys, ages 8 and 14 years, exhibited short attention span, clumsiness, and hyperactivity consistent with what has been termed 'minimal cerebral dysfunction'.

ABNORMAL MOVEMENTS Abnormal movements found in Tourette's patients were generally proximal and rapid (Table 3). Facial grimaces occurred in 12

examined and five reviewed patients. They were usually associated with brief eye closure and shoulder elevation. Abrupt rotation of the head in one direction was seen in four examined and two reviewed patients. Another examined patient had rapid to and fro rotations of head on neck resembling a negative head shake. Forward head thrusts were seen in two examined and one reviewed patient.

### Table 2

<table>
<thead>
<tr>
<th>Patient</th>
<th>Abnormality</th>
<th>Handedness</th>
<th>Medication</th>
</tr>
</thead>
<tbody>
<tr>
<td>5</td>
<td>Decreased tone and check in left UE, Ataxia</td>
<td>R</td>
<td>None</td>
</tr>
<tr>
<td>6</td>
<td>Impaired alternating movements and increased DTRs in right UE</td>
<td>R</td>
<td>None</td>
</tr>
<tr>
<td>7</td>
<td>Impaired alternating and associative movements in right UE</td>
<td>A</td>
<td>Haloperidol, benztropine</td>
</tr>
<tr>
<td>8</td>
<td>Decreased right arm swing</td>
<td>R</td>
<td>Haloperidol</td>
</tr>
<tr>
<td>9</td>
<td>Left Babinski sign</td>
<td>R</td>
<td>Haloperidol</td>
</tr>
<tr>
<td>10</td>
<td>Left sided weakness and incoordination</td>
<td>A</td>
<td>Prior insulin shock and prochlorperazine</td>
</tr>
<tr>
<td>12</td>
<td>Decreased right arm swing</td>
<td>R</td>
<td>Haloperidol</td>
</tr>
<tr>
<td>13</td>
<td>Right UE pronation and impaired alternating movements</td>
<td>L</td>
<td>Haloperidol, benztropine</td>
</tr>
<tr>
<td>17</td>
<td>Left UE pronation and drift</td>
<td>R</td>
<td>None</td>
</tr>
<tr>
<td>19</td>
<td>Increased tone and decreased arm swing on right</td>
<td>L</td>
<td>Glutethemide, propantheline</td>
</tr>
<tr>
<td>20</td>
<td>Impaired alternating movements of left UE</td>
<td>R</td>
<td>None</td>
</tr>
</tbody>
</table>

A = ambidextrous.

### Table 3

<table>
<thead>
<tr>
<th>Type of tic</th>
<th>Patients (no.)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Proximal</td>
<td></td>
</tr>
<tr>
<td>Facial</td>
<td>12</td>
</tr>
<tr>
<td>Head shaking or thrusts</td>
<td>7</td>
</tr>
<tr>
<td>Shoulder</td>
<td>13</td>
</tr>
<tr>
<td>Thigh</td>
<td>3</td>
</tr>
<tr>
<td>Jumps</td>
<td>1</td>
</tr>
<tr>
<td>Distal</td>
<td></td>
</tr>
<tr>
<td>Hands or feet</td>
<td>2</td>
</tr>
<tr>
<td>Others</td>
<td></td>
</tr>
<tr>
<td>Choreiform movements</td>
<td>7</td>
</tr>
<tr>
<td>Athetosis</td>
<td>2</td>
</tr>
<tr>
<td>Dystonia</td>
<td>1</td>
</tr>
</tbody>
</table>

Age averaged 26 years and ranged from 11 to 62 years. Four of the patients with motor asymmetries were taking haloperidol and one had received insulin shock therapy in the past. Unilateral motor changes were not related to the site of tics.

One other patient, a 16 year old boy (case 11),
Shoulder elevation or rapid 'shrugging' movements were present in 13 examined and two reviewed cases. These movements might be unilateral or bilateral and extended to ballistic abduction of one or both upper extremities in three patients. Brief flexion-adduction movements of both thighs were seen in three examined patients and sudden irregular jumps with windmill sweeps of the arms in one other.

Distal motor abnormalities were not as marked or as common as proximal ones. Isolated tics of hands or feet were seen in two examined and one reviewed patients. 'Wiping movements' of the upper extremities and 'toe curling' were described in two reviewed patients. Distal choreiform movements, usually noted only if specifically sought, were found in seven examined and two reviewed patients. In addition, athetosis was found in the right arm of the patient who had had bilateral cryothalamotomies and in both arms of a 15 year old boy taking haloperidol and benztprine. Dystonic inversion of either foot and brief torticollis were seen in the 16 year old boy described in detail above. Torsion movements of the trunk were seen in a motion picture of this patient taken at age 13 before haloperidol had been started.

Sudden patterned movements were also noted in a few patients and they seemed to be related to postural changes. One man briefly lapsed into a flexed posture during sustained muscular effort. A boy repeatedly bent forward and flexed his arms into his epigastrium and another boy suddenly struck his chest with closed fist of his right hand at irregular intervals. One reviewed patient was said to lapse into a 'knee bend' posture. None of these patients was taking haloperidol or other medication at the time such patterned movements were observed.

Haloperidol may have caused tremors that were observed in two of the examined patients. A 26 year old woman taking haloperidol, 4 mg/day, and benztprine had intermittent 2-4/sec resting tremor in her right thumb in addition to impaired rapid movements and drift of the outstretched right arm. Mild bilateral intention tremor was found in an 8 year old boy taking haloperidol, 5 mg/day. However, a 62 year old man who was taking no medications also had left upper extremity sustention and intention tremor in addition to impaired rapid movements. Three reviewed patients, aged 10, 15, and 38 years and taking no medication, were said to have tremor of outstretched hands.

VOCAL PHENOMENA Vocal phenomena were found in all patients. Initially, grunts, coughs, or 'barks' were usually associated with tics. After a period of time, usually several years, formed words became part of the vocal tic in 13 examined patients. Coprolalia supervened in 10 patients. Echolalia, the involuntary repetition of words uttered by others, was noted in seven patients. Coprolalia was recorded in two reviewed patients and one of these also had echolalia.

Aside from coprolalia and echolalia, vocal utterances sometimes took fascinating forms. Two patients exclaimed 'ow! wow! ow!'. Others interspersed hisses, spitting, and coughs. One boy inappropriately accented words in mid-sentence.

An abnormal respiratory pattern was noted in a 62 year old man with tics and shouts since the age of 6 years. Periodic tachypnoea and apnoea with rapid transition from one to the other and an overall frequency of seven per minute occurred while he was awake or drowsy. Tics usually began on the expiration of a deep breath. During sleep, regular respirations at 20-22/min were seen. Arterial blood gases, drawn with the patient awake, showed a mild respiratory acidosis and 100% oxygen saturation.

GENERAL EXAMINATION The general medical examination disclosed no consistent abnormalities. Two patients had soft non-radiating apical systolic cardiac murmurs, another a pectus excavatum deformity, and another dependent pedal oedema.

ELECTROENCEPHALOGRAM Seventeen patients who had neurological examinations also had an electroencephalogram. Twelve of these EEGs were abnormal (Table 4). Nine tracings showed paroxysmal features consisting of bilateral sharp waves in eight, and left posterior sharp waves associated with left parietal-temporal-occipital slow waves in one other. The bilateral sharp waves were posterior in four tracings, diffuse in three, and frontotemporal in one. One of these patients had simultaneous EMG monitoring of tics during his EEG and the timing of his occipital sharp waves was independent of motor activity. Sharp waves in EEG's from the other patients also seemed to be independent of tics, though EMG confirmation was not obtained. Four of these patients were taking haloperidol, 4 mg/day, and one who had undergone bilateral cryosurgery was also taking amantadine, 300 mg/day. One other patient had bilateral sharp waves associated with tic movements.

The remaining abnormalities consisted of diffuse background disorganization and slowing in awake tracings of two patients, one of whom was taking haloperidol 4 mg/day and benztprine. Five of the
tracings with paroxysmal activity also had background disorganization.

The provocative techniques of hyperventilation, sleep, and photic stimulation did not change the EEG patterns significantly.

**DISCUSSION**

The examination of these 22 patients with Gilles de la Tourette’s syndrome has disclosed three factors of possible importance in understanding the cause of this condition. These are a high prevalence of left-handedness, minor motor asymmetries, and electroencephalographic abnormalities.

A high prevalence of left-handedness (eight of 22 patients examined, or 36%, were left handed or ambidextrous) might be interpreted as evidence of left cerebral damage which might in turn be due to the underlying abnormality in Tourette’s syndrome. Though Hécaen and de Ajuriaguerra (1964) quote the frequency of left-handedness as 1 to 30% in various series, they cite the most likely frequency as between 5 and 10%. Other factors to be considered are the predominance of males both among left-handed people and Tourette’s patients and the tendency of children to acquire firm hand preference only after the first decade of life. Despite these reservations, the frequency of left-handedness in this series is high enough to warrant speculation on the relationship of early life brain injury to Tourette’s syndrome.

The finding of minor motor asymmetries in half the patients examined carefully may also indicate such a relationship. Similar findings have been associated with the syndrome of minimal cerebral dysfunction which some investigators link to birth trauma (Paine, 1962; Prechtl and Stemmer, 1962; Anderson, 1963; Twitchell, LeCours, Rudel, and Teuber, 1966). Also three of the six patients with right-sided motor abnormalities were left-handed or ambidextrous. However, the somatic asymmetry which often accompanies early life hemispherical damage was not found in our patients. Also, the motor findings were not seen in the youngest patients. Moreover, lack of a control population for examination meant that determination of motor abnormality depended solely on the neurologist’s clinical judgement. Most important, the slight arm drift, pronation, increased tone, and clumsiness found in those patients were extremely subtle in comparison with their obvious tics.

Electroencephalographic abnormalities were found in 12 of the 17 Tourette patients who also had neurological examinations. This confirms the findings of other series (Ungher, Ciurea, and Volanschi, 1962; Field et al., 1966; Fernando, 1967; Lucas, 1970), although our patients showed posterior abnormalities more often than the rolandic or parietal abnormalities described by others. No specific or characteristic electroencephalographic pattern was noted in patients with the syndrome.

The frequency of non-specific EEG abnormalities in ‘normal’ populations has ranged from 5-6% to 32% (Secunda and Finley, 1942; Corbin and Bickford, 1955, Hill, 1963), figures far exceeded by those of our patients (12 of 17 or 70%). Clinically inapparent tics could be responsible for the sharp wave abnormalities noted and we are undertaking a study with needle EMG electrodes and modified time constants to investigate this point. The background disorganization found in seven of 17 records (41%) cannot be attributed to tics. Haloperidol has been found to increase either slow and sharp or fast activity in the scalp electroencephalogram (Itil, Gannon, Hsu, and Klingenberg, 1970) and six of the patients with abnormal tracings were taking this medication. However, these patients’ abnormalities did not fall into any one category and two other patients taking haloperidol had normal records. Lucas (1970) found that haloperidol did not change his patient’s EEGs significantly.

Taken together, the high frequencies of left-handedness, minor motor findings, and electro-
encephalographic abnormalities suggest organic abnormality of the central nervous system as a concomitant of Tourette's syndrome. This suggestion is further strengthened by the results of psychological tests of patients with Tourette's syndrome. Slight to moderate indices of organic dysfunction were present in 23 of 30 patients (77%) given WAIS or WISC, Bender Gestalt, and Rorschach tests (Shapiro, Shapiro, and Clarkin, in preparation).

The nature of the neurological pathophysiology of Tourette's syndrome, if it exists, has not been ascertained by this study. Sudden patterned movements of the face, trunk, or proximal extremities suggest hyperactivity of central motor mechanisms. Whether this is due to primary irritability or removal of inhibition is unknown. Dopaminergic hyperactivity in the corpus striatum has been postulated as the pharmacological mechanism for Tourette's syndrome (Snyder et al., 1970; Messiha, Knopp, Vanecko, O'Brien, and Corson, 1971) because of haloperidol's suppression of tic symptoms and blockade of dopaminergic receptors, but this hypothesis has not yet been substantiated (Di Giacomo, Fahn, Glass, and Westlake, 1971). Vocal tics, especially coprolalia, have been cited as evidence for a psychological cause of Tourette's syndrome (Morphew and Sim, 1969), but forced utterances and respiratory tics are well described following von Economo's encephalitis (Turner and Critchley, 1925; Van Bogaert, 1934; Wohlfahrt, Ingvar, and Hellberg, 1961). Only two of our patients had any historical suggestion of encephalitis (mononucleosis and 'sleeping sickness') and none showed ocular or extrapyramidal signs which could not be attributed to medication. Also, no patient had signs or symptoms of rheumatic heart disease, which would suggest Sydenham's chorea. (Creak and Guttmann, 1935; Aron et al., 1965). Thus, there is no evidence for an infective or post-infectious cause of Tourette's syndrome, though a subclinical or slow viral infection may have been overlooked.

The consistent age and manner of onset of Tourette's syndrome in our patients could be ascribed to an inborn constitutional abnormality. The progression of tics from periorcular to facial to proximal limbs to trunk is reminiscent of the course of heredofamilial tremor (Larson and Sjögren, 1960). A patient with multiple tics and acanthocytosis without serum lipid abnormality has been described (Critchley, Betts, Nicholson, and Weatherall, 1970) but no other metabolic abnormality has been associated with tics. The family histories of our patients revealed several interesting motor disorders ("St. Vitus's dance", writhing hand movements) but no consistent pattern.

Because of the non-fatal course followed by patients with tic syndromes, there are only two necropsy reports of such patients in the literature. One was normal (DeWulf and Van Bogaert, 1941) and the other reported subtle cellular changes in the corpus striatum ("cynthohyperplethora") attributed to developmental arrest (Balthasar, 1957). Neither showed changes which one could associate with early life cerebral injury or infection.

Though examination of our 22 patients with Tourette's syndrome has disclosed subtle abnormalities suggesting a neurological disorder, we doubt that similar studies will yield more significant information. Further clarification of the pathophysiology of Tourette's syndrome, and of related movement disorders, will require more sophisticated investigative techniques, especially investigation of the pharmacology of putative neurotransmitters in patients with this perversive illness of Gilles de la Tourette.

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Neurological features of Gilles de la Tourette's syndrome


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