American Handbook of Psychiatry

MOTOR SYNDROMES OF FUNCTIONAL OR UNDETERMINED ORIGIN

Tics, Cramps, Gilles de la Tourette's Disease, and Others

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e-Book 2016 International Psychotherapy Institute

From *American Handbook of Psychiatry: Volume 3* Silvano Arieti

Table of Contents

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Eduard Ascher

Motor disturbances have always been of special interest to the clinician, because they can be readily observed and serve as indicators of the status of an underlying illness. In addition, their origin is frequently obscure, as psychogenic motor syndromes often are undistinguishable from those of an organic nature. They may affect almost every part of the body and vary in severity from mild twitchings of a single muscle group to extensive involvements of nearly every striated muscle of the body. Since the affliction of certain muscle groups is of special clinical significance, they will be singled out in this discussion, while keeping it general with respect to all other motor disturbances.

Tics

Tics are seemingly purposeless, repetitive, short-lasting, sudden contractions of striated muscles. They are involuntary and may affect but a few bundles. In severe cases, almost every part of the body is involved. The

age incidence is highest among children and most commonly affected are the face and neck.

The origin of the term was attributed by Brissaud to popular use and considered onomatopoetic by Meige and Feindel. The sound of the word conveying meaning is supported by the fact that several other languages use similar terms (*ziehen, zucken,* and *ticken* in German, tug and tick in English, *tico* in Spanish, *ticchio* in Italian).

The classification of tics depends on location and the extent of the functional determinant. “Habit tics” are those of primarily psychogenic nature and “psychic tics” of solely functional origin. Attitude tics are longer-lasting habit spasms which resemble catatonic symptoms.

Classification according to location is employed by Olson who divides children s tics into five categories: face and neck tics, those of arms and hands, shoulder and gait muscles, alimentary and respiratory, and a miscellaneous group that includes echolalia and coprolalia.

While the controversy of psychic versus organic origin continues unabated, there have been few clinicians who have failed to recognize that the functional component is ever present in tics and should be given serious consideration.

Tics are generally classified to be a hysterical conversion reaction or a compulsive disorder. Hysterical tics are similar to mannerisms and twitchings of normal individuals, according to Cameron, who considered them remnants of an attempt to adjust to anxiety while expressing through their sign language repressed conflicts of current and past significance.

According to Fenichel, tics are hysterical conversion phenomena when a well-circumscribed area of the body is affected and the patient's ego is intact. He calls the more complex tics conversion reactions also, but of a non-hysterical quality. When there is evidence of regression in such patients, he refers to them as compulsive and resembling catatonic states.

According to Bockner, tics differ from acts of a compulsive nature by the fact that they are involuntary, poorly organized, and only partially influenced by conscious effort of control. Purves-Stewart and Worster-Drought point out that tics occur automatically without the subject's attention drawn to them, while compulsive acts are very much in the awareness of the individual who cannot resist performing them.

Psychopathology

Because of the uncertain origin of tics, theories to explain them are diverse and numerous. Meige and Feindel saw in them evidence of congenital, organic, and psychological deficits, while stressing the importance of studying

each case individually. They felt that they were compulsive, inopportune, defensive, and automatic in nature.

Friedreich believed tics to have been purposive, coordinated acts which became involuntary and repetitious. Muncie calls them a motor neurosis resulting from “rut formation.” After a period of “legitimate” reaction to discomfort, the movements lose their original meaning and purposiveness by becoming habitual.

Wilder and Silbermann failing to cure tiqueurs by psychotherapy were reluctant to attribute them to psychic causes, while psychoanalysts found in them much to support their libido theory. Ferenczi believing them to be similar to catatonic stereotypes and the result of libidinal displacement named tics “cataclonia.” Deutsch, Reich, and Aarons viewed tics as masturbatory equivalents, the latter also expressing the belief that the twitchings were symbolic of a need to ward off the danger of submitting to passive wishes.

Torticollis

Because torticollis is of relatively frequent occurrence, it is described here separately. It is also known as “wryneck” and is a tic involving the neck muscles, especially the sternocleidomastoid on one or both sides. The tic is complex and consists of quick elevations of the chin while the head is turned

to one side as a result of spasmodic contractions of the neck musculature. When the contractions are quick and brief, torticollis is classified as a tic, when protracted and tonic it is considered a cramp.

While tics in general affect children more frequently, psychogenic torticollis is more common among adults. Patterson warns that torticollis in a child must be considered organic, unless proven to the contrary.

The psychodynamics of torticollis is often indicative of expressing the emotion of aversion, while Fenichel sees the condition as resulting from anal retentive tendencies, suppressed rage, and castration anxiety.

Cramps

A cramp is defined as a protracted, tonic muscular contraction of functional origin. It affects a well-circumscribed area of the body, often an extremity. Classified as being of a hysterical nature, it is related to muscular tremors in general.

Cramps are of particular interest in certain occupational groups, because they frequently interfere with a person's ability to carry out his occupational duties, especially when his livelihood is dependent on his work performance. A common form is cramp of the writing hand in individuals who may be able to use the hand for every purpose other than writing. The

condition is thought to be the result of ambivalent feeling the patient has towards his occupation; however, it may also be overdetermined and indicative of sexual conflicts that are close to conscious awareness. The sexual nature of the problem is more apparent in *blepharospasm,* which may be an expression of a defense mechanism to prevent voyeuristic impulses from becoming conscious.

Other occupation groups affected are musicians, especially pianists and violinists, seamstresses, typists, and watchmakers.

The relationship of cramps and catatonic symptoms has been the subject of much discussion. Meige described a condition which he called "tic variables" or "attitude tics." This patient shows prolonged spastic contractions of body areas and the condition may progress to catatonia.

Treatment of Tics and Cramps

The nature of the treatment will depend on the degree of impairment and what one hopes to accomplish. Many simple tics are transient and need no special care, other than rest. Other tics are very complex and require intensive psychotherapeutic intervention, often with disappointing results.

Methods of treatment may therefore be divided in those aimed at symptomatic relief and others bringing about changes in attitude and

personality structure.

Symptomatic Therapy. Includes the use of rest, massage, heat, autogenous training, progressive muscular relaxation, hydrotherapy, surgery (torticollis), counterirritants, suggestion, hypnotherapy, and relaxant drugs (meprobamate, Valium, Robaxin). Some clinicians have used bed rest with isolation; others found re-education methods of some help. Yates, Rafi, and Clark used behavior modification techniques to reduce the frequency and severity of tics. They reported good results by building up the negative habit of not performing the tic, Rafi using an apparatus based on the conditioned response principle.

The symptomatic treatment of tics has received a big boost through the introduction of the drug, haloperidol, which has proven to be highly effective in the control of tics.

The evaluation of therapeutic results is complicated by the fact that tics have a tendency to fluctuate in severity or even disappear spontaneously on occasions. It may be said, however, that the prognosis is directly related to the mental state of the tiqueur and is favorable if there is little mental deterioration. Others factors, such as chronicity and extent of muscular involvement, are also of significance as illustrated by such conditions as the variable chorea of Brissaud and Gilles de la Tourette's disease, which are

described below.

Because of the ready availability of motor symptoms for study and observation, their use for evaluation of therapeutic results has been advocated, and as body language they are considered a valid indicator of unconscious processes since they emerge and are expressed in the psychoanalytic procedure.

Variable Chorea of Brissaud

In 1899, Brissaud described a condition that resembled Sydenham's chorea in its motor movements, but differed from it through its polymorphous manifestations; the choreiform movements lack uniformity, show no regularity in their evolution, and no constancy in their duration. "They come and go, increase and decrease, stop suddenly, recur suddenly, appear to be quick at one time and slow at another."

Meige and Feindel consider the syndrome to be a separate clinical entity, but this view is opposed to that of Gilles de la Tourette, who believed the condition to be a form of the *maladie des tics.* The prognosis of the syndrome is said to be grave, even though there may be prolonged periods of remission.

Gilles de la Tourette's Disease

History and Clinical Manifestations

First described by Itard in 1825, a clinical syndrome of multiple tics was reported by Gilles de la Tourette who presented case studies of nine patients suffering from a motor disorder which he defined as "nervous affliction characterized by motor incoordination accompanied by echolalia and coprolalia." The publications of his case reports aroused considerable interest at the time and various motor disturbances were thought to be evidence of this syndrome. The "latah" of Malaya, the "myriachit" of Siberia, and the "Maine jumpers" showed sufficient similarity in their behavior to consider them victims of this motor syndrome.

Anthropological studies by Aberle have since shown that latah differs from it in certain basic characteristics. *Schlaftrunkenheit,* a condition prevalent in Germany, was also attributed to this condition which had become known as Gilles de la Tourette's disease, because of the author's original reports of his clinical observations.

The syndrome is also mentioned as *maladie des tics, koordinierte Erinnerungskraempfe*, *myospasia impulsiva, maladie des tics convulsifs, maladie des tics impulsifs, maladie des tics dégenerés,* and *mimischer Krampf.*

According to Gilles de la Tourette, the first symptoms occur in childhood and show no sex preference. Before the age of ten, the child develops tics of

the face and extremities which vary in severity and are undistinguishable from other forms of tics or basal ganglia disease. The symptom which is characteristic of the disease is the emergence of a vocal tic. It may sound like a barking noise at first, which is inarticulate. As the condition progresses, words may be formed either of a coprolalic quality or designed to censure the obscene utterances. Such expressions as “keep quiet,” “shut up,” “don’t say it,” are not uncommon. On occasions, echolalia may be present, although this is not as frequent as Gilles de la Tourette thought it to be. The same applies to coprolalia, which he considered pathognomonic of the disease, but which other authors discount as a requirement to establish the diagnosis of Gilles de la Tourette’s disease.

The course of the disease differs from patient to patient. Gilles de la Tourette believed that the condition invariably leads to mental deterioration. More recent reports indicate that there may be general improvement in symptoms during late adolescence and that “spontaneous” remissions are not uncommon. With the advent of effective methods of control of the symptoms, there is increasing evidence that mental deterioration is the exception rather than the rule in the outcome of such cases

Etiology

The cause of the disorder is unknown. Gilles de la Tourette believed it to

be a hereditary disease, as did Koester and Oppenheim. Because of the similarity to symptoms of chorea, organic lesions of the brain, especially of the basal ganglion, were held responsible for the syndrome.' Autopsy reports have not been conclusive, but they have been sparse and incomplete. EEG tracings indicate that there is abnormality in about 50 percent of the cases, but there is no characteristic pattern for the disease. Concurrent with "organic" theories were psychogenic ones, such as by Guinon who saw little difference between Gilles de la Tourette's disease and hysteria. Other psychogenic theories were advanced by Mahler, Ascher, and others. On the other hand, Wilder and Silbermann expressed the belief that the resistance of patients with Gilles de la Tourette's disease to psychotherapy was proof that it was an organic disease.

Recent Developments

After Gilles de la Tourette's publication, there were only occasional case reports in the literature, until Kovacs described the analysis of a case and Wilder and Silbermann discussed the condition in their monograph on tics. They were the first to systematically treat such patients by psychotherapy, followed by Wilson and Mahler and Rangell who used psychoanalysis in the therapy of several children.

In 1948, Ascher reported his cases and reviewed the literature. It was

followed by an increasing number of case reports which indicated that the condition was not as rare as was formerly believed. The controversy of organic versus psychogenic continued in most publications; however, there emerged a tendency to consider all possible factors in the management of patients suffering from Gilles de la Tourette's disease.

Ascher investigated psychodynamic factors and concluded that many of the symptoms could be understood in terms of the patient's relationship to authority figures. He considered coprolalia as a dissociative phenomenon, as had Meige and Feindel previously, by which the patient diverts his aggressive impulses toward the authority figure to the world in general.

Mahler and Rangell also considered conflict between repressive force and instinctual process as significant, although they could not accept psychodynamic factors alone to be responsible for the condition. They believed that such patients have a constitutionally inferior subcortical structure with a reduction in their ability to withstand "overwhelming emotional and psychodynamic forces."

Other dynamic factors were introduced by Dunlap whose patient had parents with severe tics, and by Aarons who, like Fenichel, considered the muscular twitchings to be masturbatory equivalents.

In recent years, the interest in Gilles de la Tourette's disease has

increased and there are more case reports appearing in the literature. This is partly due to the nature of the disorder, but to a greater extent, to the effectiveness of psychotropic drugs in the control of its symptoms. Investigators have polled their information in panel discussions at national conventions, and an international registry has been established in an attempt to record all cases diagnosed as Gilles de la Tourette's disease.

The effectiveness of one psychotropic drug, haloperidol, has introduced a new factor in explaining the etiology of the syndrome. Because it is a potent blocker of dopamine receptors, it is suggested that hyperactivity of dopamine systems may be a factor in the pathophysiology of Gilles de la Tourette's disease.

Titration of severity of symptoms has been achieved in one case by altering the disposition of the brain catecholamine, norepinephrine.

Treatment

The variety of treatment methods employed is an indication of the confusion as regards the etiology of the disorder. Gilles de la Tourette treated his patients with sedatives, tonics, hydrotherapy, and isolation, the latter producing the best results, though temporary. Psychotherapy has been used with discouraging results, but there are reports indicating that psychotherapy was more successful. Hypnotic therapy and electro-convulsive treatment

have been ineffective; however, leucotomy, carbon dioxide inhalation, and sleep therapy are reported as having been helpful in controlling symptoms. The question is unanswered whether the improvement was the result of the treatment or represents a spontaneous remission.

The results of drug administrations have not been consistent. Bockner and MacDonald used chlorpromazine, which reduced anxiety, but it has to be given in large amounts to be effective. Amphetamine helped one patient, but this is in sharp conflict with the experience of Meyerhoff and Snyder. Seignot" used haloperidol and his success with the drug was subsequently confirmed by Challas and Brauer. Lucas and Chapel maintained their patients on daily doses of 1.5 mg. and 3.2 mg., respectively. There are a number of other reports describing the effective use of haloperidol. Clark successfully treated three patients by behavior therapy.

Although the focus of treatment has been on the patient and his symptoms, there is an increasing awareness of the importance of helping family members as well to understand the nature of the disease. This led to a publication of a guide for parents.

The upsurge in interest in Gilles de la Tourette's disease invariably will result in better diagnosis and earlier treatment. This offers great promise that fewer patients will suffer the psychotic deterioration Gilles de la Tourette

prognosticated.

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