TICS AND RELATED DISORDERS

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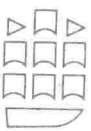

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Foreword

Early in my neurological apprenticeship I chanced upon a copy of the treatise on *Tics* by Meige and Feindel. I read and re-read the volume and my interest was all the greater because it had been translated into English by one of my chiefs. These miscellaneous movement-disorders have continued to intrigue me.

From my student days and from my residency at Great Ormond Street I was, of course, all too familiar with the diverse habit-spasms of the nervous child. But this was only the tip of an iceberg. I now became increasingly aware of the manifold mannerisms that most of my seniors seemed to show, ranging from a slow and solemn closure of one eye to a veritable saraband of sniffs, snorts and grimacing. A visiting professor of great eminence startled us all by his Jack-in-the-box display. No one among my elders seemed to be immune — academics, actors, barristers, politicians, clerics — and a subtle correlation with showmanship could be detected. Medical students were quick to seize upon the quirks, oddities and twists of their teachers and to utilize them mercilessly during the Christmas theatricals.

Then there were those instances of spasmodic torticollis that baffled us so often and so much. Did they represent symbolic gestures of aversion and abhorrence, psychologically determined? Many psychiatric colleagues were of that opinion, but none would ever embark upon their treatment. Could it be that torticollis was organic in nature and perhaps capable of alleviation by such mechanical measures as sectioning of either the muscles involved or their nerve-supply? A growing minority shared this suspicion but could not suggest any pathology other than 'deep-seated mischief'; that is, until the advent of epidemic encephalitis lethargica. The sequelae of that disorder were grave and protean, and many were in the nature of tics including torticollis. As to their intractability, James Collier used to proclaim that all muscle-attachments in the neck might be destroyed and every cervical nerve-root be severed, but the jerkings would continue, presumably through the musculature of the oesophagus.

In my experience, the only surgeon to have had any hint of success with torticollic sufferers has been Irving Cooper with his repeated and punctilious stereotactic approaches.

Craft palsies were another allied problem that worried us in those days. Their aetiology was obscure and their management perplexing. Here again the

whole subject was fraught with debates as to psychogenesis versus organicity.

Nowadays, and in some regions, it seems to be the practice to assemble all these clinical bits and pieces as though they were components of a neurological jigsaw, to be spoken of as the Tourette syndrome. Is this line of thinking justified?

Dr Lees has had the courage to grasp the nettle. He has embarked upon the exciting but hazardous task of studying in depth each representative of non-volitional movement and to discuss whether a common denominator exists, permeating the clinical aberrations: a formidable, but rewarding, assignment.

Here then is a monograph of distinction which takes up the story where the pens of Meige and of Feindel were laid down. Dr Lees has given us a survey of contemporary ideas in the context of *Bewegungserkentniss*. The contributions of science — chemical, pharmacological, electrophysiological — have added much to our stockpile of information about these enigmatical disorders. Whether a corresponding advance in our understanding of their fundamental nature has resulted is another thing.

Throughout his writing, Dr Lees has been scrupulous in defining each particular movement-disorder before proceeding to a most detailed clinical description.

May I wish this volume not only an appreciative reception, but even acclaim? Its future, I am confident, is assured.

1985 M.C.

Preface

Kinnier Wilson's translation of Meige and Feindel's monograph *Les Tics et leur Traitement* in 1907 proved to have a profound and lasting influence on the Anglo-Saxon concept of tic. In his subsequent lectures and writings Wilson reaffirmed the French School's view that tics were volitional cortical events occurring in emotionally immature and neurotic individuals. The growth of psychoanalytical theory in the early part of this century strengthened this view and, as a consequence, a whole score of fanciful theories based on case studies appeared in the literature. This alienated many psychiatrists and neurologists, and tics were discounted as unworthy of serious attention, being explained away to patients and their relatives as 'bad habits'.

The pioneering work of the Shapiros in New York in the 1960s and the resounding success of the American Tourette Association have drawn the medical profession's attention to the plight of patients with Gilles de la Tourette syndrome and have stimulated a resurgence of interest in tic disorders in the United States emulating that which occurred in Paris at the turn of the century. A new generation of neuropsychiatrists has adopted tic as its *cause célèbre*, perceiving it as the ideal paradigm for studying the interrelationship between emotions and motor behaviour. The occurrence of iatrogenic dyskinesias and tics occurring in association with structural neurological diseases has also stimulated the interest of neurologists.

The rapidly expanding literature is now widely dispersed within psychiatric, psychoanalytical and neurological journals and although there are two comprehensive multi-author textbooks devoted solely to Gilles de la Tourette syndrome, no up-to-date reference work covers the whole subject of tics and the other heterogeneous complex movements with which they are so frequently confused. In this book, I have attempted to draw together the most important strands of what remains a largely phenomenological literature in the hope of providing a modern successor to Meige and Feindel's monograph. My approach has been eclectic as befits a topic with so few established facts and I have attempted to integrate my own work on the subject impartially into the text.

I have encompassed most of the abnormal movements which currently possess no distinctive neurochemical or histological pathology. The abnormal movements seen in the psychotic, mentally retarded and the blind are covered

as they are so frequently misdiagnosed as tics, and the adult onset focal dystonias and hyperekplexias are included on historical grounds. I have arbitrarily excluded the myoclonic syndromes, generalised idiopathic torsion dystonia, paroxysmal dyskinesias, neuroleptic-induced rabbit syndrome, hereditary chin quivering and hemifacial spasm. Although drug-induced tics are rare, I have reviewed them in some detail and also covered the constellation of other iatrogenic abnormal involuntary movements. A greater understanding of these unwanted side-effects may well provide further insight into the pathogenesis of spontaneously occurring dyskinesias and tic.

Meige and Feindel hoped that their book would allot to the word tic a definite position in medical terminology. My own aspirations are equally modest and if, as a result of this book, progress can be made in the precise classification and definition of these bewildering disorders I will rest content. However, I also hope that the book may serve as a primer and reference source for those who might be tempted to work on this neglected and challenging group of movement disorders.

1985 A.J.L.

Acknowledgements

I am grateful to Mrs Joan Wolfe, President of the Gilles de la Tourette Syndrome Association of the United Kingdom, and to my collaborators Dr Mary Robertson and Dr Michael Trimble without whose help the data for the United Kingdom study on Gilles de la Tourette syndrome could not have been obtained. Dr Gerald Stern, my colleague and mentor at University College Hospital, has provided inspiration and well-timed nudges of encouragement. Dr Lieh Mak of Hong Kong and Dr Nomura of Japan were good enough to amplify their published studies for inclusion and Dr John Rothwell generously supplied the neurophysiological material depicted in Figures 1.1 to 1.3. I am especially indebted to Miss Elaine Edwards for painstakingly typing endless false starts and final drafts and to my father for proof-reading. The emotional support and forbearance provided by Juana my wife, and George and Nathalie my children, have been crucial in seeing the book through to completion.

Contents

1

Definition and Classification of Tic Disorders

The implausible nature of many of the abnormal movement disorders, their intimate connection with emotional upset, and the dearth of associated structural pathology have hampered rational classification. Semantic controversies persist despite a spate of well-meaning workshops and the recent publication by the Research Group on Extrapyramidal Disorders of an internationally agreed nomenclature (Lakke, 1981). In some respects understanding has advanced little since Charcot, exactly one hundred years ago, expressed the inherent difficulties as follows:

> Epilepsy, chorea, hysteria . . . come to us like so many sphinxes . . . symptomatic combinations deprived of anatomical substratum do not present themselves to the mind of the physician with that appearance of solidity, of objectivity, of affections connected with an appreciable organic lesion.
>
> There are even some who see in some of these affections only an assemblage of odd incoherent phenomena inaccessible to analysis, and which had better perhaps be banished to the category of the unknown.

Early 19th-century attempts at devising a working terminology depended on careful bedside observations and led to a steady proliferation of colourful eponyms such as Dubini's electric chorea, paramyoclonus multiplex of Friedreich and the variable chorea of Brissaud. Very few of these have stood the test of time as distinct nosological entities and most remain now only as monuments to the shortcomings of written descriptions in the field of abnormal movement disorders. Predictably this era of over-enthusiastic 'splitting' was followed by a sceptical volte-face in which all rapid involuntary jerks were lumped once more under a single rubric; this time myoclonus instead of chorea.

The last 20 years have witnessed modest progress in certain areas. Detection of specific enzyme defects, the discovery of physiological and neurotransmitter abnormalities and a greater understanding of underlying modes of inheritance have led to improved classifications of the choreas, dystonias and myoclonic syndromes. The capture of ephemeral dyskinesias on video-film and the greater use of audio-visual aids at medical meetings have also helped to iron out some of the prevailing dialectical arguments.

Tics, on the other hand, remain as *terra incognita*, sitting uneasily within the uncharted borderlands of neurology and psychiatry, the term being often

Table 1.1. CLASSIFICATION OF TICS AND RELATED CONDITIONS

I Idiopathic tics
- (a) Acute transient
- (b) Persistent simple or multiple
- (c) Chronic simple or multiple

II Gilles de la Tourette syndrome

III Tics occurring in association with structural brain damage
- (a) Post-encephalitic
- (b) Carbon monoxide poisoning
- (c) Head injury
- (d) Post-stroke
- (e) Post-rheumatic chorea

IV Drug-induced
- (a) Psychomotor stimulants
- (b) L-dopa
- (c) Neuroleptics

Related disorders
1. The hyperekplexias
2. Habitual manipulations of the body
3. Stereotypies
4. Mannerisms
5. Hyperkinetic syndrome
6. Adult onset focal dystonias
7. Clonic spasms

employed as a receptacle for all miscellaneous dyskinesias. They remain no more than a phenomenological concept and any attempt at tentative categorisation must insist on rigid definition and unvarying terminology. The system used in this book (see Table 1.1) aims to avoid questionable distinctions between functional and organic tics. However, its subdivisions are arbitrary and inevitably artificial. Gilles de la Tourette syndrome, for example, is distinguished merely by virtue of its severity. The tonic tics of Meige and Feindel are reclassified in the light of current thinking as focal dystonias, and tic douloureux and hemifacial spasm are considered as clonic spasms. Habitual manipulations of the body, such as thumb sucking and nail biting, are delineated from tics as are the legion of mannerisms and stereotypies so commonly encountered in mental asylums.

Definitions *A tic* is an abrupt, jerky, repetitive movement which involves discrete muscle groups. It mimics a normal co-ordinated movement, varies in intensity and lacks rhythmicity. It may be temporarily suppressed by will power and is relatively easy to imitate. It consists of a brief contraction of the prime

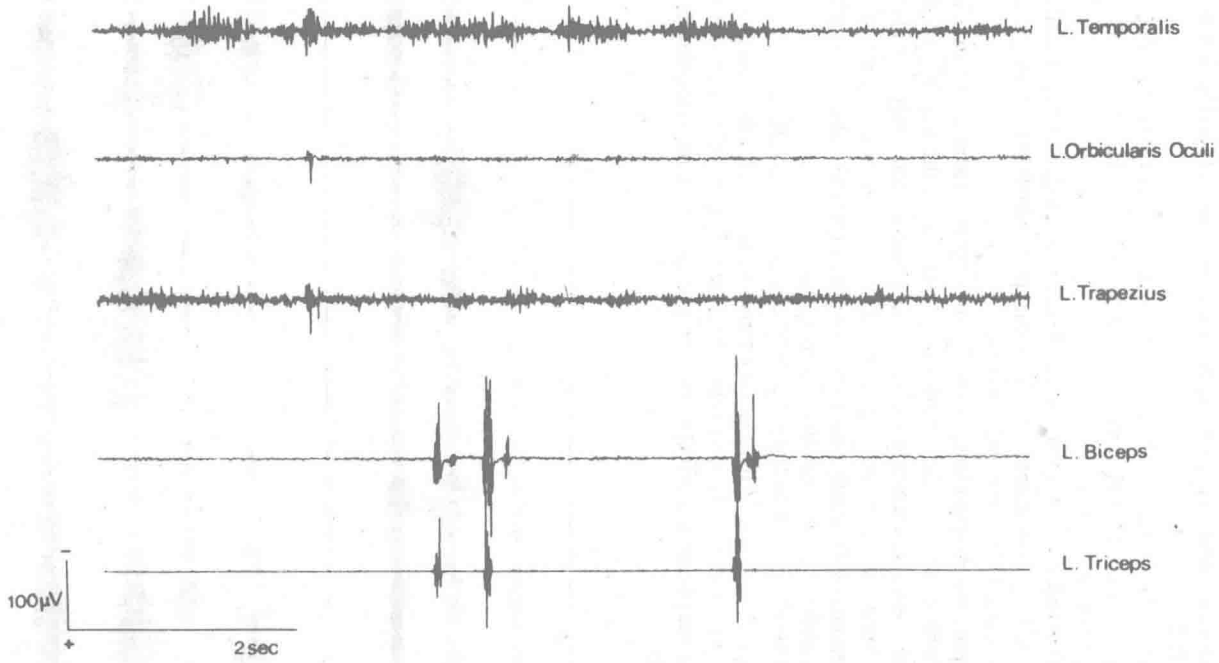

Figure 1.1 Surface electromyographic record of a 22-year-old man with simple tics in the left arm. (By kind permission of Dr John Rothwell.)

mover or simultaneous co-activation of agonist and antagonist lasting 50–500 ms (Fig. 1.1).

A mannerism is a bizarre mode of carrying out a purposeful act which usually occurs as a result of the incorporation of a stereotyped action into goal-directed behaviour.

A gesture is a culturally determined expressive movement calculated to indicate a particular state of mind and which may also be used as a means of adding emphasis to oratory.

Habitual manipulations of the body. These are self-gratifying socially offensive co-ordinated movements which occur particularly at times of anxiety, boredom, tiredness or self-consciousness.

Stereotypies are purposeless voluntary movements carried out in a uniform repetitive fashion often for long periods of time and at the expense of all other activities. In contradistinction to tics, whole areas of the body are involved.

Hyperactivity syndrome is characterised by an abnormal degree of aimless motor restlessness sufficiently severe to disturb attention and concentration and impede the ability to perform structured tasks.

Hyperekplexia is a pathologically exaggerated startle reflex.

Clonic spasms are rapid involuntary movements which follow strict anatomical localisation and can be accurately imitated only by electrical stimulation of the nerve supplying the affected muscle group. These movements may occur during sleep.

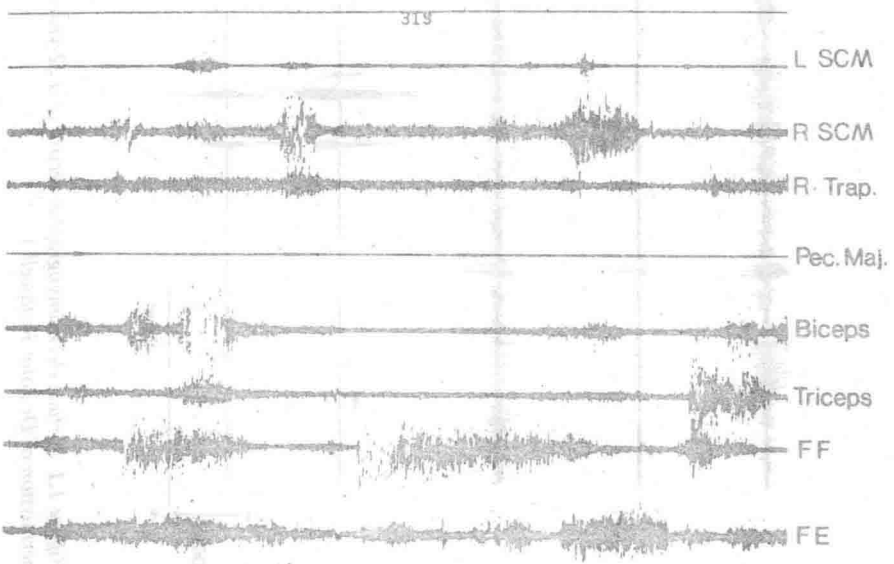

Figure 1.2 *Surface electromyographic record of an 8-year-old boy with benign hereditary chorea showing the random pattern of muscle activity. Time marker 1 s. (By kind permission of Dr John Rothwell.)*

Chorea is a forcible, rapid, irregular, involuntary and unpatterned jerk which is never integrated into a co-ordinated act but may match it in complexity. It is fleeting, unpredictable and presents a comical, playful appearance. It also tends to be aggravated by voluntary movement. Electrophysiologically it is characterised by its unpredictability. The burst of muscle activity may be brief, sustained or interrupted lasting 200 ms–1 s or more (Fig. 1.2).

Myoclonus is an irregular or rhythmical muscle jerk which originates in the central nervous system. The muscle contraction is brief resembling electric shocks and cannot be controlled by will power. The burst of muscle activity closely resembles that seen in tics.

Dystonia is a sustained involuntary torsion which may affect muscle groups of all sizes. The movements are usually slow and may be present continually or only appear during specific motor acts. When severe a relatively fixed abnormal posture may occur and secondary contractures develop. Co-contraction of agonist and antagonist occurs with a long duration of muscle activity lasting 1 s or more (Fig. 1.3). Dystonic spasm is a term applied to more rapid, repetitive, tic-like movements which lack the flowing character of chorea.

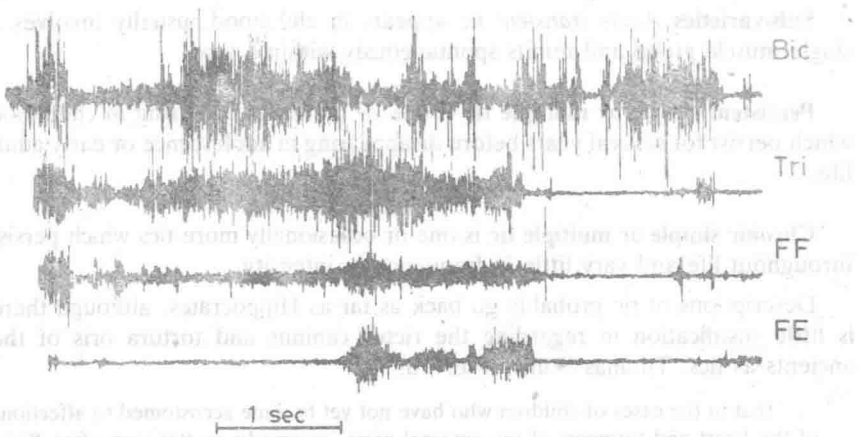

Figure 1.3. Surface electromyographic record of a 40-year-old man with dystonia of the right arm showing long co-contracting bursts of muscle activity in antagonist muscles. (By kind permission of Dr John Rothwell.)

2

Idiopathic Tic

Synonyms Habit spasm, habit chorea, variable chorea, pseudo-chorea.

Definition *A tic* is an abrupt, jerky repetitive movement which involves discrete muscle groups. It mimics a normal co-ordinated movement, varies in intensity and lacks rhythmicity. It may be temporarily suppressed by will power and because of its patterned appearance is relatively easy to imitate.

Sub-varieties *Acute transient tic* appears in childhood, usually involves a single muscle group and remits spontaneously within a year.

Persistent simple or multiple tic is one or more tics occurring in childhood which persist for several years before disappearing in adolescence or early adult life.

Chronic simple or multiple tic is one or occasionally more tics which persist throughout life and vary little in frequency or intensity.

Descriptions of tic probably go back as far as Hippocrates, although there is little justification in regarding the rictus caninus and tortura oris of the ancients as tics. Thomas Willis observed:

> . . . that in the cases of children who have not yet become accustomed to affections of the heart and torments of the external parts, spasmodic matter very often flows into the closest nerves, those of the third, fifth and sixth pairs. As a result, the area of the face and mouth is particularly distorted in these cases.

The word *ticque* was in current use in France as early as the 17th century to describe the behavioural vices of horses and in 18th century literature it was used in the sense of a 'recurrent distasteful act' particularly common in eccentric individuals. The derivation of the word is probably onomatopoeic, communicating the idea of sudden repetition as in tick-tack. *Tucken, ticken* and *tick* in German, *ticque* in French, tug or tick in English, *ticchio* in Italian and *tico* in Spanish are all probably derived from the same etymological root. André was the first to allude to tic douloureux of the face in 1756 but until the end of the 19th century dyskinesias were generally referred to as motor incoordinations and diagnosed as chorea. The first major medical work was that of

Bouteille (1810) who, in his *Traité de chorée*, distinguished a group of movement disorders he termed pseudo-chorea and which he defined as follows:

> I use the name pseudo-chorea or false chorea to distinguish those different, nervous, spasmodic, convulsive or hysterical afflictions which fail to show the characteristic features of true chorea, the only resemblance between them and chorea being facial grimacing and involuntary movements.

Sir Charles Bell used the term spasmodic twitching to describe tics in 1830, and in 1852 Marshall Hall clearly described the clonic nature of muscular tics giving examples such as violent frowning, facial distortion and shaking of the head. Trousseau (1873), however, provided the first clear description of tics although at that time he considered them a sort of incomplete chorea related to the occupational neuroses:

> Non-dolorous tic consists of abrupt momentary muscular contractions more or less limited as a general rule, involving preferably the face, but affecting also neck, trunk and limbs. Their exhibition is a matter of everyday experience. In one case it may be a blinking of the eyelids, a spasmodic twitch of cheek, nose or lip; in another it is a toss of the head, a sudden transient, yet ever recurring contortion of the neck, in a third it is a shrug of the shoulder, a convulsive movement of diaphragm or abdominal muscles — in fact the term embodies an infinite variety of bizarre actions that defy analysis.
>
> These tics are not infrequently associated with a highly characteristic cry or ejaculation — a sort of laryngeal or diaphragmatic chorea which may of itself constitute the condition; or there may be a more elaborate symptom in the form of a curious impulse to repeat the same word or the same exclamation. Sometimes the patient is driven to utter aloud what he would fain conceal.

Friedreich (1881) regarded tics as remembrance spasms suggesting that at the time of an intense fright in childhood the ticqueur had reacted with a series of muscle movements and that when external events replicated this experience spontaneous reproduction of the original movement occurred, and sustained excitability finally led to the occurrence of co-ordinated spasms without an external stimulus. Charcot was the first to appreciate that impulsive involuntary ideas or psychical tics commonly occurred together with bodily tics. Railton (1886) considered that occasional discharges from irritable nerve cells near the motor and speech areas in the cerebral cortex might underlie tics and Hammond (1892) developed this idea, postulating an irritative lesion of the motor cortex and subcortical structures as the cause of tics. Despite these early attempts to implicate a structural cause for tics the prevailing view was that they were psychologically determined and difficult to distinguish from hysterical convulsions. The monograph by Meige and Feindel published in 1902 consolidated the view that tics were a psychiatric illness. Their definition of tic runs as follows:

> A coordinated purposive act, provoked in the first instance by some external cause or by an idea; repetition leads to it becoming habitual, and finally to its involuntary reproduction without cause and for no purpose; at the same time as its form, intensity and frequency are exaggerated; it thus assumes the characters of a convulsive

> movement, inopportune and excessive; its execution is often preceded by an irresistible impulse, its suppression associated with malaise. The effect of distraction or of volitional effort is to diminish its activity; in sleep it disappears. It occurs in predisposed individuals who usually show other indications of mental instability

The psychological nature of tics was subscribed to by eminent neurologists such as Brain and Kinnier Wilson and was taken a stage further by Ferenczi (1921) who attempted to determine the underlying psychical traumas by psycho-analysis. Influenced by Freudian concepts he believed that masturbatory conflicts were at the root of tic production whereas others subsequently blamed narcissism or sublimated aggression. A wide range of incompatible doctrinaire theories appeared over the next 40 years, most extrapolated from observations on single patients.

In the last 20 years greater attention has been paid to the epidemiology of tics and there has been a swing towards a more eclectic view on their nature and how they should best be managed. The recent revelation that dopamine may modulate mood and behaviour in subcortical structures has provided further fuel for those who have always believed tics to be organically determined, and has led to the hypothesis that tics might result from a dopaminergic preponderance in an as yet undetermined region of the brain.

EPIDEMIOLOGY

Tics are primarily a disorder of childhood, the majority appearing between the ages of 6 and 8 years. They are common, occurring in at least one in 20 children but are a relatively rare reason for psychiatric referral (Zausmer, 1954; Debray-Ritzen and Dubois, 1980). Chronic simple tics may develop for the first time in adult life although an antecedent history of acute transient tics is often unearthed.

The first epidemiological survey was conducted by Boncour (1910) on 1759 French children aged between 2 and 13 years. An overall prevalence of 24% was found with an unexplained peak frequency in the 54 12-year-old boys of 50%. Tics occurred approximately equally in boys and girls and rudimentary tic-like movements were noted in children below the age of 6 years. Contrary to the accepted view at the time, Boncour found no appreciable difference between ticqueurs and normal children in scholastic achievements or conduct. Unfortunately, this study is marred by the failure of the author to define his diagnostic criteria and there is no information as to whether the tics were transient or persistent.

In the National Child Development Study, examination of 7970 healthy 7-year-olds revealed tics in 4% with an equal sex incidence (Kellmer Pringle et al, 1967). An overall frequency of 4% was also found in a large American study with a peak incidence of 10% in 6- and 7-year-olds (MacFarlane et al, 1954). 12% of a randomly selected group of 482 children living in Buffalo, aged between 6 and 12 years, were found to have tics. Again an equal sex incidence