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ADULTS WITH TOURETTE SYNDROME

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An Abstract of

A dissertation submitted to the Faculty of the Graduate School of Emory University
in partial fulfillment of the requirements for the degree of
Doctor of Philosophy

Graduate Institute of the Liberal Arts
2009

ABSTRACT

Adults with Tourette Syndrome

By Shlomit Ritz Finkelstein

The neuropsychiatric disorder Tourette syndrome (TS) is characterized by motor and vocal involuntary stereotypical tics. Its adult population (ATS) is little studied and often excluded from research. Only little medical help is available for ATS and many of them have limited access to health care due to lack of health insurance or inability to drive to a clinic.

ATS is the focus of this study. Its phenotypic presentation is heterogeneous with no reliable phenotype that serves as a basis for research. Instead there is a phenotype *spectrum* that may or may not include obsessive compulsive disorder (OCD), attention disorders like attention deficit disorder (ADD) and attention deficit hyperactivity disorder (ADHD), and more.

This study of ATS is interdisciplinary, drawing on the history of medicine, cognitive psychology, and neuroscience. The historical dynamics of the elusive TS include changes in its definitions and treatments from the 19th century of Gilles de la Tourette to the present. The dissertation is contextualized within this historical dynamics and aims to contribute to it.

In this qualitative study, data are gathered from video-interviews with sixteen adults with TS, video-interviews with their relatives, self reports, and medical evaluations. Based on these data, tics are classified in three ways: by modality, complexity, and triggers. A close study of coprolalia -- involuntary cursing afflicting some with TS -- is performed as well, with attention to the contribution of culture to this phenomenon and the possibilities and hindrances to its amelioration. The psychological, neurochemical, and neurosurgical interventions administered to the participants are studied and reflected upon.

This study suggests three major areas for closer and quantitative future investigation: (i) the role of the visual system in TS; (ii) the role of the somatosensory system in TS; and (iii) the possible application of mindfulness to the treatment of TS.

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To David and Aria, again

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PREFACE

> One morning in March of 1989, just before my fifth birthday, I woke up as a normal, healthy boy. By that afternoon, I had an irresistible urge to shake my head – continually – and the course of my life changed in ways few people had ever seen or could begin to understand. Before long, my body became an explosive, volatile, and unpredictable force with a mind and personality of its own. It jerked and twisted, bent in half, and gyrated without warning until I was almost always in motion. I bit down on my teeth until I actually broke them and howled in pain because of the exposed nerves. I twisted my back around with such force that I tore muscle tissue and had to be drugged asleep to stop myself from doing it. My mind fed me thoughts so frightening I couldn't even talk them to my parents. It didn't take long before I saw myself as the oddest person in my town."

This is how Cory Friedman introduces his Tourette syndrome (TS) (Patterson & Friedman, 2008:vii).

Why do I study TS? My initial drive was to understand the mind-body relation; a relation that has been explored from both dualist and monist perspectives for millennia. Already Socrates, Plato's teacher, expressed his dualist view in *Phaedo*, "[A]re we not part body part soul?... The soul is most like that which is divine, immortal, intelligible, uniform, indissoluble, and ever-consistent and invariable; where body is most like that which is human, mortal, multiform, unintelligible, dissoluble, and never self-consistent" (Plato, 1961:79b - 80b). But Plato's student, Aristotle the biologist, expressed a different view. In *De Anima* Aristotle presented the question of whether the soul has any parts that can do without the body. His answer was, "[T]here seems to be no case in which the soul can act or be acted upon without involving the body… it cannot be so divorced at all, since it is always found in a body" (Aristotle, 1984, *De Anima* 403a15).

I do not sketch here the history of the monist-dualist debate, but I do position myself in the camp of the monists. It has its own differences and disagreements about whether the mind (monists do not speak of souls) emerges from the brain, whether it is an epiphenomenon of the brain, or whether mind and brain are two different manifestations of something else, yet to be named. The challenge of studying any human phenomenon from a monist perspective is that one needs to simultaneously account for the biological and for all else that is human, including self and culture.

My doctorate in physics did not prepare me for this challenge. I had to return to school, where I decided to explore the mind-body relation by using TS as a window into the relations among language, culture, and neurology. The logic was the same as that of lesion studies. Since in TS, language, social behavior, and motor behavior all demonstrate various levels of malfunctioning, the disorder might express a nexus in which they all meet. If better understood, it would hopefully shed some light on the larger question of the mind-body relations.

Specifically I have been interested in the relations of language and culture to the *sensorimotor* system -- the system through which we perceive the world and act on it; the system that participates in every interaction between us and our environment. In addition, it is phylogenetically old and therefore likely to have been recruited by the phylogenetically-younger culture and language.

I started to interview people with TS. And as I met them face-to-face I became compelled to better understand their disorder, for its own sake, not as a window. For my current study I have given up my original goal of studying *through* TS. Instead I am

studying *it*. What started as an attempt to learn *from* Tourette syndrome has become a study *of* Tourette syndrome; in fact, of *people* who are afflicted with it.

INTRODUCTION

Tourette syndrome (TS) is a neuropsychiatric disorder characterized by involuntary stereotypical tics -- motoric and vocal. The motoric include, and are not limited to, eye blinking, shoulder shrugging, squinting, and hopping. The vocal include, and are not limited to, throat clearing, sniffing, and barking (James F Leckman, King , & Cohen, 1999). But the behavior for which TS is widely known and the one that has contributed to its bad reputation is called coprolalia. Of those who are diagnosed with TS nowadays, less than 20% suffer from coprolalia (Freeman, 2007).

The 19th century French neurologist Georges Gilles de la Tourette, after whom the syndrome is named, coined *coprolalia* after the Greek *kopros* dung, and *lalia*, speech. People who suffer from it curse involuntarily, with expletives that are often sexual, religious, or racial. Strangely and mysteriously, even though involuntary, the behavior demonstrates great sensitivity to the culture and to the specific situation. The sensitivity to the culture is demonstrated by violating what the culture forbids. For example, Japanese people with TS and coprolalia often curse with their prosody, not necessarily with their vocabulary, in accord with their culture, which interprets certain prosody as socially inappropriate (Baron-Cohen & Robertson, 1998). The sensitivity to the specific situation is demonstrated by the relevance of the violation. For example, racial slurs are typically uttered in racially mixed company and seldom otherwise (Chiten, Medley, & Russel, 1994).

A smaller percentage than the coprolalia population is of those with copropraxia – socially unacceptable gestures -- which is also sensitive to the culture and the situation. In the US it can be raising the middle finger, or touching one's own or another's genital

regions. Echolalia – repeating other's words, and echopraxia – mimicking other's movements, are also signs of TS.

The onset of TS is typically between the ages of 5 and 7. Its course is unpredictable as it tends to wax and wane, but many recover before adulthood (James F. Leckman & Cohen, 1999); those who do not, typically suffer from severe signs and symptoms[1]. The prevalence of the disorder is evaluated between 1-5% of the child and adolescent population (Comings, 2001; Mason, Banerjee, Eapen, Zeitlin, & Robertson, 1998), and is not known for adults. TS afflicts more males than females, and as with every aspect of it, there is disagreement on the sex ratio. The most extreme ratios that have been reported are 9:1 among children and over 3:1 among adults (Tanner, 2005).

TS has no biological markers. But studies of animal models and the response of patients to drugs, have led to consensus that the basal ganglia and the dopamine receptors and pathways are involved in the disorder. And therefore medications targeting the dopamine receptors of the basal ganglia have been administered to people with TS. Alas, the responses vary. Some people are helped; others have adverse side effects that outweigh the amelioration of signs and symptoms. A different treatment altogether should be administered if TS is caused by molecular mimicry[2] triggered by streptococcus infection, as is suspected by some. This path is studied for a pediatric population but little progress has been made so far. Psychogenic approaches were abandoned after being the

[1] Signs are observables, like temperature, rash, or a bacterium that can be identified in a lab test. Symptoms are subjective and known only through the report of the patient, like pain, or disturbing thoughts.

[2] Molecular mimicry is a hypothesis that a susceptible host acquires an infection with an agent that has antigens that are immunologically similar to the host antigens but differ sufficiently to induce an immune response when presented to T cells. As a result, the tolerance to auto-antigens breaks down, and the pathogen-specific immune response that is generated cross-reacts with host structures to cause tissue damage and disease (Albert & Inman, 1999).

main paradigm during the 19th century, and recently have regained interest (A. Peterson, 2007).

The phenotypic presentation of TS is heterogeneous (Alsobrook & Pauls, 2002). Despite a DSM-IV list of diagnostic signs of the disorder (*Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition*, 2008), there is an ongoing dispute over which signs to include in TS, and there is no reliable phenotype that serves as a basis for research. Instead there is a phenotype *spectrum* that may or may not include attention disorders – attention deficit disorder (ADD) and attention deficit hyperactivity disorder (ADHD) – alcoholism, affective disorders, obsessive compulsive disorder (OCD), and more. The search for the genotype has been mostly unsuccessful (D. Pauls, Cohen, Kidd, & Leckman, 1988; D. Pauls, Raymond, Leckman, & Stevenson, 1991; D. L. Pauls, Alsobrook, Gelernter, & Leckman, 1999).

The definition of the phenotype of TS has changed since Gilles de la Tourette, and the most severe cases have been marginalized over the years (H. I. Kushner, 1999). They are often excluded from research since their comorbidities make the interpretation of their data difficult; little medical help is available for them; and since many of them are adults, their access to health care is often limited due to lack of health insurance or inability to drive to a clinic. (H. I. Kushner, 2008b).

Adults with TS (ATS), who suffer greatly and are hardly studied and helped, are the focus of my research. As other psychiatric disorders, TS requires an interdisciplinary approach. I have drawn primarily on the disciplines of history of medicine, cognitive psychology, and neuroscience – each an interdisciplinary field in its own right.

Outline

The work is divided into ten chapters, each starting with two navigational aids: a table of contents, and an abstract-like paragraph "In this chapter." Chapters 2 and 3 explore Tourette's as a syndrome whose etiology is not known, rather than a disease whose cause is known and can be, for example, a bacterium. Chapter 2 is historical and explores the changes in the understanding and treatments of Tourette's since Gilles de la Tourette until our time. Chapter 3 concentrates on the present. It describes the challenges of selecting diagnostic criteria for TS and how these challenges are dealt with in the current *Diagnostic and Statistical Manual of Mental Disorders.* Then the chapter describes the onset, prevalence, and course of the disorder. Despite its unknown etiology, there are theories about the familial, neurological, and psychological nature of TS. They are summarized, and the interventions that are based on them are described. The spectrum nature of TS, its possible links to OCD and ADHD, and the severe phenotypes of adult TS (ATS)[3], which is the form studied in my research, are discussed.

The complexity and elusiveness of TS lead to difficult decisions about how to study it. In Chapter 4 I reflect on the tension between qualitative and quantitative methods and on how to extract hard conclusions from soft data. My study is explorative and primarily qualitative. Its purpose is to observe TS and generate specific questions and hypotheses that can be studied in following phases quantitatively. The interviews and questionnaires that supported my observations are described as well as the manners in which the data are documented and analyzed.

[3] I use ATS either for adults with TS, or to the subcategory of TS as experienced by adults. The context determines the use.

Chapter 5 introduces the participants. Each participant is described briefly, and then some of the interview is shared, letting the participants speak for themselves. In Chapter 6 I start analyzing my observations: I classify the tics along two axes – modality (vocal and motor) and complexity. The modality classification highlights some fine points about the gray zone between vocal and motor tics debated in the community. I also observe there that tic modalities tend to not mix. Only seldom does a person tic vocally and motorically at the same time. The complexity classification refines the current one and identifies simple, elaborate, symbolic, and violent tics. Chapter 7 classifies tics by what triggers them. The visual and somatosensory stimuli are important as well as the social and the internal worlds. The acoustic seem not to trigger tics and sometimes even ameliorate them. Some situations and activities ameliorate the tics or even cause them to disappear altogether.

The socially isolating coprolalia and copropraxia are discussed in Chapter 8, "Symbolic tics." I argue that they are not limited to the sexual and the religious spheres but instead violate cultural taboos. Therefore I propose replacing "copro" by "forbidden." *Coprolalia* is replaced by *forbidden speech*, *copropraxia* by *forbidden gesture*, and *coprophenomena* by *forbidden language*. The chapter studies the prevalence of the phenomenon, its social consequences, and whether and how it can be ameliorated. Finally, the chapter discusses possible neurological contributions to forbidden language.

The main goal of studying any disorder is to develop treatments for it. This is especially difficult for the elusive disorder of TS. Chapter 9 discusses the specific interventions that the participants in my study have undergone, their efficacy, side affects, and the contribution of the patients to the success or failure of their treatments. I

close this chapter with a personal reflection. In the last Chapter 10, “Future directions,” I propose general future directions for the research of TS. Three studies of special interest to me are: (i) the role of the visual system in TS; (ii) the role of the somatosensory system in TS; and (iii) the possible application of mindfulness to the treatment of TS.

I thank many in the “Acknowledgments” chapter. One special acknowledgement, to the 19th century British neurologist John Hughlings Jackson, runs through the whole work. He combined astute medical observations with deep philosophical considerations, and has been a source of inspiration for me. His words open some of the chapters.

TOURETTE'S – A SYNDROME NOT A DISEASE: HISTORICAL BACKGROUND

> *I fear we too frequently arrange our thoughts on cases according as the symptoms approach supposed types – such a genuine epilepsy, real chorea, etc. Then we hear it discussed whether genuine epilepsy is always attended by loss of consciousness or necessarily implies spasm of muscle... if the convulsions of children are true epilepsy or not, if epilepsy be hereditary… and frequently, of a particular case, whether it be a case of genuine epilepsy or not. Indeed I fear the student – it was so in my early medical career – imagines there is some entity of which epilepsy is the proper name.*

J. Hughlings Jackson in "Note on the comparison and contrast of regional palsy and spasm," *Lancet* 1867.

CHAPTER CONTENTS

IN THIS CHAPTER I discuss the difference between a disease and a syndrome. I then introduce the evolution of the definitions, understandings, and treatments of Tourette's[4], starting at the middle of the 19th century -- the time of Gilles de la Tourette -- and ending at the second half of the 20th century.

DISEASES AND SYNDROMES

To paraphrase Hughlings Jackson, we should be wary of the notion that there is some entity of which Tourette's is the proper name. Rather than a static labeling of a well-defined entity, "Tourette's" represents a dynamic process of understanding and constructing a syndrome, in which signs, symptoms[5], causes, diagnoses, interventions, and their relationships continue to mutually inform and affect each other. Syndromes are intricate and considered by many to be bio-psycho-social phenomena. Their very construction keeps changing, affected by and affecting their diagnoses and treatments.

A good starting point is a brief account of the concept of a disease. Unlike a syndrome, its cause, or causes, which are typically specific biological agents, are known, serve as its diagnostic criteria, and inform its treatments.

Diseases and their etiologies

The study of any illness strives to (i) be able to identify it; (ii) understand its cause or causes; and (iii) cure it; the former two leading hopefully to the latter. A great triumph of 19th century Western medicine was the understanding of bacterial diseases. Bacteria

[4] Tourette's was not always understood as a syndrome. Therefore, when I do not commit to a specific era or definition of it, I use *Tourette's* rather than *Tourette syndrome*, or *TS*.
[5] As was mentioned in the Introduction: Signs are observables, like temperature, rash, or a bacterium that can be identified in a lab test. Symptoms are subjective and known only through the report of the patient, like pain, or disturbing thoughts.

have been identified as causing diseases like cholera, diphtheria, tuberculosis, and others. The German physician Robert Koch (1843-1910), who discovered the bacteria that cause anthrax and tuberculosis, postulated two requirements to establish etiology of a disease: (i) the biological agent needs to be present at every occurrence of the disease, and (ii) the agent does not occur in other diseases as a fortuitous and nonpathogenic parasite[6] (Evans, 1993:163).

Such a one-to-one relation sometimes proved to be wrong for viral infections. The example of the poliomyelitis virus and the streptococcus bacterium is given by Thomas Rivers in his presidential address before the American Society of Immunology (1937): Some claimed that the streptococcus bacterium was the cause for poliomyelitis, also known as polio or infantile paralysis. Their claim was built on the fact that individuals recovering from poliomyelitis possess antibodies against the streptococci, and the bacterium was found in cultures obtained from patients with the malady. Importantly, when injected to monkeys and rabbits these cultures produced paralysis. We now know that poliomyelitis is a viral rather than bacterial disease, caused by the poliovirus, not by the streptococcus bacterium. What about the monkeys and the rabbits that were paralyzed? They did not have poliomyelitis, "Paralysis is not a characteristic sign of a single disease" (6). Both the streptococcus bacterium and the poliovirus can produce paralysis. And in general, various agents can produce the same signs or symptoms; this is not a one-to-one relation but rather a many-to-one relation.

[6] Koch originally stipulated three conditions. The third one was that after isolating the agent from a body, it should be repeatedly grown in pure culture and will succeed in inducing the disease anew. But later in his career he realized that this requirement can be a hindrance rather than a parameter for determining etiologies and adhered only to his first two conditions.

The reverse is also true. For example, each of the two species of the herpes virus family, HSV-1 and HSV-2, causes a variety of illnesses, like cold sores, chickenpox, shingles, encephalitis, and various cancers. This is a one-to-many relation. The case of swine influenza that was discovered in 1931 by Shope is a good example of many-to-many relation in which an infectious disease is caused by two agents – a virus and a hemophilic bacterium (cited in Evans, 1993).

In summary, the idea of a one-to-one causal relation between a specific biological agent and a specific disease is often simplistic, and needs to be replaced by a many-to-many paradigm: A single cause can lead to a spectrum rather than to a single clinical presentation; different mechanisms can cause similar presentations; and a synergy or combination of more than one mechanism can lead to a spectrum of clinical presentations. Epidemiological studies further add to the complexity by showing that the predominant agent of a certain disease may vary by geographical location and the type of the population (e.g., age or military vs. civilian population) (Evans, 1993)[7]. In spite of this complexity, diseases are still linked to known biological agents; and as such they are simpler and less mysterious than syndromes.

Syndromes and etiologies

Complex or simple, one-to-one or many-to-many, the 19th century celebrated a triumph over diseases. Many bacterial diseases, like tuberculosis, were successfully diagnosed and cured with antibiotics, and many viral diseases, like polio, were successfully prevented by a vaccine. This biological success and its promise led people

[7] Many other complexities, like the mutations of the agents, are not even mentioned here. I only intended to shake-off the one-to-one paradigm in establishing causal relations between an illness and its etiology.

like Freud to seek a theory that would account for psychiatric disorders in the same way (Hacking, 1995). Such a search for the Philosopher's Stone is also reflected in the work of the American psychiatrist Arthur Shapiro and his colleagues. They opposed considering interaction between organic susceptibility and psychopathology in Tourette syndrome (TS) since it "does not appear to us to be parsimonious." (1988:162). Alas, in spite of the elegance of parsimonious explanations, it seems so far that Tourette's should be understood as a network of causes and a spectrum of signs and symptoms. In other words, it is a syndrome, not a disease.

Understanding its web of signs, symptoms, interventions, and causes, is a *dynamic process*. Our current ignorance is not total; our knowledge and understanding are incomplete. We have some insights, among them that the causes of the syndrome are not only biological. Psychological, social, and cultural aspects contribute as well, sometimes in subtle ways. A great challenge is the question of the phenotype, the signs and symptoms that make up a presentation of the disorder. The phenotype is not robust, and not two patients are alike.

Diagnoses, etiologies, and interventions

Diagnostic criteria, knowledge of etiology, and interventions are intricately linked, and they all participate in the ongoing process that seeks to improve them. The main source of knowledge is the patient; each adds to the understanding of the disorder. Theories of the syndrome's etiology influence interventions. When these interventions prove helpful the success reinforces the causal explanation, and otherwise their failure diminishes the credibility of the proposed etiology. This is a trial-and-error process, in which the individual may pay a price, sometimes high. While the treatments are intended

to heal, they do not always do: Side effects can be worse than the original complaints, and often choosing one kind of therapy closes the gate to others. In this unavoidable trial-and-error process, the "error" can increase the person's suffering.

The theoretical framework is simultaneously a guide and a bias. Explanations are typically biased in favor of either the biological or the psychogenic. As cognitive neuroscience continues to evolve, the gap is narrowing. For a growing number of scientists and clinicians the psychogenic and the biological are not in dichotomy anymore. Yet the road ahead is long and interventions that include both are still rare or non-existent.

Finally, in the absence of known etiology, diagnosis becomes nosologic, relying on signs and symptoms. There is a need for a robust phenotype. Which signs and symptoms should be the diagnostic criteria of TS? Some other disorders, as we will soon see, have similar signs, yet are otherwise classified, based on etiological knowledge or on the course of the disorder. In addition comorbidities further complicate the picture. Tourette syndrome seldom comes alone. Often it is accompanied with obsessive compulsive disorder (OCD), attention deficit hyperactivity disorder (ADHD), and self injurious behavior (SIB).

THE HISTORY OF THE DEFINITIONS OF TOURETTE SYNDROME

Kushner observes that "[t]he longer a syndrome's causes remain unidentified, the more disagreements can emerge over which signs and symptoms are necessary to authorize a diagnosis." This is partly because "the constituents of a syndrome can vary over time and in different places" (2008a:552); and partly because new data and new possible explanations emerge. Studying the history of TS reinforces Kushner's

observations and in addition reminds us of long-forgotten ideas from which we can benefit, and guards us from those that were proven wrong.

I will not attempt to repeat the detailed and deep study *A Cursing Brain? The Histories of Tourette Syndrome* (H. I. Kushner, 1999). Instead I will summarize some of Kushner's historical insights about how we have reached our current understanding and definition of TS. I start in 19th century Paris. The topic in question is that of chorea. The word is derived from the Greek *Khoreia*, meaning dance, and applies to a collection of clinical conditions of movement disorders. The questions debated are: (i) Are all choreas alike? (ii) Is chorea hereditary? ("Genetic" had no meaning in the 19th century). (iii) Is chorea caused by infection? (iv) Is chorea a psychopathology? (v) How to differentiate choreas and diagnose them?

I take here three movement disorders with some symptomatic resemblance – Tourette's, startle effects, and Sydenham's chorea. Through them I will present some of the chorea discourse. I start with the one that is the focus of this study.

La maladie des tics de Gilles de la Tourette

La maladie des tics was named after Gilles de la Tourette (1857-1904) by his famous mentor, Jean-Martin Charcot (1825-1893), chief physician of the Salpêtrière Hospital in Paris (H. I. Kushner, 1999). For Gilles de la Tourette, the disorder bearing his name was a disease -- *maladie*, not a *syndrome.* His starting point in studying the disorder was the phenotype.

He collected nine case studies of patients with convulsive tics and compared them with three studies of Startle disorders that were observed in Malaysia, Siberia, and Maine. He relied in part on reports of others; only two of the people described in his publication

were seen and examined by him. Some first-hand descriptions were published more than half a century prior to Gilles de la Tourette by the French neurologist Jean Marc Gaspar Itard (1775-1838). One of Itard's patients was the Marquise de Dampierre.

Itard examined her at the age of 26, already eighteen years after the first outbreak of her symptoms. He saw her only briefly, and we know nothing about the later course of her disorder. She became famous in part due to her high social status and in part due to the fact that she lived to the age of 85. More importantly, she had become the emblem of the syndrome for Gilles de la Tourette, even though he never examined her himself (H. I. Kushner, 1999). This is how Itard described the marquise's behavior:

> In the midst of a conversation that interests her extremely, all of a sudden, without being able to prevent it, she interrupts what she is saying or what she is listening to with bizarre shouts and with words that are even more extraordinary and which make a deplorable contrast with her intellect and her distinguished manners. These words are for the most part gross swear words and obscene epithets and, something that is no less embarrassing for her than for the listeners, an extremely crude expression of a judgment or of an unfavorable opinion of someone in the group (cited in and translated by H. I. Kushner, 1999:10).

Gilles de la Tourette named such involuntary cursing and verbal violation of social conventions *coprolalia*, from the Greek *kopros* (dung) and *lalia* (talk). For Charcot and Gilles de la Tourette, coprolalia was a necessary diagnostic criterion for the disorder; so was echolalia, the involuntary echoing of the speech of others. The syndrome was "a nervous affection characterized by lack of motor coordination accompanied by echolalia and coprolalia" (cited in Teive, Germiniani, Coletta, & Werneck, 2001: 725).

In an 1885 Italian translation of one of the weekly public talks given by Charcot at the Salpêtrière, the translator Melotti[8] presents the Charcot definition of Gilles de la Tourette's disease as having "three major elements: tics, echolalia and coprolalia. The tics differ from simple tics because of their intensity and because of the complexity of their movements, which simulate chorea. They are isolated movements of the face and of the arm on the affected side, or general movements on one side of the body. Movement can be so extreme that the patient even slips on one side" (H. I. Kushner, Luzzatti, & Finger, 1999:9).

Both Charcot and Gilles de la Tourette understood *la maladie des tics* as a hereditary disorder whose underlying pathology could be identified in the familial history of the patient. Hereditary mechanisms were not understood at the time, and it was not clear what exactly was inherited; but family trees were drawn showing the transmission of hereditary disposition for the disease. The disease was understood as a progressive illness. And while it could wax and wane, on the whole it would get worse with age, and would eventually include coprolalia and echolalia. It would also resist all treatments (H. I. Kushner, 1999).

In his last communication at 1899, Gilles de la Tourette summarized the disease named after him: (i) Its signs are motor and vocal tics; (ii) it is hereditary; (iii) its onset is between the ages of seven or eight; and (iv) it affects "the sexes… equally." (cited in Meige & Feindel, 1907/1990:224). And while he stated hereditary disposition, he admitted that the course of the disease was "[u]nder the influence of causes whose action we are, in the majority of case, powerless to appreciate" (224). Nevertheless, he did

[8] Kushner and colleagues question some of the authenticity of Melotti's translation. But based on other writing of Charcot, they accept the quotes that I bring in this discussion.

attribute the cause of "a complete absence of the moral sense" to coprolalia and considered its impulse "psychical" (224). Since no anatomical lesions were found among patients of the disease, the accepted explanation by Charcot and his circle was that the course of the disease was due to psychological degeneration (for more see H. I. Kushner, 1999: 26-44).

Exaggerated startle effects

"Exaggerated startle effect syndromes" is a collective name for syndromes whose signs are evoked by a sudden touch, a sudden sound, or a sudden motion. Some reports about startle effects caught the attention of Gilles de la Tourette who considered them occurrences of *maladies des tics* (Gilles de la Tourette, 1884), since they shared the involuntary echoing of others' and one's own speech and actions.

Jumping is the name that was given to the excessive startle response among the French population of northern Maine, especially in the region of Moosehead Lake. The behavior was first documented in 1878 by the American psychiatrist George M. Beard, who saw 50 Jumpers, 14 of them belonged to 4 families. It was claimed that they were all of French descent and of Canadian birth. Most of them were occupied in lumbering in the Maine woods. (Stevens, 1965). The signs of their syndrome included jumping, screaming, echolalia, echopraxia[9], coprolalia, copropraxia[10], and involuntary obedience. One example of the echolalia and involuntary obedience of one Jumper, is described by Beard: "While sitting in a chair, with a knife in his hand, with which he was about to cut tobacco, he was struck sharply on the shoulder, and told to 'throw it.' Almost as quick as

[9] Echopraxia is involuntary echoing of other's actions.
[10] Copropraxia is involuntary socially unacceptable gestures, like rising the middle finger.

the explosion of a pistol, he threw the knife, and it stuck in a beam opposite; at the same time he repeated the order 'throw it' with a certain cry as of terror or alarm" (1880:171).

Myriachit is a startle effect observed in Eastern Siberia. The report of three American Navy officers on their way from the Pacific through Asia to the United States caught the attention of Dr. William Hammond, who treated many patients with convulsive tics (G. M. Hammond, 1892). He cited the Navy officers' description and proposed some neurological speculations (W. A. Hammond, 1884). *Myriachit* runs in families, and Hammond reports of one family in which four children repeated words and motions that were performed in their presence. In a summary of ten *Myriachit* studies, Caroline Tanner and Jerry Chumberland report that coprolalia was observed in nine of the ten studies (Tanner & Chamberland, 2001).

In his book *Latah in Southeast Asia: The History and Ethnography of a Culture-bound Syndrome,* Robert Winzeler describes the Malayan *Latah* (1995). *Latah* (the word stands both for the behavioral signs and the afflicted person) typically runs in families and includes involuntary cursing and utterances of obscenities. The *latah*s are tolerated in their society but often become the target of mockery:

> I asked a series of questions about who, if anyone, deliberately teased them… A very few people replied by saying that no one tried to make them *latah* while a great many more said that *semua orang* ('everyone') did… Only in one or two instances did *latah* persons say that they were only provoked by persons who were 'outsiders' (*orang luar*), persons with whom they had no significant relationships of one sort or another." (p. 93).

The extent of the involuntary nature of *latah* and its echopraxia is demonstrated by the tragic story documented in the 1883 notes of the British adventurer H. A. O'Brien

> This cook [the *latah*] was dandling his child forward one day; one of the crew came and stood before him with a billet of wood in his arms., which he began nursing in the same way as the *latah* was nursing his baby. Presently he began tossing the billet up to the awning, and the cook tossed his child up also, time for time. At last, the sailor opened his hands wide apart and let the wood fall upon the deck, and the cook immediately spread out his hands away from the descending child, who never moved again after striking the boards" (1883:11).

While *Jumping, Myriachit,* and *Latah* share much, they also differ. For example, most of the anecdotes about Jumpers involve men. *Latah* and *Myriachit*, by contrast, affect mostly middle-aged women (Chapel, 1970). The startle effect syndromes also differ enough from Tourette's to agree with Charcot and doubt Gilles de la Tourette's speculation that they are the same: (i) Charcot pointed to the lack of motor tics in startle effects; (ii) Unlike *Latah* and *Myriachit*, the Tourette's population is overwhelmingly male. Of this, Gilles de la Tourette was not aware "for the sexes are affected equally" (cited in Meige & Feindel, 1907/1990:224); (iii) In TS the onset of the tics is early. *Latah* often emerges late in life (Simons, 1983); and finally (iv) Startle effects are triggered by sudden touch or a loud sound. By contrast, Tourette's seems to not be triggered by sound, and its response to touch is not universal and is not as extreme as in the startle syndrome. I will return to this important point in the chapter "Tic classification," where I discuss the phenotype of TS as it appeared in my pilot study.

All these syndromes, including Tourette's, run in families, which might point to their hereditary nature. But in families behaviors are learned as well. Do the Jumpers of Maine suffer from a hereditary disease; or as suggested by P. M. Yap (1951), are they all members of a religious group that learned and automated certain patterns of behavior?

Sydenham's chorea

Sydenham's chorea is a movement disorder whose signs resemble the ecstatic dancing that was widely practiced in Europe in the 15th and 16th centuries by people who made pilgrimage to the shrine of St. Vitus in the hope of being healed. This resemblance led the Swiss scientist, physician, and occultist Paracelsus (1493-1541) to name it "St. Vitus dance." In 1686, the British physiologist Thomas Sydenham (1624-1689) commented on St. Vitus chorea and restricted it to a specific chorea, *chorea minor,* which later was named after him. Here is an extract from his writing, translated from the Latin in 1848:

> This is a kind of convulsion, which attacks boys and girls from the tenth year to the time of puberty. It first shows itself by limping or unsteadiness in one of the legs, which the patient *drags*. The hand cannot be steady for a moment. It passes from one position to another by a convulsive movement, however, much the patient may strive to the contrary. Before he can raise a cup to his lips, he makes as many gesticulations as a mountebank; since he does not move it in a straight line, but had his hand drawn aside by spasms, until by some good fortune he brings it at last to his mouth. He then gulps it off at once, so suddenly and so greedily as to look as if he were trying to amuse the lookers on. (cited in Finger, 1994:222).

Meige and Feindel observed that "If we were to confine ourselves to this description by Sydenham… differentiation between tic and chorea would not be a matter of any complexity" (Meige & Feindel, 1907/1990:279). And indeed the unilaterality of Sydenham's chorea and its difficulty in completing purposeful motions are not characteristic to tics. In addition the vocal tics, with their most florid forms being coprolalia and echolalia, are not necessarily present in Sydenham's chorea. The difficulty in differentiation, according to Meige and Feindel, is that "in practice one constantly

meets with conditions suggesting alike the gesticulations of chorea and the convulsive reaction of tic" (279).

Etiologically, Sydenham's chorea is interesting and might be important to the understanding of other movement disorders. By the 19th century, based on concomitance with involuntary tics following rheumatic fever, and autopsies that revealed heart damage, the general consensus was that *rheumatic fever* is the cause for Sydenham's chorea. Rheumatic fever was understood by many as a disposition not only for Sydenham's chorea but for a whole range of choreas, depending on which organ in the body was affected. Based on many case studies on both sides of the French Channel, the distinguished French neurologist Armand Trousseau wrote in 1862 that "of all the predisposing pathological states [of choreas], rheumatism is assuredly the most marked and the least questionable"(cited in H. I. Kushner, 1999:34).

Chorea or choreas?

In the talk translated by Melotti and cited above, Charcot said,

> Last year I had a patient in the ward, who had a tic with echolalia and coprolalia, phenomena which are features of these exotic diseases [Jumping, *Latah*, and *Myriachit*]. Gilles de la Tourette, at that time my intern, studied these cases, which he collected with some similar ones, and tried to describe their history, believing to have encountered a new disease that he called *motor incoordination with echolalia and coprolalia*. I would prefer to call it *convulsive tic with coprolalia and echolalia* and I do not believe it to be a special disease. On the contrary, I think it is a mistake to consider it the same phenomenon as *jumping* and *latach* (sic) in which tics are not mentioned, whereas in Gilles' (sic) disease tics are the essential element (H. I. Kushner et al., 1999:8).

Charcot adopted a nosological[11] differentiation between *maladie des tics de Gilles de la Tourette* and the startle effect disorders *jumping* and *latah*. While for *maladie des tics de Gilles de la Tourette* tics were the essence, for the startle effect disorders tics were not mentioned. At the same time Charcot considered similar hereditary disposition for different choreas. He compared the hereditary transmission to a tree with various branches, with each branch standing for a specific and differentiated movement disorder. His Sydenham's chorea branch included rheumatic fever but the familial branch of *maladie des tics de Gilles de la Tourette* did not.

Tourette's – congenital, or infection-induced?

Like his teacher, Gilles de la Tourette adhered to a hereditary etiology with psychological degeneration, insisting that no anatomical pathology was to be found in those who suffered from *maladie des tics*. This diverted attention away from the possibility of an infection[12]. The case of the patient G. D. is telling. Three months after contracting tonsillitis -- inflammation of the tonsils -- the 15-year-old G. D. was seen by Gilles de la Tourette with severe motor spasms and coprolalia. But even though the boy mentioned his previous tonsillitis, Gilles de la Tourette did not inquire about past occurrences of infections and their possible relations to the tics and their severity (H. I. Kushner, 1999:40). Other similar stories cited by Kushner portray a battle of ideas in which possible link between infections and choreas got pushed away from the mainstream of psychiatry. And finally, "by the early twentieth century the connection

[11] Nosological diagnosis or differentiation depends on signs alone.
[12] A possible link between rheumatic infection and chorea is understood today in terms of molecular mimicry. This explanation was not available at the time of Gilles de la Tourette.

between convulsive tics, Sydenham's chorea, and rheumatic fever almost totally disappear[ed]" (43).

In spite of deep disagreements about the etiology and the differentiation diagnoses of choreas, there was a consensus that the etiology of any chorea included hereditary transmission and its course was due to psychopathology, to "psychological degeneration" (39). The greatest support for this view, Kushner tells us, came from "the publication of Henry Meige and E. Feindel's extremely influential 1902 *Les Tics et leur Traitment*" (44), which was later published in English in 1907 (Meige & Feindel, 1907/1990). Under its influence, the view that the phenomena depend on acquired lesion was removed from the medical discourse of tics. In other words, lesions caused by an infection were not considered as a possible etiology for tics. However, it is important to acknowledge that Meige and Feindel did not deny the possibility of biological origins, as long as they were congenital rather than infection-induced: "[W]e inclined to believe that they [the tics] represent some congenital anomaly, some arrest or defect in the development of cortical associations paths or subcortical anastomoses [(networks)], minute teratological [(abnormal in growth or structure)] malformations that our medical knowledge is still unhappily powerless to appreciate" (117).

Staying loyal to "the doctrine hallowed by the authority of Charcot, and since advocated by Professor Brissaud" (xiii)[13], Meige and Feindel propagated the psychogenic theory of tics and accordingly advocated behavioral and psychological therapies. They also posited that "[t]he prognosis in a case of a tic *depends solely on the mental state* of

[13] This adherence is not surprising since Meige studied with Charcot at the Salpêtrière, and later worked with Édouard Brissaud, himself a student of Charcot, and the one who wrote the preface for the book. E. Feindel was a patient of Brissaud after he had been found comatose in a Paris street. Later he was encouraged by Brissaud to pursue his studies, earned his medical degree, and became an important researcher of neurology ("Obituary. E.Feindel, M.D.," 1930).

the patient" (293, my italics). Their chapter about treatments is grim. In it they cite many practitioners who feel helpless. Among them is Charcot: "We cannot say that cure is certain, but we may count on longer or shorter intervals of arrest, either spontaneous or as a sequel to the employment of serviceable measures such as hydrotherapy or rational gymnastics" (cited in p.300). Meige and Feindel qualify this helplessness, though, only to cases of "graver nature." After summarizing treatments by medications, diet, massage, electrotherapy, suggestion, surgeries, and orthopedic treatments, they resort to propose "treatment by re-education."

Their re-education involves a range of therapies: Trousseau "extolled the value of exercises systematically applied to the muscles involved in non-dolorous tics" (315). Letulle "advises an appeal to the intelligence, good sense, and will of the patient in the endeavor to provoke an inverse effort at the moment when the tic begins, or even before" (315). The French psychiatrist Pierre Janet (1859-1947) advocates "[e]ducation of movements by some form of drill" (316). An example of reeducation is provided by Meige and Feindel: "One of our patients, suffering from facial tic, was directed to perform, as far as practicable, the opposite movements to her grimaces. If her mouth was drawn to the right, she forthwith made a corresponding twitch to the left; is her mouth was shut spasmodically, she was instructed to open it widely and quickly" (322).

In addition Meige and Feindel emphasized the need to provide therapy within a supportive structure and advocated "a daily routine for the patient to follow regularly and punctually" (320). This is how they described and interpreted the condition of one of their patients: "His mental disarray is patent not merely from his disorders of motility, but in the unmethodical and changeable habits of his everyday life" (320). An "excellent

result" was reported by Tissié – "a complete cure" of a very severe case of a "young man [who] had suffered for eleven years from generalized tics… Every few seconds, violent twitches of electric-like rapidity seized the muscles of his head, trunks and limbs, to the accompaniment of abrupt cries and inarticulate growls" (cited in 326). The therapy addressed the respiratory system, and its success was attributed by Meige and Feindel to the "bestowal of the attention of the allotted task" (327). Tissié disagreed; he saw a conflict between deep respiration and attention.

The harshest treatment recommended by Meige and Feindel's was isolation that they admitted was "a rather severe proceeding," but nonetheless "one must not hesitate to utilize in rebellious cases, or if the patient's mental state precludes the possibility of prolonged application of systematic discipline" (341).[14] For them "psychotherapy is… of capital importance" (343). In addition, when considering both isolation and psychotherapy they emphasized the indispensability of supportive human milieu, especially of the psychiatrist and the parents. The parents were advised to "appreciate the importance of discipline and unite, intelligently and assiduously, in the task of education," and the physician "need have no fear of damaging his professional prestige by the simplicity of his methods" (345), which often characterizes the methods of reeducation.

In spite of the immense influence of Meige and Feindel with their behavioral psychology approach, theirs was not the only voice. French, German, English, and American physicians strongly debated over psychological, congenital, or infection-

[14] Isolation, in which the patient is removed from his environment and typically admitted to an asylum, deserves its own discussion. Foucault and Goffman expressed their strong criticism. But this is not the right place for this discussion.

induced etiologies. But the psychogenic narrative prevailed and paved the road for the psychoanalytic approach (H. I. Kushner, 1999).

Even against Freud's acknowledgement that the origin of convulsive tics was most likely organic, the Hungarian psychoanalyst Sandor Ferenczi resorted to psychoanalysis as a treatment for tics. His student Margaret Mahler (1897-1985) escaped the Nazis to the USA and became the most prominent and influential psychoanalyst to treat tics, working mainly with child and adolescent population. Kushner dedicates a detailed chapter to describing her work and some of the patients that she treated. A few points deserve attention: (i) Like Gilles de la Tourette, Margaret Mahler found *maladie des tics* to be progressive; (ii) Like Meige and Feindel that found some of their patients to be "rebellious," Mahler found some of her patients to be "resistive to giving up defensive mechanism" (H. I. Kushner, 1999:117), and through this description put some of the blame on them; and finally (iii) "[T]here was no direct correlation between recovery from the tic syndrome and the length, method, or thoroughness of treatment" (118).

The three schools, of psychogenic, congenital, and infection-induced etiologies, continued to debate up to the late 1960's. Then, great advancements in psychopharmacology and the leadership of the American psychiatrist Arthur K. Shapiro (1923-1995) and his wife Dr. Elaine S. Shapiro, supported by their successful interventions with haloperidol, brought about what Kushner calls "the triumph of the organic narrative." And this is how it still is in our current time. The mystery, however, has not been removed and Tourette's, as was observed by the Italian Robert Massalongo in 1895, "is not one morbid entity but a *clinical syndrome*; etiologically there are many

choreas, as there are many epilepsies" (H. I. Kushner, 1999:39). There are also many interventions, alas none with great efficacy.

NEXT

I describe the current state of affairs…

TOURETTE'S – A SYNDROME NOT A DISEASE: THE CURRENT STATE OF AFFAIRS

CHAPTER CONTENTS

IN THIS CHAPTER I introduce the current definition of Tourette syndrome, which is articulated in the *Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition*, also referred to as DSM-IV. I then discuss issues of reliability and validity of the manual. The course, onset, and prevalence of the disorder, as they are defined today, are then described.

The search for etiology has produced much, even if not complete, knowledge about the familial, neurological, and psychological nature of the disorder. This knowledge is reflected in and contributing to interventions – psychological, pharmacological, and neurosurgical. I describe them too.

Finally, Tourette's is often not by itself. As a spectrum disorder it is linked with other disorders either by comorbidity or by belonging to the same spectrum. I discuss the disorder of obsessive-compulsive disorder. And I pay special attention to adults with severe phenotypes

THE CURRENT STATE OF AFFAIRS

Currently, Tourette's is considered a syndrome, not a disease. It is understood mainly in congenital biological terms. Psychological conditions like stress or concentration respectively contribute to the exacerbation or amelioration of its signs and symptoms, but are not considered etiological. The possibility that it is infection-induced is studied only for children, under the title PANDAS -- Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal infections. And the classification and diagnostic criteria for Tourette syndrome (TS), as for other psychiatric

disorders, are in the fourth edition of the *Diagnostic and Statistical Manual of Mental Disorders* (DSM-IV).

Nosological diagnostics -- the DSM and the IDC

The ongoing changes in the understanding of Tourette's have not happened in a medical and intellectual vacuum. For the last 150 years, these changes have been quite attuned with the changes of the *zeitgeist*. And for psychiatric disorders, this *zeitgeist* influenced and was influenced by two manuals, the *International Classification of Diseases* (ICD) and the *Diagnostic and Statistical Manual of Mental Disorders* (DSM). ICD is now in its 10th edition and the DSM is in its 4th, while people are already working towards the 5th. These indicate the difficulty of defining psychiatric disorders, the disagreements in the community, the unavoidable shortcomings of any approach adopted, and the ongoing changes in understanding of psychiatric disorder, not only in terms of their diagnostic criteria, but also in their classification and relationships to each other.

During their evolutions, the ICD and the DSM have been converging. Therefore I will not discuss the ICD. Nor will I introduce the history of DSM; I will only highlight a few points that help contextualize the construction of TS. A major one is that the DSM-III, published in 1980 under the leadership of Robert Spitzer of Columbia University, has been a turning point in the evolution of the DSM, changing the manual from its psychoanalytical DSM-II into an "atheoretical descriptive approach" (Spitzer, 1987:3). This change parallels the change in the understanding and treatments of TS, which switched from the psychological perspective of Meige and Feindel and the psychoanalytical perspective of Mahler to the biological perspective pioneered by Shapiro.

Like all the disorders in DSM-III and its following revision, and the DSM-IV and its text revision, the classification and diagnostic criteria of TS are nosological – by clusters of signs and symptoms that might reveal putative distinct disorders with an underlying mechanism, but with no presumptions about what this mechanism is -- what the disorder's etiology is.

Many of the TS signs included in the DSM-IV were not there in the past; they were too mild. And some of the severe ones are not mandatory any more. Opening the gate for mild signs is part of a process of medicalization -- an aspect of our own *zeitgeist.* It has changed the definition of TS as well as of other psychiatric disorders, notably depression. There are a few reasons for this change of the phenotype, including the power of the pharmaceutical industry over current psychiatry, on which I will not elaborate[15]. The historian of medicine Edward Shorter attributes this process in part to the fact that "[p]hysicians prefer to diagnose conditions they can treat rather than those they can't" (Shorter, 1997:291). A similar observation, pertaining specifically to TS, is made by Kushner who points out that expansion of the phenotype by including milder cases leads to an impression of success of treatments, since many mild cases, some of which would have recovered spontaneously, appear to be successfully treated. But the greater harm is that the florid cases, those who are desperate for help, are marginalized and are typically dropped from research and medical care (H. I. Kushner, 2008b).

Those florid cases of TS typically suffer from coprolalia and echolalia, which fit the criteria of Charcot and Gilles de la Tourette but are no longer mandatory for the diagnosis of TS. Other involuntary behaviors like inappropriate gestures (copropraxia),

[15] Important writings about this unsettling relations are pioneered by David Healy and Eliott Valenstein (for example see Healy, 1998; Valenstein, 2000).

repeating other's motions (echopraxia), and repeating one's own words (palilalia) are not even mentioned in the DSM-IV. The sufferers almost always continue to have these severe signs throughout their lives. But in the competition on medical attention, resources, and empathy they often lose to the pediatric and adolescent populations, in which many present only mild phenotypes.

DSM-IV and the diagnosis of Tourette's syndrome

There are four diagnostic criteria for TS as published in section 307.23 of the Text Revision DSM-IV-TR *(Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition, 2008)*:

Diagnostic criteria for 307.23 Tourette's Disorder

A. Both multiple motor and one or more vocal tics have been present at some time during the illness, although not necessarily concurrently. (A *tic* is a sudden, rapid, recurrent, nonrhythmic, stereotyped motor movement or vocalization.)
B. The tics occur many times a day (usually in bouts) nearly every day or intermittently throughout a period of more than 1 year, and during this period there was never a tic-free period of more than 3 consecutive months.
C. The onset is before age 18 years.
D. The disturbance is not due to the direct physiological effects of a substance (e.g., stimulants) or a general medical condition (e.g., Huntington's disease or postviral encephalitis).

The criteria (A) and (B) describe the signs of TS and their frequencies. According to them, Tourette syndrome is characterized by involuntary and sudden tics, which can be

motor or phonic[16]; and both need to be present. In the section "Diagnostic Features," the tics and their anatomical locations are described:

> The anatomical location, number, frequency, complexity, and severity of the tics change over time. Simple and complex motor tics may affect any part of the body, including the face, head, torso, and upper and lower limbs. Simple motor tics are rapid, meaningless contractions of one or a few muscles, such as eye blinking. Complex motor tics involving touching, squatting, deep knee bends, retracing steps, and twirling when walking may be present. The vocal tics include various words or sounds such as clicks, grunts, yelps, barks, sniffs, snorts, and coughs.

As was mentioned above, Gilles de la Tourette's diagnostic criteria of coprolalia and echolalia are no longer mandatory. Instead, "Coprolalia, a complex vocal tic involving the uttering of obscenities, is present in only a small minority of individuals (less than 10%) and *is not required for a diagnosis of Tourette's Disorder*." (my italics). And indeed, in an analysis of a large international database, the Canadian neurologist Roger Freeman reports than among 6805 participants diagnosed with TS, only 13% suffer from coprophenomena -- coprolalia or copropraxia (Roger D. Freeman, 2007: Table 4).

Criterion (C) of the DSM-IV-TR is about the onset time, which needs to be before the age of 18. The last criterion (D) touches upon etiology. While it cannot and does not specify the etiologies of TS, it excludes the etiologies of substance-use or Huntington's disease, whose genetic origin is known. The exclusion of postviral encephalitis strongly suggests that TS-like tics following infections, including streptococcus infections, are also to be excluded. This is in the spirit of Gilles de la Tourette, who, as mentioned above, paid no attention to the tonsillitis of his patient G. D. This would exclude

[16] Like many in the community, I use here "vocal tics" and "phonic tics" interchangeably. Their difference is discussed in the chapter "Tic classification." The current trend in the community prefers "phonic" but as I will show later, there is room for both "vocal" and "phonic."

Sydenham's chorea from possibly being TS. However, growing understanding of molecular mimicry has made room for other opinions.

Reliability and validity in the DSM-III and DSM-IV

The consensus is that starting with DSM-III the manual has followed in the footsteps of the German psychiatrist Emil Kraepelin (1856-1926). And in some ways it has. Like Kraepelin, who divided all psychiatric illness into 13 big groups (Shorter, 1997), the DSM has also classified psychiatric illness; but with over 200 disorders, a much greater number of classes than 13. Like Kraepelin's, the DSM's classification is nosological. And like Kraepelin, who understood the construction of a psychiatric disorder as a *process,* and expressed this understanding through publishing successive versions of his textbook (105), so the DSM has ongoing publications of new versions of the manual, each with modified or new definitions.

As with any other scientific endeavor, diagnosticians attempt to achieve reliability and validity. A strong criticism of the DSM-IV is that it sacrifices validity for reliability. Reliability measures the agreement among diagnoses by different practitioners. The nosological definitions of the DSM-IV support reliability, even though the TS diagnosis has remained "an uncertain art" (Towbin, Peterson, Cohen, & Leckman, 1999:118). Having a list of signs is insufficient. The experience of the practitioners matters; the more experience, the greater the agreement among diagnosing physicians (cited in Burke, 1987).

The question of validity is tightly linked to the definition of the disorder, and is more challenging. How do we know that what we diagnose actually exists? How do we know that what we diagnose is what we claim we diagnose? The validity of the diagnosis

of a psychiatric disorder is elusive until the etiology of the disorder is understood. Since etiology for psychiatric disorders might never be found, the pragmatic test for the validity of diagnostic criteria is whether they "carv[e] nature at the joints," to use Shorter's paraphrase of Kraepelin (Helzer & Hudziak, 2002; Shorter, 1997:105). To this end Kraepelin built a database. His database was a collection of cards in which he documented his observations of his patients, and data gathered from patients (Shorter, 1997:105). In other words, he collected signs and symptoms. By contrast, the DSM-IV definition of TS relies only on signs. While the symptomatic obsessions of obsessive compulsive disorder (OCD) are among the diagnostic criteria of OCD, the symptomatic urges that precede many tics are not. They are only briefly described in the "Differential Diagnosis."

The course of Tourette syndrome

Kraepelin considered the course of a disorder as a diagnostic criterion too. This has not been adopted by the DSM, but it is described in the manual.

> The duration of the disorder may be lifelong, though periods of remission lasting from weeks to years may occur. In most cases, the severity, frequency, disruptiveness, and variability of the symptoms diminish during adolescence and adulthood. In other cases, the symptoms actually disappear entirely, usually by early adulthood. In a few cases, the symptoms may worsen in adulthood. The predictors of this course are not known (*Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition*, 2008:Tourette's Disorder, Course).

A more detailed description of the course is given by Gilles de la Tourette. But unlike the DSM, Gilles de la Tourette included in it the emergence of coprolalia and mentioned the

urge to tic. Since very little of his writing has been translated into English, I bring here most of the Meige and Feindel citation of his description of the course of the disorder:

> About the age of seven or eight a little boy or girl... begins to exhibit a series of tics… the twitches are limited preferably to the facial musculature. At this stage, too, expiratory laryngeal noises are occasionally superadded.
>
> The movements may be confined for a long time to the face, but under the influence of causes very difficult to determine they gradually invade the shoulders and the arms. First one shoulder is shrugged and then the other, then the trunk is inclined *en masse* to right or left; then the patients waives his hands or his arms, or bends backwards and forwards, or jumps up and down. Flexing the knees alternately and tapping with his feet. The muscles of the larynx sometimes participate in the abnormal functioning, when it is that many sufferers from tic give vent to quick expiratory "hems" and "ahs" which coincide often with the twitches of trunk and limbs.
>
> The disease may be limited to this stage, but it is not uncommon to find, a few months or years after the beginning of the facial movements, that the inarticulate laryngeal sound becomes organized and develops in a particular direction, thus, is a sense, showing a pathognomonic value. Under the influence of cause whose actions we are, in the majority of cases, powerless to appreciate, the patient gives vent one day to a word or short phrase of a quite special character, in as much as its meaning is always obscene. These words and phrases are exclaimed in a loud voice, without any attempt to restraint. There must be a complete absence of the moral sense where there is coprolalia such as this[17]; at the moment of the ejaculation some irresistible physical impulse must drive the patient to utter filthy words unreservedly and with no consideration for other people.
>
> Another psychical stigma – echolalia – is occasionally, though less frequently, observed in these cases. (cited in Meige & Feindel, 1907/1990:224).

[17] A somewhat different understanding of coprolalia is discussed in the chapter "Symbolic tics."

Meige and Feindel used the course to differentiate among tics. Following Brissaud, they distinguished the disease of Gilles de la Tourette syndrome from chorea by its "uniformity of its symptomatology and *the regularity of its evolution*" (228, my italics).

The onset of Tourette syndrome

Another differentiation is by the onset of the disorder. The onset time of Tourette syndrome is a diagnostic criterion in the DSM-IV-TR. While it sounds like a clear and simple criterion, it is not. Nothing in TS is simple. To begin with, the age 18 of the DSM-IV, which is for most purposes the legal switch from adolescence to adulthood in the USA, is not universally accepted. For example, the British psychiatrist Mary Robertson sets the limit for TS onset at the age of 21 (2000). And even though according to many statistics, the most common onset is between the ages of 2 and 7 years (for example, see James F Leckman, Bloch, King, & Scahill, 2006; James F Leckman, King , & Cohen, 1999; Mary M. Robertson, 2000), these determinations typically neglect the complexity of the disorder, its comorbidities, and its etiology. How are these factors play into determining the time of onset? The two examples below demonstrate.

Donald Cohen and colleagues (1982) present the case of Mark, who at the age of 3 was in non-stop motion. He was thought to have ADHD and needed medication at the age of 4. A few months later he started with eye blinking, followed later by other motor and vocal tics. In school, Mark demonstrated behavioral difficulties and failed to learn basic arithmetic and reading skills. When was the onset of Mark's TS? If ADHD is a disorder comorbid with TS but separate from it, the age of almost 5, when his eye blinking started, was the onset time of Mark's TS. But if ADHD and TS are aspects of

one, more complex disorder, the onset of Mark's TS was at the age of 3, when the first signs of his ADHD showed up.

Another example comes from one of the participants in my study. On November 6th, 2006, in the hospital room where the 16-year-old Daniel recovered from his deep brain stimulation (DBS), in which two electrodes were implanted in his brain, his mother told me, "The surfacing of his Tourette's was at the age of 7, with mild tics that seemed rather like bad habits." But, she added, "I noticed from birth that he was different. He would not fall asleep unless he put his fingernails under mine tightly." Was Daniel's onset at the very beginning of his life when he behaved as a "different baby" with no tics but with atypical way of being lulled to sleep; or was his TS's onset only when it "surfaced" with mild tics at the age of seven? Moreover, for Daniel, his mother added, "a bad turn happened at the third grade after severe chicken pox, and he has never been the same." Was his onset then at the age of 7, when the mild tics might have been only "bad habits" or at the third grade when after his chicken pox "he has never been the same?"

While TS often runs in families, there are no robust findings of its genetic origins. Even studies of monozygotic twins have shown that much is left for interaction with the environment (Shapiro & Shapiro, 1981). At what age does such interaction happen and what is its nature? Many with TS, like Daniel with the chicken pox, report that the onset of their TS was preceded by an infectious disease. If indeed the infection triggers the TS in a person with genetic disposition, how does the gene-environment interaction depend on the person's age? Is there a time-window for the gene-environment interaction that enables the TS or brings it up?

Prevalence of Tourette syndrome

Any statistics of the prevalence of TS in the population is burdened with the possibility of ascertainment bias. Among adults, many mild cases, in which interference of the tics with everyday and social life is manageable, may never make it to the doctor. The same might be true for the extremely severe cases. If, for example, the severity of a person's signs leads to unemployment and no health insurance , the person may never visit the costly doctor and will be missed by the statistics Those who are afflicted with more than one disorder, might also bias the statistics. For example, people who suffer from OCD or ADHD in addition to their TS, might be diagnosed as having OCD or ADHD rather than TS. Even when diagnosed with TS, they are often excluded from TS studies for the sake of reliability. For adults then both ends of the severity spectrum of TS are often excluded from the statistics.

By contrast, studies of children and adolescents can rely on school population, and are therefore more likely to capture complete TS samples and conduct comparisons with well matched control population. In a 2005 review paper, Harvey Singer reported the prevalence of TS among children and adolescents to be between .1%-3% (2005). Higher figures of 5% were reported by David E. Comings and colleagues in 1990 (Cited in H. I. Kushner, 1999:4). Mason and colleagues (Mason, Banerjee, Eapen, Zeitlin, & Robertson, 1998) examined the whole population of 9th graders in a randomly selected high school in West Essex, UK, and found that 5 out of 166 students (2.9%) satisfied the DSM-III-R criteria for TS.

Out-of-school "[o]nly one population-based study of TS has been published." (Tanner, 2005:401). This is the study of population in Rochester, Minnesota, which took

more than a decade to complete, and was published in 1982 in *Advanced Neurology*. In it, AR Lucas and colleagues identified 4.6 people with TS per1 million. But this study was based on population that sought medical help; and the public awareness of TS was still low. The estimated numbers have grown greatly since.

How little studied the adult population is can be learned from the telling example of the Index of the 2005 handbook of Tourette syndrome, which has many entries for children and adolescents, but none for adults (Kurlan, 2005). And this is even though Ruth Bruun and Cathy Budman comment in their paper in this very volume that a few decades ago TS was considered a lifelong disorder. Shapiro too considers TS to be a lifelong disorder (1988). Singer and Walkup (1991) report that "one third of cases will remit completely during late adolescence or early adulthood, another third will show a significant improvement (in both amount and severity) of tics, whereas the remaining third will continue to be symptomatic throughout early adulthood and middle age."

One statistics is quite established. TS is distributed unevenly between the sexes. It afflicts more males than females and the ratio reported in the literature is usually between 3:1-4:1 . A higher ratio of 4.3:1 male:female is reported in 2000 by Roger Freeman and colleagues based on their study of 3500 individuals with TS in 22 different countries (Roger D Freeman et al., 2000).

Not all tics are signs of a disorder. Tics are quite common among school-age children and affect 12%-24%. Most are isolated or transitory. Today the number of TS population (as opposed to tic population) is believed to range between 4% and 6% (Scahill, Williams, Schwab-Stone, Applegate, & Leckman, 2006). This large range, suggests Kushner (1999), is due to the fact that each phenotype is unique.

Even though most of the studies cited above are from the USA, TS appears to be distributed worldwide and is reported in studies from the US, Europe, Japan, China, Hong Kong, the Middle East, and Brazil (Tanner, 2005). Comparisons across countries, however, are difficult to draw due to different methodologies and the effect of culture on the symptomology of the disorder, but can be revealing and important in understanding the interactions between Tourette syndrome and culture.

The familial nature of Tourette syndrome

As mentioned above, Charcot and Gilles de la Tourette already observed the "hereditary" nature of TS based on familial pedigrees. Since then a great attention has been paid to pedigrees. Meige and Feindel listed many neurologists who included pedigrees in their diagnoses and patient records (1907/1990:98). They also noticed that often in the same family there is a rich variety of people with pathologies – ticcers, hysterics, alcoholics, epileptics, etc. This observation is shared by the geneticist David Comings (Comings, 2001)[18].

Studying family trees is challenging. To begin with it is difficult to interview all family members (Rosario-Campos & Pauls, 2005:366). But even when this is overcome, the interviews must ask questions whose answers are apt to be concealed, like "Uncle Ben's alcoholism and wife abuse" (Comings, 2001:30). Then there always remains the difficulty of deciding whether certain behaviors are learned in the family or inherited.

[18] David Coming's work is controversial in the TS community. In a "Letter to the Editor," the Yale group of Pauls, Cohen, Kidd, and Leckman strongly criticizes Comings, who includes in the TS spectrum not only ADHD and OCD but also conduct-disorders, manic-depression, sleep-disorders, schizoid-behavior, and phobias. Their criticism questions the statistical quality of his studies and ends with the harsh words, "All of them [the statistical weaknesses of Coming's studies] make it impossible to accept as valid any of the results presented and raise serious concerns regarding the integrity of the peer review process for these papers." (D. Pauls, Cohen, Kidd, & Leckman, 1988).

The former challenge of data gathering was first met in 1986 by DL Pauls and colleagues (cited in D. Pauls, Raymond, Leckman, & Stevenson, 1991), who interviewed all first degree relatives of 32 TS probands. In 1991, Pauls and colleagues designed a study that included, in addition to the biological first-degree relatives of the TS probands, a control group of non-biological first-degree relatives of adopted TS probands (D. Pauls et al., 1991). In these two populations of first-degree relatives they documented TS, chronic tics (CT), and OCD. For age-corrected rate, their findings are:

B. Age-corrected Rates

Diagnosis	Biological Relatives of TS Probands	Relatives of Control Probands	*P*
TS	.087 ± .016	0	.02
CT	.173 ± .021	.027 ± .027	.004
OCD	.115 ± .019	.025 ± .025	.05

All of the rates are significantly higher in the biological relatives of TS, CT, and OCD probands than in the relatives of controls. These results support the hypothesis that TS, CT, and OCD are familial. Yet the possibility of a learning effect cannot be removed.

Twin studies have further supported the hypothesis about the genetic nature of TS. In 1985 Price and colleagues carried out the largest twin study. They mailed out 8000 questionnaires to registered TS patients and found for 43 same-sex pairs the following results. The identical twins concordance for TS is significantly higher than for fraternal twins, suggesting a genetic contribution to the disorder.

	Identical twins	**Fraternal twins**
Number	30	13
Concordant for TS	53%	8%

In 1987, Pauls and Leckman reported that they could not find any identical twins with only one having TS while the other having no evidence of tics. (cited in Comings, 2001:46). But again, as Comings observes, identical twins are more likely to share environments than fraternal twins. And even though the twin studies are highly suggestive of genetic etiology, we should notice also that the identical twins differ, leaving room for the role of the interaction with the environment.

Based on segregation analysis[19], the most wide spread hypothesis is that a single major locus best explains the patterns of transmission of TS in the family (Comings, 2001; D. L. Pauls, Alsobrook, Gelernter, & Leckman, 1999). But some suggest that more than one gene involved, while others suggest additive genetic effects through carriers whose phenotype carries no abnormalities (Barr, 2005; Rosario-Campos & Pauls, 2005).

Recent studies attempt to deal with the elusive nature of the disorder's phenotype. The few pedigree and genetic studies that have been conducted in the last decade divide each cohort by phenotype (Mary M. Robertson, Althoff, Hafez, & Pauls, 2008). For example, Alsobrook and Pauls (2002) used symptom-based factor analysis to identify four significant factors among 85 probands diagnosed with TS: 1) aggressive phenomena (e.g., kicking, temper fits, argumentativeness), 2) purely motor and phonic tic symptoms, 3) compulsive phenomena (e.g., touching of others or objects, repetitive speech, throat clearing), and 4) tapping and absence of grunting. These four factors accounted for 61% of the phenotypic symptom variance in the disorder's probands and their first-degree relatives. Three of these factors may indicate the presence of heritable components of the

[19] In *segregation analysis* pedigree data are heuristically analyzed against Mendelian theory to suggest a best fitting for dominant, recessive, or multifactorial inheritance.

Tourette's disorder phenotype. The correlation between the proband and relative factor scores was significant for three of the factors, all but factor 2 of purely motor and phonic tics symptoms.

As for the genetic loci, there are mainly two methodologies: (i) *Linkage analysis* attempts to identify a DNA marker[20] that is inherited significantly more often in affected family members than in unaffected members. Different studies have differed in their phenotypes and have identified different chromosome locations. There is no convergence yet of various studies with two exceptions. A region on the chromosome 2p32.2 has been identified both in sibling pairs and in large families of TS patients. And the chromosomal region 11q23-24 has been identified in TS patients in two independent studies (Leary, Reimschisel, & Singer, 2007).

(ii) Another approach associates specific allele polymorphism in TS patients and their family members and compares them to controls. This has not converged into conclusive results either. (iii) Nor have studies of chromosomal abnormalities led to conclusive findings yet. In addition, in all these genetic studies the causality is questioned since it is possible that epigenetics plays an important role (Leary et al., 2007).

In their discussion of the psychiatric disorder of schizophrenia, Levi Ledgerwood and his colleagues argue that evidence generally accepted as demonstrating genetic causation can be explained by hypotheses of infectious causation, while some of the evidence implicating infectious causation cannot be explained by genetic causation. Therefore, they argue, this asymmetry should lead to more studies exploring the possibility of infectious causation of psychiatric disorders. (Ledgerwood, Ewald, &

[20] A DNA marker is a DNA sequence of "letters," representing a DNA molecule or strand, with a known location on a chromosome and associated with a particular gene or trait.

Cochran, 2003). Indeed, we noted above that for Sydenham's chorea such causation has been established. The field of PANDAS studies pediatric population and investigates the possibility of their infectious causation for TS. However, not much progress has been done in this field and I will not discus this topic further.

Tourette syndrome and the brain

The triumph of the organic understanding of TS is linked mainly to the names of the Shapiro's who were convinced that all psychiatric disorders, including TS, were organic rather than psychogenic. Prior to them, already in the 1950's and 1960's some practitioners reported their success in treating tics, coprolalia, and echolalia with pharmacological and surgical interventions. The organic interpretation of TS was not limited to TS. Many other psychiatric disorders started to be treated with various levels of success with drugs and psychosurgeries (H. I. Kushner, 1999). Due to the elusive nature of psychiatric disorders, many of the new treatments were and still are administered in a trial-and-error fashion, in which the patients are not only the beneficiaries but often the test cases, the guinea pigs.

The clinical treatments themselves took part in what Elliot Valenstein calls *the war of the soups and the sparks*, meaning the chemical (soups) and the electrical (sparks) (Valenstein, 1988, 2005). It is not a war anymore and no one today doubts that both electrical and chemical messaging operate in the brain. Neither of them, though, is completely understood. But there is enough progress that guides interventions. The chemical interventions are in the form of pharmacological drugs aiming at the synaptic connections, with their neurotransmitters and receptors. The electrical intervention is

through the insertion of electrodes to the brain, like a pace maker. The procedure is known as deep brain stimulation (DBS).

The regions targeted for TS by both these treatments are mainly in the basal ganglia (BG). However, it is likely that rather than mere regions, the effect is on circuits. Indeed, the electrodes of DBS are sometimes placed in a thalamic region, rather than a BG region, to affect the circuit. The understanding of the BG, their neurotransmitters, and their various receptors relies mainly on animal models. But the implication of the basal ganglia (BG) in TS derives mainly from responses of patients to psycho-pharmacology and psychosurgery and from postmortem neuroanatomy studies.

The basal ganglia and cortico-basal ganglia-thalamocortical circuits

The basal ganglia (BG) are a group of subcortical nuclei, which includes four nuclei: The striatum (which consists of the caudate nucleus, the nucleus accumbens, and the putamen), the globus pallidus (with two parts – internal GPi, and external GPe), the substantia nigra (with two parts – compact SNc, and reticular SNr), and the subthalamic nucleus (STN). Figure 3.1. is a coronal (horizontal) slice showing the BG.

The BG are the principal subcortical components of a family of parallel circuits, illustrated in figure 3.2. Informed by animal studies, Alexander, DeLong, and Strick describe a model of these circuits as "discrete, essentially non-overlapping parts of the striatum, globus pallidus, substantia nigra, thalamus, and cortex." (1986, p. 359). The circuits are named after their paths cortico-striatal-thalamo-cortical (CSTC). Four circuits have been identified by functionality (T. Wichmann & M. DeLong, 2006): Motor,

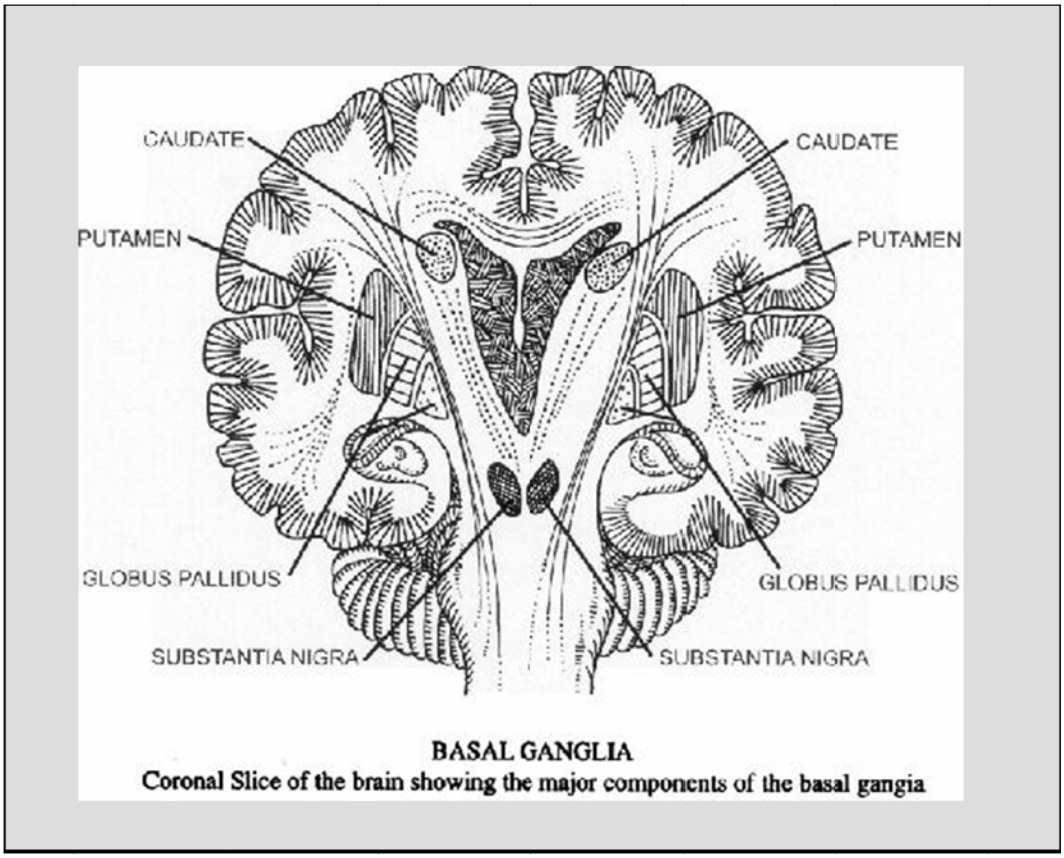

Figure 3.1[21] A coronal slice showing the basal ganglia

oculomotor[22], prefrontal, and limbic. Grossly, each passes through the cortex, the striatum, the pallidum and sunstantia nigra, then the thalamus, and back to the cortex. However, thanks to the somatotopic organization[23] of many brain regions, the specifity of the target within each region is much finer.

The messaging between the cortex and the BG, and the cortex and the thalamus are excitatory and glutamatergic. The inhibitory connections in the CSTC are

[21] From (H. Kushner, I., 1999)
[22] Oculomotor – relating to the movements of the eyeball
[23] Somatotopic organization in the brain respects the spatial relationships of the body. For example, in a somatotopic map of the hand, the brain region representing the pinky is next to the region representing the ring finger. However, the size allocated for each does not reflect the physical size of the body part, but rather its use. For example, in the motor cortex, the space allocated for the representation of the leg is smaller than that allocated for the hand.

GABAergic[24] (T. Wichmann & M. R. DeLong, 2006). But important connections inside the BG are dopaminergic. Their receptors are of two kinds. D1 receptors are the gate

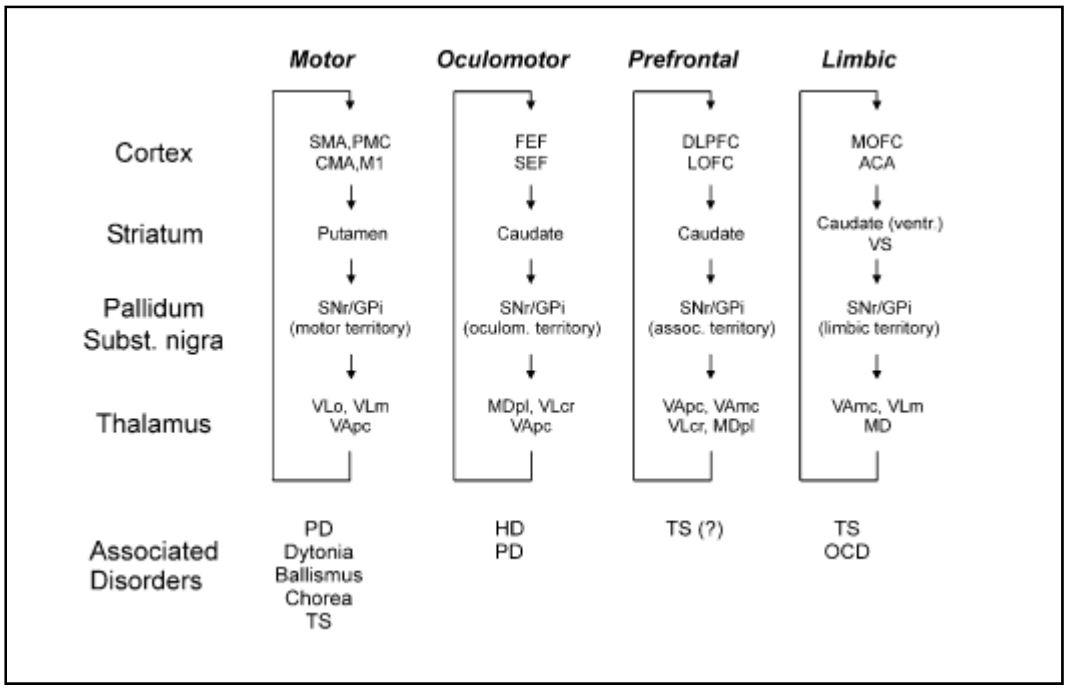

Figure 3.2[25] Parallel cortico-striatal-thalamo-cortical circuits. **Abbreviations:** ACA, anterior cingulate area; CM, centromedian nucleus of thalamus; CMA, cingulate motor area; DLPFC, dorsolateral prefrontal cortex; FEF, frontal eye fields; HD, Huntington's disease; LOFC, lateral orbitofrontal cortex; M1, primary motor cortex; MDpl, mediodorsal nucleus of thalamus, pars lateralis; MOFC, medial orbitofrontal cortex; Pf, parafascicular nucleus of the thalamus; PMC, premotor cortex; SMA, supplementary motor area; SEF, supplementary eye field; VApc, ventral anterior nucleus of thalamus, pars parvocellularis; VAmc, ventral anterior nucleus of thalamus, pars magnocellularis; VLm, ventrolateral nucleus of thalamus, pars medialis; VLo, ventrolateral nucleus of thalamus, pars oralis; VLcr, ventrolateral nucleus of thalamus

keepers of a direct pathway that facilitates movements. Its activation disinhibits the thalamus thereby increases thalamo-cortical activity. The D2 is inhibitory and enters an indirect pathway. But even though it is inhibitory, the final results of D1 and D2 and their respective pathways are the same in that they both lead to reducing inhibition of the

[24] The common abbreviation for μ-aminobutyric acid is GABA
[25] From (T. Wichmann & M. R. DeLong, 2006).

thalamo-cortical neurons and thus facilitate movements initiated in the cortex (DeLong, 2000). Figure 3.3 illustrates the dopaminergic circuits of the BG.

TS and the morphology of the BG

Postmortem studies have found abnormalities in the caudate and the putamen. A 1956 postmortem study by K. Balthazar found these regions of the basal ganglia of a TS patient's brain to be hypoplastic. In other words, the size and packing density of the neurons in the putamen and caudate nuclei resembled those of young infants. Another postmortem study of four adult TS patients reported in 1991 by Singer and colleagues found increased numbers of pre-synaptic dopamine-uptake sites in the caudate and the putamen. In addition a 1992 study by Anderson and colleagues found in these four cases reduced glutamate levels in the projection areas of the STN -- GPi, GPe, and SNr.

In-vivo structural brain imaging found morphological abnormalities as well. While for right-handed people, the left BG, is volumetrically bigger than the right, Peterson and colleagues found in 1993 that for 14 TS right-handed patients, this asymmetry was reduced, especially in the GP (B. S. Peterson et al., 1999). But none of these studies have been translated into interventions.

TS and neuropharmacology

The beginning of psychopharmacology as we know it today is marked by the discovery of chlorpromazine (Thorazine) by a French team headed by Henri Laborit. As with other psychiatric drugs, the studies of the chlorpromazine were carried on psychiatric patients, sometimes with devastating effects. This is a deeply disturbing aspect of the history of the development of psychiatric drugs. Even though

chlorpromazine and the whole group of phenothiazines, to which chlorpromazine belongs, were first investigated as synthetic dyes, it was observed that chlorpromazine could act as antihistamine. It was first administered by Laborit as analgesic for surgical patients but was noticed in the late 1950's as producing Parkinsonian symptoms -- rigid movements, opposite from those of ticcers (H. I. Kushner, 1999; Valenstein, 1988). Even though its use greatly increased, psychiatrists qualified their praise of it. The dominant paradigm was still psychoanalytic. But marketing by Smith Kline & French and positive reports about the effects on psychiatric patients generated voluminous sales of Thorazine. In 1955 The First International Colloquium on Chlorpromazine and Neuroleptics Drugs was held in Paris. The effectiveness of chlorpromazine was established in the US with a 1961 six-week study conducted by the National Institute of Mental Health (NIMH). Chlorpromazine and two other phenothiazines were compared with placebo treatments in nine hospitals. 75% of the psychiatric patients were "much improved" or "very much improved" and 50% were judged as no longer even "mildly ill." None was reported to have become worse after the drug treatment, which is in great disagreement with later findings of many adverse side effects like parkinsonism, dyskinesia, and motor restlessness. But the 1964 publication of the NIMH study with its endorsement of chlorpromazine caused a wide adoption of it for treating psychiatric disorders (H. I. Kushner, 1999:133).

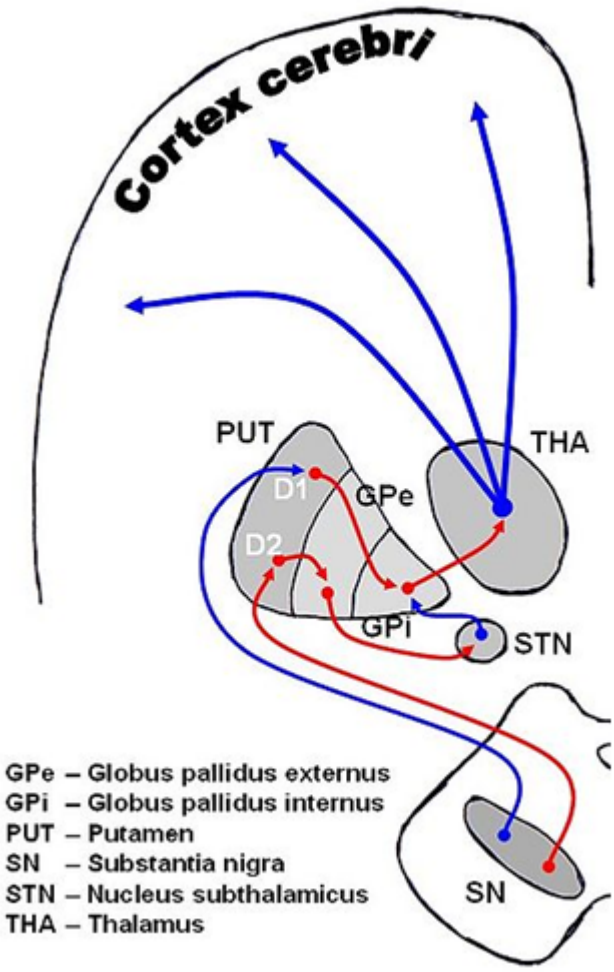

Figure 3.3[26] An illustration of the dopaminergic pathways in the BG in one hemisphere. Red is inhibitory, blue is excitatory.

The positive effect of chlorpromazine on TS, and the conclusion from it that TS is organic and is due to brain lesions, was published in 1962 by two Polish psychiatrists, Roman Dolmierski and Maria Kloss. In an article entitled "de la Maladie de Gilles de la Tourette" they reported of two (out of five) patients who were treated successfully with chlorpromazine in their clinic in Gdansk. A 12-year-old boy with multiple motor tics, coprolalia, and echolalia was placed on 225 mg chlorpromazine per day and all his severe

[26] From http://en.wikipedia.org/wiki/Parkinson%27s_disease

signs were reduced to eyebrow movements, slight facial grimaces, and slight muscle movements of the shoulders and neck. A 55-year-old woman with neck and shoulder tics, coprolalia, and obsessive symptoms was put on 100 mg of chlorpromazine per day. Her symptoms were not eliminated but were controlled. In both cases, when the drug was withdrawn the tics reappeared. Thus, concluded Dolmierski and Kloss, TS is most likely organic because "it is known that chlorpromazine acts on the spinal cord and on the sub-cortical neurons and that in high doses results in Parkinsonian symptoms" (Meyer & Quenzer, 2005).

At the time, it was not understood how chlorpromazine was working. Today, though, we know that it inhibits dopamine transmission throughout the brain. By contrast, haloperidol (Haldol), a modification of chlorpromazine, is selective in its action and inhibits dopamine transmission only in the D2 receptors (Valenstein, 1988). Based on an animal model that was used to test potential antipsychotic drugs, haloperidol was found to be much more effective than chlorpromazine in blocking amphetamine-induces stereotyped behavior in animals (H. I. Kushner, 1999:135).

The haloperidol's success with TS was first published in 1961. The French psychiatrist M. Jean N. Seignot reported a 35-year-old man who underwent every possible treatment for his *maladie des tics*, including lobotomy[27], with no effect. His violent and self-injurious motor tics as well as his vocal tics and coprolalia were intense and frequent. His only relief came from .6 mg per day of haloperidol, with "remarkable" and immediate results. There was a great reduction in his tic intensity and frequency, and his coprolalia disappeared completely. Similarly good reports came from the psychiatrists

[27] In a lobotomy, some brain fibers, typically fibers that connect the frontal lobe of the cortex to a subcortical region, are surgically severed to relieve some mental disorders.

Georges Challas and William Brauer of Iowa State University. In 1963 they published a report about the effective results of administering 1.5 mg of haloperidol per day to a 16-year-old male with motor tics and coprolalia and to a 10-year-old boy, also with motor tics and coprolalia. The haloperidol was successful in both cases but had to be interrupted and reinstituted for the 16-year-old depending on his liver toleration of the drug. (H. I. Kushner, 1999:138-42).

There were more reports of case studies. The case of a 13-year-old female was reported by the neurologist Janice R. Stevens and the psychiatrist Paul H. Blachly of the University of Oregon Medical School. The patient who had intense and frequent motor tics and hissing tics had undergone behavioral, psychological, chlorpromazine, and other drug therapies. All to no avail. But 2.5 mg four times daily of haloperidol greatly decreased her motor tics and hissing, and 3 mg four times daily caused them to entirely disappear. When unknown to her, the haloperidol was replaced with a placebo, her tics returned, then again disappeared after returning to the drug. Stevens and Blachly were persuaded that the motor tics and the hissing were "organic." Moreover, they speculated that due to the chemical similarity of haloperidol to the inhibitory neurotransmitter GABA it decreases dopamine transmission in the brain's basal ganglia. More reports of successful treatments of TS with haloperidol arrived from Europe (Shapiro et al., 1988:327). Whether the excessive dopaminergic activity is due to the receptors or the production of dopamine is unclear. But in either case, the excess would increase the thalamic disinhibition and therefore would lead to hyperstimulation of the frontal cortex (Leary et al., 2007).

A few studies have suggested the contribution of serotonin to the abnormality of the CSTC in TS. They include studies that show abnormally low level of 5-HIAA[28] in the cerebrospinal fluid (CSF); postmortem studies of TS patients showing decreased concentration of 5-HIAA in the BG; and blood and urine analyses showing low levels of serotonin in TS patients (Leary et al., 2007). Serotonin re-uptake inhibitors[29] are often prescribed to OCD patients and serotonin abnormality is a strong hypothesis in the understanding of OCD (Alvarenga, Hounie, Mercadante, Miguel, & Rosario, 2007; Geller, 2007). Later I will speak about the question of whether OCD is comorbid with TS or belonging to the same spectrum.

Since haloperidol, a whole host of pharmacological drugs for tic suppression have been developed. Table 3.1 gives a list of antipsychotic medications (but not all of them) used for tic suppression. They are divided into typical and atypical. The typical are hypothesized to block the post synaptic dopamine D2 receptors. But even though they are efficacious in reducing tics, they have many adverse side effects and are used less now than in the past. Atypical neuroleptics have a combined affinity to the excitatory receptors for serotonin, 5HT-2, and the D2 receptors. They have less side effects and there is growing evidence to their efficacy in tic suppression. When the tics are understood as autoimmune-based resulting from infection, they are not treated with neuroleptics but instead with penicillin (Harrison, Schneider, & Walkup, 2007).

[28] 5-HIAA is the principal serotonin metabolite.
[29] Reuptake is the absorption of a neurotransmitter by the pre-synaptic neuron. This is a "clean up" process, which removes the remaining neurotransmitter that has not passed to the post-synaptic neurons, from the synaptic cleft. Blocking this process increases the availability of the neurotransmitter to the post-synaptic receptors.

Generic name	Brand name	Dosage (mg/day)	Side effects
Typical neuroleptics			
Haloperdiol	Halidol	1-5	Sedation, weight gain, extrapyramidal symptoms[30]
Pimozede	Orap	2-8	QTc prolongation[31] , sedation, extrapyramidal symptoms
Fluphenazine	Prolixin	1.5-10	Sedation, weight gain, extrapyramidal symptoms
Atypical neuroleptics			
Risperidone	Risperidol	1-3	Sedation, weight gain, elevated prolactin[32]
Olanzapine	Zyprexa	5-10	Sedation, weight gain
Quetiapine	Seroquel	200-500	Sedation, weight gain
Aripripazole	Abililfy	10-20	Insomnia, Akathesia, tremor

Table 3.1[33] Antypsychotic medications used for tic suppression

As has been repeatedly mentioned, one of the greatest challenges of TS is its heterogeneity. Clinically complex patients might not have a robust response to only one medication. They might however benefit from a combination of medications. Such combination requires a series of ongoing trials with each patient in search for the optimal solution. It requires a strong support system of family-patient-physician relations. It also requires a knowledgeable and up-to-date psychiatrist. As the number of medications increase, the psychiatrist is challenged to know their effects and possible side effects. Meeting all the mentioned requirements is difficult and not meeting them adds to the risk when experimenting with a patient to identify the optimal treatment (Harrison et al., 2007).

[30] The extrapyramidal system is part of the motor system involved in the coordination of movements. Extrapyramidal symptoms are symptoms resulted from affecting the extrapyramidal system and are by themselves various movement disorders. They are often side effects of antipsychotic drugs and include repetitive, involuntary muscle movements, urge to be moving constantly,

[31] QT interval is the time between the start of the Q wave and the end of the T wave in the heart's electrical cycle. QTc is prolongated QT and is associated with increased risk of sudden death.

[32] Prolactin is a peptide hormone primarily associated with lactation.

[33] Table from (Harrison et al., 2007)

TS and neurosurgery

Some extremely severe patients are refractory and do not enjoy any amelioration of symptoms despite all treatments. These patients are sometimes treated surgically. In 1949 the Portuguese neurologist Antonio Egas Moniz won the Nobel prize in physiology and medicine "for his discovery of the therapeutic value of leukotomy in certain psychoses" (*Nobel prize in physiology or medicine*, 1949). The prefrontal leukotomy introduced by Moniz in 1936 severs the white matter connecting the front lobes to subcortical regions. It was applied mainly to schizophrenia patients. The procedure was modified by the American neurologists Walter Freeman and James Watts and the word *leukotomy* was replaced by *lobotomy*, since the American procedure encompassed not only the destruction of neuronal tracts in the white matter, but also the destruction of actual neuronal cells in the brain. In the 1940s, Freeman streamlined the procedure, replacing it with transorbital lobotomy, in which a picklike instrument was forced through the back of the eye sockets to pierce the thin bone that separates the eye sockets from the frontal lobes. The pick's point was then inserted into the frontal lobes, where the connections between the lobes were severed. Freeman performed this procedure very quickly, sometimes in less than 10 minutes (Pressman, 1998).

In 1955 Watts performed the first transorbital lobotomy on a refractory TS patient. The goal of the surgery was to sever the connections between the prefrontal lobes and the deeper limbic structures. The limbic structures, considered as the "emotional brain," would eliminate the emotional outbursts of some psychiatric disorders. After a two-year follow up of the 37-year-old male patient, Watts found a "decrease in the frequency, duration, and amplitude of the motor tics and the compulsive swearing" (cited

in H. I. Kushner, 1999:128-9). But in 1981 the German child psychiatrists Uta Asam and W. Karrass published a follow up study of 16 Gilles de la Tourette patients who had undergone psychosurgeries. According to their report, good outcomes were reported only if the follow up time was short. Otherwise the two psychiatrists warned against "severe side effects" (cited in H. I. Kushner, 1999:130).

Today ablative approaches have been replaced by the non-destructive and reversible high frequency electrical deep brain stimulation (DBS) (T. Wichmann & M. R. DeLong, 2006). In DBS, electrodes are inserted to a target location, which depends on the disorder, and send electrical impulses. This is why some call these electrodes *brain pacemaker*. The tuning of the electrodes (voltage and frequency) is done through an external battery-operated controller, and the parameters are determined individually by an iterative error-and-trial process. The procedure has been approved in the US by the food and drug administration (FDA) for Parkinson Disease but is still considered experimental for TS.

The theory supporting the practice of DBS emphasizes circuits, not regions. And based on imaging and the phenotypes of TS, the limbic and the motor circuits are targeted (See figure 3.2). On these circuits a few targets have been tried in the pallidus, the thalamus, the subthalamic nucleus, and the pontine nucleus of the brain stem. They are starred with greater specificity in figure 3.4. The populations selected for DBS is typically refractory and adult. The published studies report more than 70% reduction in vocal or motor tics with disappearance of the urge that often precedes TS tics (T. Wichmann & M. R. DeLong, 2006).

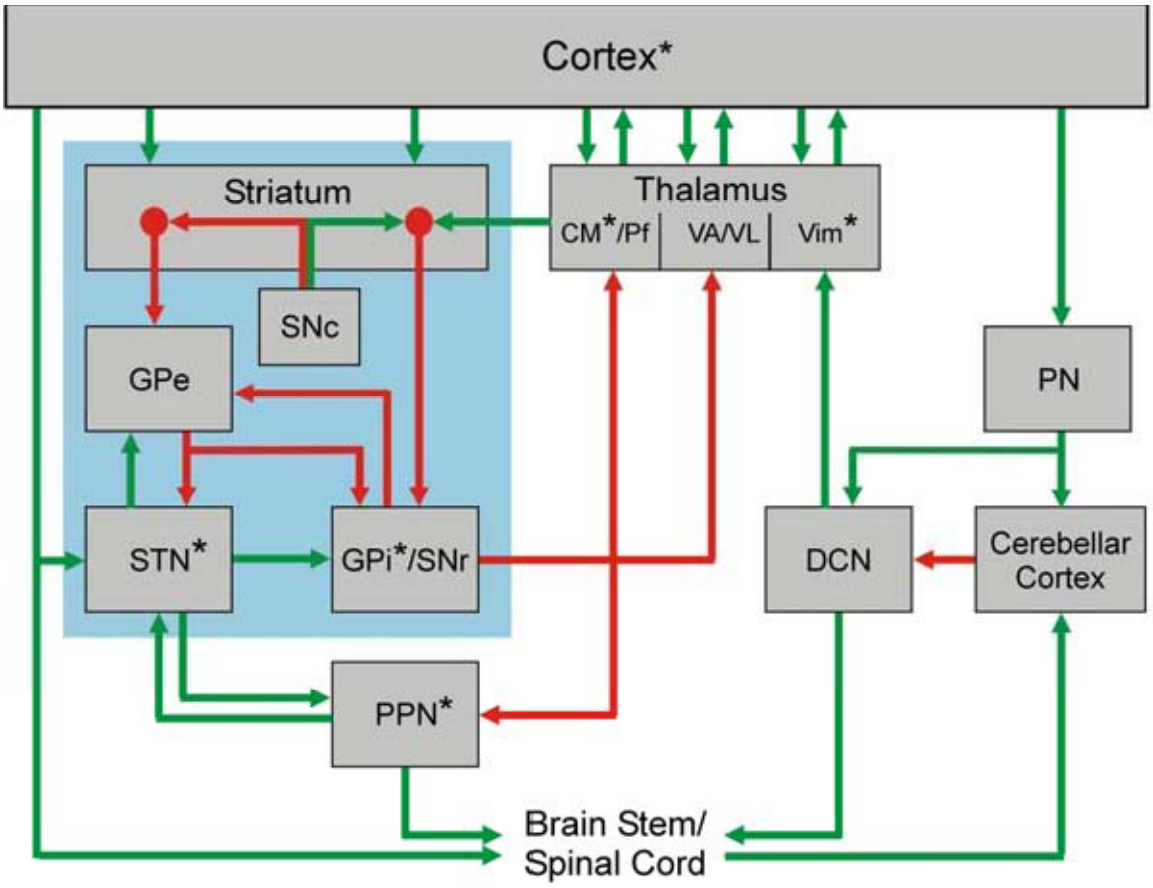

Figure 3.4[34]. Intrinsic anatomy of Cortico-Subcortical Circuits. Inhibitory (GABAergic) connections are shown as red arrows, and excitatory (glutamatergic) connections are shown as green arrows. The basal ganglia are shown in the blue rectangle. Common and experimental DBS targets are marked with asterisks. PN, pontine nuclei. For other abbreviations, see legend to Figure 1 and text.

TS and the cortex

Investigating the cortical contribution to TS has been possible in the last two decades with positron emission tomography (PET) and functional magnetic resonance imaging (fMRI). In a PET study, Braun and colleagues compared 16 adult TS patients with controls and found *decreased* metabolic rates of the TS probands in the orbitofrontal regions, insula, parahippocami, ventral striatum, and midbrain. *Increased* metabolic activity was observed in the supplement motor area (SMA), lateral premotor area, and

[34] Figure 3.4 is from (T. Wichmann & M. R. DeLong, 2006)

primary motor cortex. A later study with more severe phenotypes and OCD showed increased metabolism in the orbitofrontal regions (reported in Butler, Stern, & Silbersweig, 2006).

There are a few fMRI studies investigating motor tics, the urge, and tic suppression. Due to the involuntary nature of tics and the limited control over them, it is difficult to have an experimental paradigm which is ecologically valid. But with this caveat, some fMRI studies further support the role of the BG in TS and the participation of the CSTC circuits in the disorder. In addition studies converge to show abnormal activation of the premotor and the motor cortices (Butler et al., 2006). But as with chemical and electric treatments, so in imaging, we are still far from having a coherent picture either about the very nature of the disorder or about its treatments. As in phenotype, so in brain imaging, no two TS patients are alike.

The return of psychosocial approaches

Some "non-pharmacological interventions" have made their comeback. Their very name points to the supremacy of the organic, but this supremacy does not exclude other kinds of interventions (A. Peterson, 2007), and can be applied in conjunction with them. One goal of the psychosocial approach is to decrease the social and psychological impact on the patient. The target of this approach is not the patient as much as his or her human environment. Educating the family and, for children, their classmates can increase social acceptance and therefore decrease the suffering that comes from social mockery and rejection (Conners). I will speak later about how the attitude of families and friends affect the participants of my study.

Another goal is to reduce and ameliorate the tics. The most common approach towards it is *habit reversal* (HR). It includes various components designed to increase awareness of self and the environment, increase relaxation, and get skilled at *competing response* (A. Peterson, 2007). The latter, which is also the major component of HR, seems like a revival of the behavioral therapy that was administered by Meige and Feindel, with some success. In competing response the individual responds to an urge with a movement that disrupts the tic, or applies deep breathing to reduce rhythmic eye movements or vocal tics. I will return to psychosocial therapy in the context of my own study. I will especially highlight the role of the sense of agency and active participation of the patient in such treatments.

Psychosocial treatments are challenged like all other interventions by the great variability among individuals. In addition there is no metric to measure their efficacy. The current guidelines of the clinical psychology division of the American Psychological Association determine the efficacy of a certain treatment as "well established" or "probably efficacious," by the number and type of publications that support them. According to this criterion, HR would be classified as "probably efficacious."

TOURETTE SYNDROME AND OTHER PSYCHIATRIC DISORDERS

The complexity of TS increases when considering its comorbidities (Shytle & Wilkinson, 2007:48). Frequently TS comes with other disorders, notably obsessive compulsive disorder (OCD), attention deficit disorder (ADD), attention deficit hyperactivity disorder (ADHD), and anxiety disorder (AD). In my study I concentrated on the comorbidity of OCD. This is reflected in my theoretical considerations as well as in the data that I collected from interviews, patient self-reports, and doctor evaluations of

tics, obsessions, and compulsions. In exploring the nature of TS, I ask with many others whether OCD is a separate disorder, comorbid with TS, or whether it is in the same spectrum that makes TS a spectrum disorder?

The same question is also asked by clinicians and scientists who treat and study OCD. But they ask the mirror question, in which OCD defines a spectrum and the question is whether TS belongs to this spectrum. A paper titled "Obsessive-compulsive spectrum disorder" includes a section "Tourette's disorder and OCD" and examines the comorbidity of OCD and TS (Moore, Mariaskin, March, & Franklin, 2007:25). In this paper the studied patients are those who had been diagnosed with OCD. TS is their comorbidity. A similar OCD-centered approach is expressed in another article in the same OCD handbook, with a section, "Comorbidity: Tic disorders and Tourette's syndrome" (Berrios, 1996:153 n46).

The graphical representation of Venn diagrams brings greater symmetry. In it the comorbidities are neither TS-centered, nor OCD-centered; being atemporal, the graph does not inform which diagnosis came first. It only shows areas of overlapping or separateness. An example of a Venn diagram conceptually presenting possible relations between TS, OCD, and ADHD is in Fig. 3.5. The overlapping regions represent cases with more than one diagnosis – some have TS and OCD, others TS and ADHD, others yet ADHD and OCD, and some have them all; while the non-overlapping regions stand for cases with only one diagnosis -- only TS, only OCD, or only ADHD. The shared regions in the Venn diagram that represent overlapping of comorbidities can be also considered for the possibility of shared neurological substrata, similar interactions with the environment, or shared genetic causes.

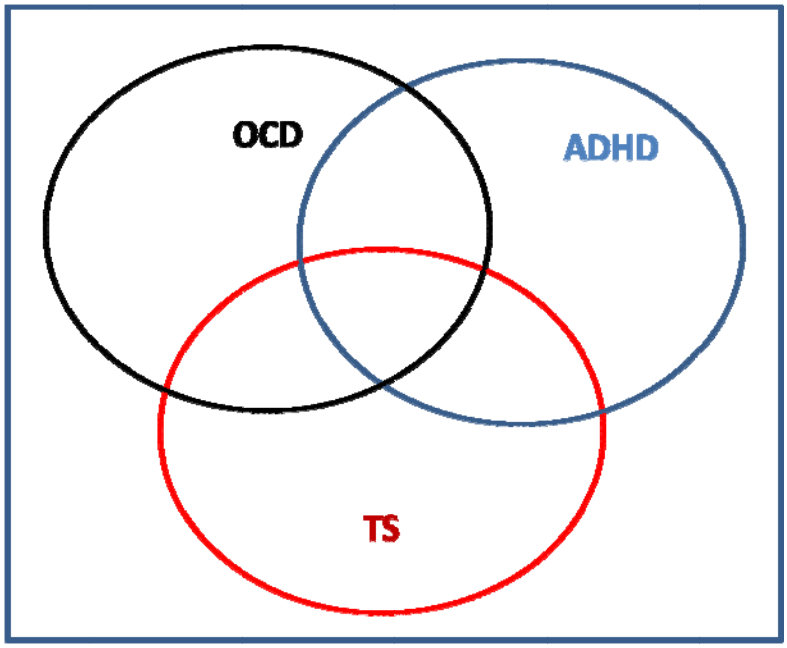

Figure 3.5: Comorbidity

Obsessive-compulsive disorder

There are some historical similarities in the studies of OCD and TS. Like Gilles de la Tourette, a student of Jean-Martin Charcot the chief physician of Salpêtrière Hospital in Paris, so was Jean-Etienne Dominique Esquirol (1772-1840) half a century earlier, a student of Phillipe Pinel (1745-1826) the chief physician of the Salpêtrière from 1795 to his death at 1826. And as the Marquise de Dampiere was the emblematic case for Gilles de la Tourette's *maladie des tics* , Mademoiselle F. was the emblematic case for Esquirol's *monomania*. Mademoiselle F. was seen by Esquirol when she was 34 years old. Her illness started at the age of 18 when she started to fear that she might take in her pockets objects belonging to her aunt. She was also afraid that when touching money, some of it might stick to her fingers. She started to fear that her hands would touch things. When this happened she rubbed them against her clothes as to get rid of something. She frequently washed her hands and as her signs progressed, she had to be

fed by her servant since her fear of touching included food. (H. I. Kushner, 1999). She described her thoughts as "irresistible" even though she had the "insight" that they were not hers (143). Esquirol "oscillated between explaining her complaint as a disorder of ideas or of the will" (143).

Georges Guinon, a Salpêtrière neurologist at the time of Jean-Martin Charcot and Gilles de la Tourette, considered *maladie des tics* as part of what we call today a spectrum disorder; and obsessive compulsive disorder, according to Guinon, belonged to this spectrum (for a historical overview of OCD see Alvarenga et al., 2007). From then on, disagreements continued about the relation, or lack of it, between TS and OCD. Also, along with the *zeitgeist* and as for TS, the presumed etiologies of OCD fluctuated between psychological and biological.

Meige and Feindel, citing Charcot that tics are "psychical disease in a physical guise", considered obsessions as related to TS and reasoned that "[t]he frequency with which obsessions, or at least a proclivity for them, and tics are associated, cannot be a simple coincidence" (Meige & Feindel, 1907/1990:82). In their book, they never use the word "compulsion," but briefly discuss "impulsive tics" and refer to a "typical example of Tourette's disease… described by Köster as 'disease of impulsive tics'" (227).

Margaret Mahler grouped compulsions and tics in contradistinction from impulsions, which she defined as "instances in which the ego condones objectionable motor release with little or no inner conflict" (Mahler, 1944:434). For Arthur Shapiro tics were organic movements unlike the compulsive movements of OCD. And he proposed that "some Tourette's disorder symptoms resembling OCS[35] be referred to as impulsions"

[35] OCS – obsessive compulsive symptoms

(Shapiro et al., 1988:220). He differentiated impulsions from the tics of TS since "they are intended rather than involuntary" (493). But even though he acknowledged that 10%-20% TS patients have such impulsions, and that this number is significantly higher than in the general population, he considered them as separate and objected to linking TS, OCD, and impulsions to one spectrum. This is much like Charcot who on one hand grouped tics and compulsions in the same "psychiatry insanity" and considered them motorically and ideationally analogous, yet on the other hand insisted that they were different and unrelated (H. I. Kushner, Luzzatti, & Finger, 1999). Currently "impulsions" are not to be found in the DSM-IV-TR, but most agree that impulsions have a voluntary, intentional, aspect while compulsions are irresistible. The definitions of obsessions and compulsions are given in the manual.

Current definition of OCD

Obsessions are defined in the DSM-IV-TR as

> recurrent and persistent thoughts, impulses, or images that are experienced, at some time during the disturbance, as intrusive and inappropriate and that cause marked anxiety or distress; the thoughts, impulses, or images are not simply excessive worries about real-life problems; the person attempts to ignore or suppress such thoughts, impulses, or images, or to neutralize them with some other thought or action; the person recognizes that the obsessional thoughts, impulses, or images are a product of his or her own mind (not imposed from without as in thought insertion).

Compulsions are defined as

> repetitive behaviors (e.g., hand washing, ordering, checking) or mental acts (e.g., praying, counting, repeating words silently) that the person feels driven to perform in response to an obsession, or according to rules that must be applied rigidly; the behaviors or mental acts are aimed at preventing or reducing distress

> or preventing some dreaded event or situation; however, these behaviors or mental acts either are not connected in a realistic way with what they are designed to neutralize or prevent or are clearly excessive.

In the DSM-IV-TR, OCD and TS are considered as being often comorbid, but they are different and belong to different groups. OCD is in the Anxiety Disorders group and TS is in the group of Disorders Usually First Diagnosed in Infancy, Childhood, or Adolescence. But in the Differential Diagnosis section of TS, the manual acknowledges the phenotypic resemblance and the TS-OCD comorbidity. Moreover, the manual acknowledges that "[c]omplex tics may be difficult to distinguish from compulsions… making this distinction is all the more challenging because Obsessive-Compulsive Disorder is common in individuals with Tic Disorders."

The manual further differentiates between tics and compulsions:

> Tics... and stereotyped movements … must be distinguished from compulsions. A *tic* is a sudden, rapid, recurrent, nonrhythmic stereotyped motor movement or vocalization (e.g., eye blinking, tongue protrusion, throat clearing). A *stereotyped movement* is a repetitive, seemingly driven nonfunctional motor behavior (e.g., head banging, body rocking, self-biting). In contrast to a compulsion, tics and stereotyped movements are typically less complex and are not aimed at neutralizing an obsession. Some individuals manifest symptoms of both Obsessive-Compulsive Disorder and a Tic Disorder (especially Tourette's Disorder), and both diagnoses may be warranted.

In the chapter "Tic classification" I will further compare tics and compulsions.

> According to the DSM-IV-TR, 35%-50% of TS patients suffer from OCD.

However, the reverse documents only 5%-7% of OCD children as having TS, even though 20%-30% have reported current or past tics. This is in disagreement with the

review of Moore and colleagues that cites 80% comorbidity of tics disorder and TS in children with OCD (Moore et al., 2007).

Familial studies support etiological link between TS and OCD

The rate of OCD in first-degree biological relatives of individuals with TS is significantly higher than that in the general population. The significant level is such that many, especially in the Yale Family Study data led by James Leckman, consider OCD as a variant expression of TS (Barr, 2005; Rosario-Campos & Pauls, 2005). Interestingly, in contradistinction to TS, OCD by itself appears to occur more frequently in female than in male relatives of TS probands (D. Pauls et al., 1991).

Self Injurious Behavior (SIB)

The repetitive nature of self-injurious behavior makes it a good candidate for OCD. However, not much attention has been paid to this highly penalizing behavior. Those who speak of self-injurious *compulsion* often consider it as an OCD phenomenon. Others include it in TS. The British neurologists Mary Robertson and Michael Orth observe that self-mutilations "often have an obsessional quality to them" (2006:55). Nonetheless, Robertson and Orth link SIB to TS, and define SIB as "self-injurious behaviors encountered in people with TS" (55). The 2007 *Handbook of child and adolescent obsessive-compulsive disorder* has only one mention of SIB, and it is under the section "Tourette disorder and OCD" (Shytle & Wilkinson, 2007:49).

Armando Favazza defines self-mutilation as "the deliberate destruction or alteration of one's body tissue without conscious suicidal intent" (Favazza, 1996:xviii). And Robertson and Orth too distinguish between SIB and suicide. While suicide is "intentional self-inflicted death" (2006:54), SIB "on the whole [is] not associated with

desire to die, rather merely to inflict injury" (55). Whether this is indeed the case deserves further investigations. In her review "Self-injurious behavior as a separate clinical syndrome," Jennifer Muehlenkamp cites statistics according to which 28%-41% of people who reported SIB, had attempted suicide (Muehlenkamp, 2005).

In a summary of six studies, 225 out of 862, 26% of TS patients, had SIB. The individual studies showed a range of 15%-53%, and the largest study with 555 TS patients had 34% with SIB (Mary M Robertson & Orth, 2006:55). Jennifer Muehlenkamp observes that there is evidence suggesting that the incidence of SIB is increasing among adolescents and college students. This observation is linked to a question that was raised earlier in this chapter about the definition of a disorder. How to interpret this rise in numbers? Are there behaviors that are currently labeled as SIB, but were not considered SIB in the past? In addition there is the question of ascertainment. How do we access and identify the SIB population for the purpose of computing the statistics of prevalence? And finally, the question of whether SIB is within the TS spectrum, or a separate disorder often comorbid with TS, is still debated (Muehlenkamp, 2005).

Sub-categories of TS

While the search for a spectrum is driven by the phenomenon of comorbidity, a reverse perspective has led some to further divide the TS spectrum into sub-syndromes. Some argue in favor of SIB being a separate disorder; maybe a subtype of OCD or TS. In their trade book about Tourette syndrome, Simon Baron-Cohen and Mary Robertson consider dividing the spectrum of TS. They ask, "Is there more than one type of Tourette syndrome?" (Baron-Cohen & Robertson, 1998:25). And suggest subdividing TS into three categories: (i) "Pure" TS with predominantly motor and vocal tics; (ii) "full blown

TS," which includes coprophenomena, echophenomena, and paliphenomena; and (iii) "TS plus," which has comorbidities like OCD or ADHD (Mary M. Robertson, 2000).

Such spectral analysis is far from being trivial. "A major difficulty… is the inability to definitely diagnose a true case [of TS]" (Tanner, 2005:400). Interestingly, to overcome this difficulty, Towbin and his colleagues recommend diagnosing TS by its *symptoms* rather than its *signs* -- the premonitory sensations that precede many tics (1999). And a similar suggestion is expressed by Neal Swerdlow (2005).

Joseph Jankovic (1997) proposes a new classification for TS. He distinguishes between Definite Tourette syndrome and two types of Probable Tourette syndrome. His Definite TS requires frequent motor and phonic tics for at least one year and before the age of 21. The course of the disorder further contributes and needs to present dynamic change in the patterns and anatomical locations of the tics. Jankovic's Type 1 Probable TS relaxes the onset and dynamical requirements; and his Type 2 Probable TS relaxes the requirement of both multiple and motor and phonic tics and accepts instead either single motor tic with phonic tics or multiple motor tics with possible phonic tics. He also defines some tics as compulsive, a topic to which I will return in the chapter "Tic classification."

Adults with Tourette syndrome

Another possible subtyping of TS might be by the course of the disorder. For a majority of afflicted children tics and vocalizations wane as they grow into adulthood (James F Leckman et al., 2006). In a histogram combining two studies, one with 36 probands and one with 46, Leckman and colleagues present the worst ever tics between the ages of 8 and 12; no tics at the age of 17; and almost no tics at the age of 18. In a

large study across Africa, Asia, Australia, Europe, the Middle East, North America, and South America, Roger Freeman and colleagues created a database with 6805 probands who suffer from Tourette syndrome. They found out significant *decline* with age in the comorbidity of ADHD and in the signs of sleep problems and anger control. However, they found significant *increase* with age of OCD, mood disorders, anxiety disorders, coprolalia and copropraxia, SIB, and mental retardation (Roger D. Freeman, 2007). Similarly, Leckman and Cohen observe that "Paradoxically, although tics usually diminish in frequency and intensity by early adulthood, the most severely affected individuals are adults whose symptoms may include self-injurious motor tics (e.g., being blinded by a detached retina secondary to hitting tics), coprolalia (shouting obscenities), and markedly disinhibited speech and behavior" (James F. Leckman & Cohen, 1999:159).

In agreement with the findings or Freeman and the observations of Leckman and Cohen, my pilot study suggests that those whose signs and symptoms continue into adulthood have the most florid clinical presentations, including coprolalia and copropraxia, SIB, and OCD. This is the least studied population, and for it neither medications nor other interventions prove effective over time. Their signs and symptoms are debilitating and isolating. Moreover, because the focus of diagnosis and treatment centers around the pediatric population, adults with florid TS (ATS)[36] find only few clinicians who are able to treat them. In addition, given that many of this population have difficulty in sustaining employment, they rely on Medicaid which further limits their access to treatment. Treating ATS is also frustrating for the clinician. As medications

[36] I use ATS for "adults with Tourette syndrome" and for "Adult Tourette syndrome," depending on context.

generally prove ineffective, patients often turn to addictive substances for self-medications, and, as a result, treatments get complicated and success is elusive (H. I. Kushner, 2008b).

My pilot study of 16 ATS suggests that there are unique challenges for this group, not shared with others. Their phenotypes and challenges will be discussed in the chapter "Tic classification." I consider subtyping ATS as *adults who suffer from TS and OCD*. Of the 16 ATS in my study 100% suffer, in addition to their TS, from OCD, even though OCD was not a selection criterion for the study. A correlation between the severity of TS's signs and symptoms and the presence of OCD has been also found in an adult study by (Cath et al., 2000). In this study, four groups were studied: Probands with TS and OCD (14), with TS only (18), with tic-free OCD (21), and controls (29). The signs and symptoms were ranked by their severity, which was measured by the time per day consumed and by the amount of distress and interference induced by each item. The items were: aggression, sexual thoughts, contamination, rumination, mental play, echophenomena, washing, checking, counting, symmetry, touching, reading, impulsive behavior, praying, and miscellaneous. Three levels were defined: symptomatic repetitive behavior, subthreshold symptoms, and just being present. The distinction among obsessions, compulsions, and impulsions was guided by the presence or lack of anxiety. Obsessions and compulsions were defined as accompanied with anxiety, unlike impulsions, which were not accompanied with anxiety. Obsessions were found to be the most distressing and interfering. Phenomenologically, those with TS and OCD were found to be more similar to TS than to OCD. And Cath and colleagues concluded that

these differences "possibly represent differences in underlying pathophysiology between Tourette's and tic-free OCD" (2000:505).

There are other differences: In the chapter "Tic Classification," I will propose a few classifications of tics by various criteria. One of them is the trigger of the tics. Some tic triggers are *external*, as simple as light; others are as complex as social situations. By contrast, many obsessions that lead to compulsive behavior are *internal*, self-generated. Dark thoughts, what the neurologist Jorge Juncos calls "the alien within" (private communication), are both triggered and carried out internally.

Compulsions are observables expressed in the external world. In my pilot study, touching every third car, counting over and over to 5, and establishing symmetry were quite common. By contrast, the internally-triggered excessive need for cleanliness expressed in compulsive hand-washing is rare or non-existing among TS+OCD patients. as is reported in the studies of Cath and colleagues (Cath et al., 2001).

The OCD of ATS might differ from the usual OCD also in relations to anxiety. In OCD, it is understood that anxiety drives the obsessions and compulsions. In ATS, it is possible that anxiety is the outcome of the obsessions and compulsions. This agrees with the clinical intuition of Juncos, based on treating many ATS (private communication). This is also in agreement with the 2000 study of Cath and colleagues, according to which people with TS and OCD have more impulsive, anxiety free, behavior. In their 2001 study Cath and colleagues compared the phenotype of TS+OCD, ticless OCD, TS without OCD, and controls. They found more echophenomena, trichotillomania (hair pulling), SIB, and touching and symmetry behavior among TS+OCD than among ticless OCD (2001).

Unlike in the 2000 Cath et al study, in which only 36% of the TS+OCD patients, 28% of the TS-only patients, and 19% of the tic-free-OCD patients were on neuroleptic or antidepressant, in my study 14 out of 16 participants, 87.5%, are on medications. This can explain a greater symptomatic severity in my study. On the other hand, some adults, who can manage their TS, choose, like two in my own study, to stay away from medications and their side effects altogether.

The possibility of age-based subtyping has been raised for OCD too. But, unlike with ATS, in which the proposed subtyping relies on the *course* of the disorder, for OCD the proposed subtyping is by *onset time*. It has been observed that even though the clinical presentations of child and adolescent onsets are similar to those with adult onset, the sex distribution is different. For childhood and adolescence onset, there is, like in TS, male predominance with 2.5-3.5:1 ratio. For adult onset, the sex distribution is equal according to some studies or with female predominance in others. Those with early-onset OCD are considered to be most likely linked to TS (see review by Eichstedt & Arnold, 2001).

The ATS subtyping by the course of TS will hopefully prove to be productive for a population, which suffers from severe presentations, and is often marginalized, hardly studied, struggles to be employed, and socially isolated.

CONCLUSION

Since the time of Gilles de la Tourette, Tourette's has undergone different constructions. Its very phenotype has changed. Milder cases were added while the severe signs including coprolalia and echolalia have been pushed away, often excluded from research. The population with severe phenotype is often marginalized and without

adequate treatment. For lack of research dedicated to ATS, its percentage is unknown. But many adults, who continue to suffer from the pediatric disorder of TS, or TS+OCD, are with severe phenotypes.

The century-old questions about the etiologies of TS are still not answered. But some progress has been made. Genetic, neurophysiological, and neurochemical avenues have been explored, as well as behavioral and psychological ones. Progress often depends on iterative trial-and-error interventions, in which new theories and interventions are derived from the responses of patients to treatments.

While there is a wide consensus about the role of excessive dopamine in TS and the implication of the basal ganglia in the disorder, its origins are still debated. Unlike with the clinical definition of TS, in research each study defines its own phenotype. There is no convergence of genetic studies even though pedigree studies strongly point to the hereditary nature of the disorder. The contribution of infection, especially the streptococcus bacterium, is still debated. The contribution of epigenetic influences is not understood and hardly explored.

As TS is a spectrum disorder, it touches on other disorders, especially OCD and ADHD. Are they comorbid with it or do they all define one complex spectrum? Or maybe subtyping is a way to gain a better understanding of the disorder? Discussing TS in the singular as though it is one homogenous entity is misleading. With its spectral nature and different phenotypes, each patient calls for an individual study. Indeed we are reminded by Hughlings Jackson to not fall into the trap of thinking that there is some entity of which TS is the proper name.

METHODOLOGIES

CHAPTER CONTENTS

IN THIS CHAPTER I describe my methodologies, procedures, data sources, and data analyses. The information is necessary for a critique of my work, but not for understanding it.

Every study of humans suffers from the tension between qualitative and quantitative approaches. Qualitative studies are informed by human interactions between those who study and those who are studied. They are often described with words and are holistic presentations of personal narratives. They are simultaneously enriched and biased by the subjective interpretation of the researcher. Quantitative studies seek the commonalities among people and use the language of numbers. They typically focus on specific aspects of people, be it a specific trait, a specific brain region, the response to a specific medication, etc.

Each highlights some perspectives and neglects others. For example, in studying TS, a quantitative study that compares a certain medication to a placebo treatment would lead to a pharmacological therapy, if there is a significant difference in favor of the medication. However, the great variability in phenotypes might prove this specific therapy useless or even harmful for a certain individual. A careful attention to the phenotype of this individual might lead to the realization that his or her suffering requires different intervention altogether. The cultural context of the individual can play a role too. While dark thoughts might be exorcized successfully in one culture, they would be better treated with a message from the ancestors in another culture, or with a new drug that had been identified in a series of quantitative studies (Kleinman, 1988).

To capture the complexity of a syndrome, both the quantitative and the qualitative are needed. This is well articulated by the neurologist VP Ramachandran in his

discussion of how to study the brain: "A tension exists in neurology between those who believe that the most valuable lessons about the brain can be learned from statistical analyses involving large numbers of patients and those who believe that doing the right kind of experiments on the right patients – even a single patient – can yield much more useful information" (Ramachandran & Blakeslee, 1998: xii). The resolution, he observes, "is obvious: It's a good idea to begin with experiments on single cases and then to confirm the findings through studies of additional patients." (xii). In my study, Ramachandran's "single cases" are the phenotypes, and they are what I have studied.

I am carrying out a multi-phase study, of which the dissertation is the first one. This phase is a qualitative study whose data were gathered in video-recorded interviews with adult TS patients (ATS) and their family members or friends; and from questionnaires. The outcomes of this phase include insights and hypotheses of neurological, cultural, linguistic, and psychological nature about Tourette syndrome. Testing these hypotheses quantitatively on large populations will be sketched in the last chapter "Future directions."

THE CURRENT STUDY

Qualitative studies, like this one, are not exactly repeatable. However, some of the planning and technical details of my study can be of use in similar studies. I therefore share them here.

IRB approval

This research was approved by a full board committee of the Emory IRB on 6/19/2007, and was assigned the number IRB00002397.

The participants and their recruitment

The sixteen TS patients of this study were recruited as a *purposive sample* (Bernard, 2002: 186). That is, they were not randomly selected. Instead, they met the criteria of DSM-IV for the diagnosis of Tourette syndrome (TS), and were individually identified by Dr. Jorge Juncos[37] to meet the study's criteria and goals. Fourteen of them brought another close person -- referred to as *partner* – who was an immediate relative or a close friend of many years. The rationale for having the partner was to provide an additional subjective perspective of an outsider who belongs to the patient's immediate social and cultural environment and cares enough to share observations and folk theories about the patient's condition. No financial compensation was offered to the participants.

Out of the sixteen TS patients, thirteen were male. This sex distribution reflected the distribution in the total TS population. The patient ages ranged between 18- 65 years, and averaged 39 years. The participants included representatives from a wide severity spectrum, from simple motor and vocal tics to coprolalia, copropraxia, and self mutilation.

All the participants but two are under medical care and being treated with medications. This simultaneously informed the study about how they responded to their medications, and masked the presentation of the un-intervened disorder. Table 4.1 summarizes the profiles of the sixteen TS participants.

[37] Dr Jorge Juncos is an associate professor of neurology at the School of Medicine of Emory University, and a neurologist and psychiatrist in the Clinic of Movement Disorders of Emory University. He is a member of the Medical Board of the Tourette Syndrome Association.

#	Participant	Name[38]	Sex	Age	Medications	DBS[39]	Partner	Date of interview
01	SRA	Steven	M	19	X		X	9/28/07
02	LRA	Lionel	M	65				11/07/07
03	TQA	Ted	M	41	X		X	8/06/07
04	KGA	Kyle	M	25	X		X	9/21/07
05	DQA	Dylan	M	29	X	X	X	8/01/07
-6	HBA	Henry	M	59	X		X	7/20/07
07	SZA	Stuart	M	29	X		X	10/26/07
08	EEA	Elliot	M	37	X		X	8/13/07
09	CBA	Chuck	M	40	X		X	8/24/07
10	NFA	Neil	M	50			X	8/22/07
11	DLA	Danielle	F	31	X		X	8/17/07
12	NYA	Nick	M	55	X			12/17/07
13	CFA	Claire	F	45	X		X	12/21/07
14	DGA	Donna	F	43	X		X	2/08/08
15	LLA	Louis	M	39	X		X	5/30/08
16	DGB	Daniel	M	18	X	X	X	6/04/08

Table 4.1 The profiles of the participants

The procedure

After their successful recruitment, the patient and the partner came for one visit at the Emory Clinic for Movement Disorders. I explained the study to them and gave them the opportunity to ask questions. Two consent forms were signed, one by the patient and one by the partner. In addition, the patient signed the HIPAA form. After that, Dr. Juncos observed the patient and filled in the YTGSS and the YBOCS medical evaluation forms. Then the patient filled in two self reports -- about tics, and about obsessions and compulsions. The questionnaires and the interviews will be described in greater detail later.

[38] The participant names are fictitious to protect their identity
[39] DBS stand for the procedure of deep brain stimulation

The patient and the partner were then interviewed sequentially and separately by me, in video-recorded interviews. Most of the interviews lasted one hour, none longer; and all lasted only as long as the participant was comfortable. This completed the interaction with the patient and the partner. The cameraman, Rob Poh of Georgia State University, and I stayed briefly after each interview to critique the interview and learn from its strengths and weaknesses and improve the following ones.

Setting and equipment

All the interviews were conducted in the same room at the Emory Clinic for Movement Disorders. Three people were present in each interview: The participant, Finkelstein the interviewer, and Poh the cameraman.

The semi-transparent curtains on the windows were pulled down to control the light, and a few blankets were hung as drapes to soften the sound. A vase of flowers was provided to counter the medical atmosphere of the room. The camera was focused on the participant for the whole interview, and was positioned for maximal view of the face, torso, and arms. Most of the time, the camera was stationary, except when the participant demonstrated leg, knee, or foot tics that the cameraman judged as worth capturing.

The camera was a Canon XL-2 with 20x Zoom lens. Subjects were shot from 10 feet away. Lighting was two standard overhead ceiling fixtures each with 4 daylight fluorescent tubes. Audio was recorded simultaneously with the Canon's on-board audio, using the standard Canon front-mounted stereo microphone.

The video and audio were recorded onto MiniDV digital video tape. Video and audio were captured and edited using Final Cut Pro 5 running on a Quad G5 Power Mac. DVDs were produced on DVD Studio Pro 4, running on the same computer. Completed

DVDs were burned on DVD-R or DVD +R formatted disks. The video files were stored on a Western Digital My Book external Firewire hard drive.

Data Sources

Four kinds of data were collected in this study: Medical evaluations, self-reports, medical records, and interviews.

Medical evaluations

For 10 of the participants, Dr. Juncos filled in two semi-structured[40] medical observational reports. Both have earned high rates for reliability and validity. The questionnaires served two purposes: (i) to determine the form, frequency, and intensity of the tics; (ii) to find out if the patient, in addition to TS, also suffered from obsessive-compulsive disorder (OCD).

The *Yale Global Tic Severity Scale* (YGTSS)[41] is a second generation semi-structured instrument for the assessment of tic severity in children, adolescents, and adults. It was developed by the members of the Tic Disorders Clinic at Yale (Leckman et al., 1989) based on data collected from 105 subjects aged 5 to 51 years. The YGTSS provides an evaluation along five dimensions: number of tics, frequency, intensity, complexity, and interference of motor and phonic symptoms. For each of these five dimensions there is a six-point ordinal scale, in which 0 stands for no tics 5 for the most severe. The instrument yields three scores: total motor, total phonic, and total tic scores.

[40] Semi-structured interviews or questionnaires are conducted with a fairly open framework which allows for focused, conversational, two-way communication. They can be used to give and receive information (Patton, 2002).
[41] For the YGTSS evaluation form see Appendix III

It allows the observer to incorporate direct observations with historical information. And it requires medical training and experience with tic disorders.

Reliability of the YGTSS was measured by the group that developed it. The intraclass correlation coefficients for three raters and 20 subjects, with $p<0.0001$ are: 0.78 for total motor scores; 0.91 for total phonic scores; and 0.84 for total tic scores. For validity[42], the subscales scores of YGTSS were compared with the 1984 Tourette Syndrome Global Scale (TSGS). With $p<=0.0001$, the correlation was 0.86 for motor; 0.91 for phonic; and 0.10 for total tics.

The well-established *Yale-Brown Obsessive Compulsive Scale* (Y-BOCS)[43] is another important instrument (Goodman et al., 1989). Like the YGTSS, is a semi-structured instrument for measuring severity of tics. Y-BOCS has a five-item list for obsessions and a five-item list for compulsions. Each has a five-point scale 0-4, with 0 for not present and 4 for most symptomatic. It has slightly different versions for adults and for children and adolescents due to different symptomatology between these populations. Both versions yield a total obsession score (0-20), a total compulsion score (1-20), and a combined total (0-40). "Most investigators consider the Y-BOCS to be the best available instrument for rating obsessive-compulsive symptomatology." (Scahill, King, Schultz, & Leckman, 1999:315). And indeed, in the Goodman et al. 1989 study, based on 42 patients, the intraclass Pearson coefficient[44] among four raters, with

[42] As was discussed in the chapter "Tourette's – a syndrome not a disease," validity is difficult to determine. Therefore, while the above study attests to the validity of YGTSS, it only confirms the fact that the YGTSS and the TSGS measure the same thing, with high probability.
[43] For the Y-BOCS evaluation form see Appendix IV
[44] Pearson coefficient measures correlation. When the measured correlation is among raters, it amounts to a measure of the instrument's reliability.

$p<0.0001$ was 0.97 for obsession subtotal; 0.96 for compulsion subtotal; and 0.98 for the total score.

Even though the above two instruments present their results in numbers, they are nominal; moreover they are qualitative. The data recorded in them are subjective, and while the data can be presented graphically and afford comparisons, YGTSS and the Y-BOCS should not be considered as objective quantitative instruments. Calling them "instruments" does not turn these evaluation forms into objective thermometers or odometers. Their authority and scientific value draw on their reliability.

The self reports

Each TS participant filled in two self-reports. The *Tic Symptom Self Report* (TSSR)[45] is a two-part questionnaire. The first part addresses motor tics and has 18 questions with an ordinal scale of the values 0-3: 0 = No symptoms at all this past week; 1= Tics were infrequent and nor forceful; 2 = Tics were frequent and forceful; and 3 = Tics were very frequent and very forceful. The second part addresses vocal tics with 16 questions, and with a similar ranking.

The Y-BOCS self evaluation[46], titled *Y-BOCS Compulsions Checklist*, is a two column checklist of compulsions – one for the past and one for the present. A checkmark in the appropriate column indicates the presence of the compulsion.

Like all self reports, the TTSR and Y-BOCS Checklist are subjective. Some past events have been forgotten; others that had never occurred are confabulated; some are self-denied; others get flourished. As with all autobiographical data, they tell us much

[45] For the TSSR questionnaire see Appendix I
[46] For the Y-BOCS self evaluation see Appendix II

about how the person understands himself or herself; or how the person selects to present herself or himself. Often the subjective self-image and self-presentation correlate with more objective data. But not always.

The medical records

Medical records are written by a medical practitioner, typically a physician or a registered nurse. In addition, the records include the prescribed medications, their dosage, and the patient's response to them. Occasionally they include some reflections of the physician about the patient in general and the medications and their effect. Medical records were available for 13 patients. In addition, the stereotactic coordinates and the electrode settings of Dylan and Daniel, the two participants who had undergone deep brain stimulations (DBS), were available too.

The data from the interviews

An interview gives the researcher the opportunity to observe signs of the disorder and hear from the patient about his or her *symptoms* -- the unobservable experiences. What the individual shares about his or her own perspective and subjective world is complex and depends on the quality of the individual's self reflection, honesty, and articulation. Therefore complementing self reports partner perspectives is valuable. In addition, if the partner is a parent, data from early age can be gathered too.

I used *topic guided interviews*[47] in a video-recorded, face-to-face setting, and in an informal and conversational style. While on its surface the interview took the form of a free-flow conversation, all the topics in the Interview Guide were covered. The free

[47] For the Interview Guides see Appendices VI-XI.

format allowed for the questions to be sensitive to the interviewee's attention, mood, and responses. It brought a sense of ease, trust, and openness, and in addition made room for the emergence of new topics that were not included in the original Guide. Unlike in structured interviews, the wording and the order of the questions have not been uniform. Moreover, since sometimes additional topics arose beyond the Guide, the interview's contents differed among individuals, never missing themes that were in the Guide, but allowing for additional ones.

The opportunity given to an individual to speak and be heard might have been an important motivation for the interviewees to participate in the study. As the participant Lionel complained, "The doctor never asked *me*." (Finkelstein et al., 2007-08-01). Some questions were hard and painful. When too embarrassed to answer orally, most agreed to answer in writing, on a pad which I offered.

The advantages of video-recording an interview are obvious. Nevertheless I will mention some explicitly. (i) The most obvious one is that tics, with varying modalities, frequency, duration, and intensity can never be faithfully documented in any other form. (ii) Video-recording liberates the interviewer from concerns of documenting the interview. Having only the responsibility of interacting with the interviewee, my presence was not compromised and I could be fully attuned to the other person and the nuances of our interaction. (iii) The DVD's have been visited by me over and over again. Unlike with notes, which select and screen what is documented at the moment of the note-taking, the full situation presents itself in each viewing, revealing each time new nuances and providing new insights. (iv) Finally, the danger of compromising the sense of trust and

safe space by having a third person in the room was avoided thanks to Poh's professionalism that made him "invisible" shortly after the beginning of the interview.

The camera was kept stationary for most of the time, and only on the interviewee. This served several purposes: (i) no tics were lost (ii) the TS restlessness was not mixed with a jittery camera, (iii) maybe most important is the minimal interpretation of the cameraman. With a relatively free-of-motion recording, the interpretation occurs during the data analysis. And finally, (iv) the video-recordings can, and hopefully will, be studied independently by others in the future. This is as close to objective documentation as possible.

Additional data sources

Some individuals with Tourette syndrome published their own stories (two such examples are Cohen & Wyscocky, 2005; Seligman & Hilkevich, 1992). They are revealing and moving, and I included some data and insights from them. I also expanded my exposure to TS patients through psychiatric reports of the present and the past. I have exposed myself to as many individuals and phenotypes as I could.

Data analyses and presentations

The data used in this study include: (i) quotations from the interviews, (ii) the answers to the questionnaires, and (iii) the medical-evaluations. Their use is self-explanatory throughout the dissertation. However, the temporal analysis of the video-recordings requires more explanation[48]:

[48] After I had determined the code and the fashions of analyses, much of the coding itself was performed by Jocelyn Taylor, who was trained in tic recognition by me.

Temporal coding of the interviews

The DVD's were played back with QuickTime 7.6.. The observations were recorded in Excel:mac 1. The platform for both the viewing and the recording was Macintosh X10.5.6. The time resolution was 1 second.

The codes[49] and their documentation

To document the behaviors observed in the DVD's, I first created a dictionary – a set of codes for the tics and their intensity. All motor tic codes start with "M." All vocal tic codes start with "V." The full code is presented in Table 4.2. The value of the intensity is 1 for a mild, 2 for a moderate, and 3 for a forceful tic.

The time axis of the Excel table was expressed by the rows. Each row represented 1 second. (e.g., 1 minute of an interview was recorded into a table with 60 rows; an interview of one hour was recorded into a table with 3600 rows.). The first three columns of the table recorded the passing time in an hours:minutes:seconds format. The fourth column recorded the passing time in seconds (e.g., 0 hour:10 min:7 sec would be recorded as 607 seconds). The fifth column recorded the motor tics using their codes, and the sixth column recorded the tic's intensity. Similarly, the seventh and eighth columns were used for the vocal tics and their intensity. The ninth column was used for comments (for an example of the Excel table of the first 14 seconds of the participant Steven, see Table 4.3).

[49] I thank Prof. Kim Wallen and Andrew Fischer for helpful discussions about the coding system.

Score		Motor		Vocal	
0	No symptoms	**MA**	Trouble starting actions	**VB**	Animal noises (barking)
1	Very weak	**MB**	Touching face	**VC**	Coughing
2	Moderate	**MC**	Touching part of the body	**VD**	Saying single words
3	Very forceful	**MD**	Shoulder jerks	**VE**	Repeating other's speech
		ME	Eye blinking	**VG**	Grunting
		MF	Other facial tics	**VH**	Bird noises (hooting)
		MG	Picking at things (clothing, etc.)	**VI**	Insults (lack of inhibition)
		MH	Head jerks	**VM**	Humming
		MI	Pressing hands and fingers	**VO**	Other vocal tics
		MJ	Touching objects	**VR**	Repeating own words/sentences
		MK	Knee clapping	**VS**	Saying single syllables
		ML	Kicking leg movements	**VT**	Throat clearing
		MM	Miscellaneous motor tics	**VU**	Gulping
		MN	Finger or hand movement	**VV**	Ch-sound (train like)
		MO	Touching other people	**VW**	Whistling
		MP	Finger or hand tapping	**VX**	Obscene words (coprolalia)
		MQ	Lip biting/ pressing lips	**VY**	Sniffing
		MR	Arm movements	**VZ**	Stutter
		MS	Stomach jerks		
		MT	Tensing parts of the body		
		MU	Hurting self		
		MV	Align head and neck with hands' aid		
		MX	Copropraxia		
		MY	Playing w/tongue		
		MZ	Swaying/ bending		

Table 4.2: Tic codes

In the few events where more than one tic appeared simultaneously, a greater resolution was used to tease the tics apart. The software Cut Pro 5 was used for this. Its resolution is over 29 frames per second. In other words, each second analyzed by CutPro was divided into units of less than 34 milliseconds. No other study, to the best of my knowledge, has coded 16 hours of TS behavior with such high resolution.

Presenting and analyzing the data

The temporal coding of the interviews provides a wealth of data that can be studied in various ways. A complete study of the co-occurrence of motor and vocal tics

rs:min:sec			Time in Secs	Motor		Vocal		Comments	1_SRA
0	0	0	0						9/8/2007
0	0	1	1						
0	0	2	2						
0	0	3	3						
0	0	4	4						
0	0	5	5						
0	0	6	6						
0	0	7	7						
0	0	8	8						
0	0	9	9						
0	0	10	10	MI	1			[I am not sure that it's a tic]	
0	0	11	11	MI	1	VY	3	[The sniffing is shorter than a…	
0	0	12	12	MI	1			[The sniffing is accompanied…	
0	0	13	13			VZ	1		
0	0	14	14						

Table 4.3: A sample of tic coding

was inspired by a few samples of stack graphs of the data. The full study and its results are described in chapter 6, in the section that classifies tics by their modality. The graphs were generated by SPSS-11 based on data that were exported from the spread sheet. The stack graph in Figure 4.1, for example, presents the first 96 seconds of Steven's interview[50]. The height of a bar represents the tic's intensity. Vocal tics are green and motor tics are red. When vocal and motor tics co-occur at the same second, the two bars are stacked. In the example of Figure 4.1 there is such an event only in second 10. Co-occurrence of vocal and motor tics appears to be rare.

To statistically test this impression I conducted two within-subject chi-square comparisons. One compares vocal and motor tic co-occurrence with random co-occurrence vocal and motor. The other compares the co-occurrence of tics with the co-occurrence of gesture and speech. The speech-gesture behavior was coded for duration of

[50] I thank Rob O'Reilly for technical support with SPSS.

10% of the total time of the interview and started in a randomly selected time. As discussed in chapter 6, most of the participants, showed significant difference of mixed-modalities between tics, and random expectations or linguistic behavior.

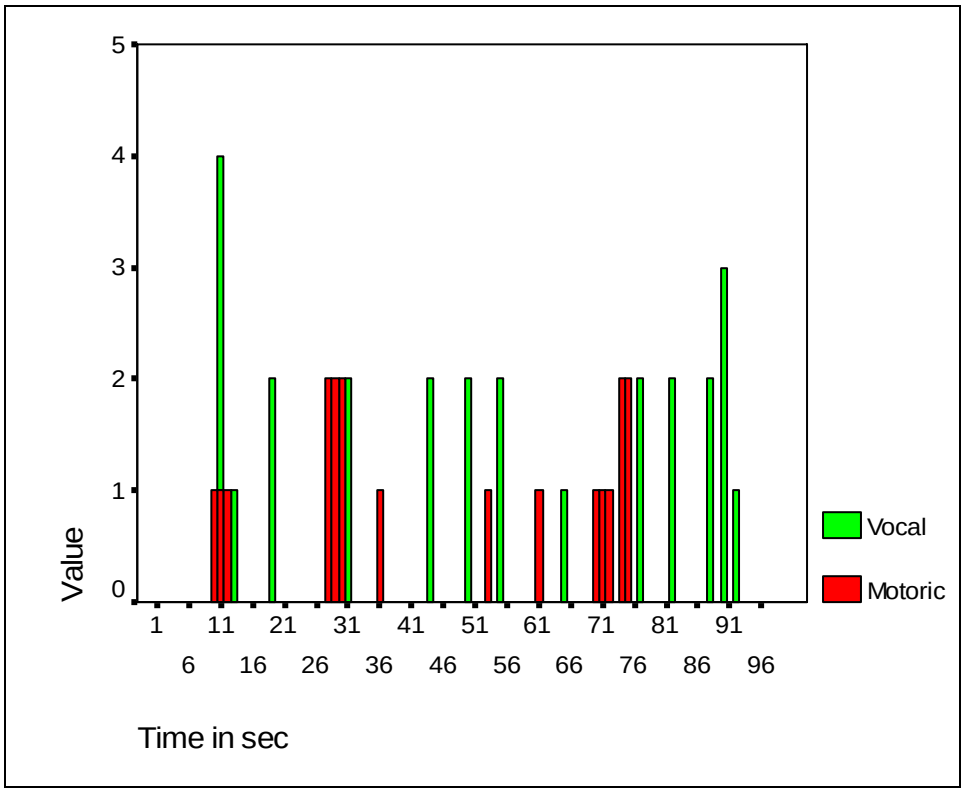

Figure 4.1: A graph of the first 96 seconds of SRA

More can be learned from this graphic presentation. We can notice a repeated patterns of 3 seconds of motor tics followed by one second of vocal tic, all at the same intensity, in the interval of (9-12), (28-31), and (74-77) seconds. I plan to further analyze these data in the future.

THE PARTICIPANTS: IN THEIR OWN WORDS

The account is purposely given nearly in the man's own words, so that the opinions expressed and the theoretical terms used are the patient's and not the physician's.

J. Hughlings Jackson in the "Case of convulsive attacks arrested by stopping aura," *Lancet* 1868.

CHAPTER CONTENTS

IN THIS CHAPTER I invite the participants of my study to tell their stories, from their own subjective perspectives, and in their own words. The reader is invited to listen and appreciate the complexity of the syndrome, its many challenges, and the different ways in which the afflicted live with their challenges. Reading the stories, one is often tempted to compare and contrast them. I refrain. This would have interfered between the people and the reader.

As was briefly described earlier, in 2007 and 2008 I interviewed thirteen men and three women, ranging in age from 18 to 65. They were selected for the study since they were diagnosed with Tourette syndrome (TS), but during the interviews were found to also suffer from obsessive compulsive disorder (OCD). They make up a heterogeneous population. Some suffer from anxiety; some from attention deficit. Their Tourette's motor tics vary from mild facial tics to severe and frequent body tics; their vocal tics vary too, from coughing and sniffing to coprolalia.Accordingly, their treatments vary. Two receive no treatment at all; the rest are treated by Dr. Jorge Juncos of the Emory Clinic of Movement Disorders with medications; and two underwent deep brain stimulation (DBS) by Dr. Mahlon DeLong[51]. Their phenotypes are partially masked by their treatments.

All are given pseudonyms here. Their other details -- age, gender, marital status, occupation, and the course of their disorder – are correct. The purpose of this chapter is to introduce them as people, not as data sources, and to listen to their voices. Admittedly, I could not disappear completely. The interviews, intended for exploration, were guided, like all explorations, by my preconceptions – some conscious, others not. Similarly, my

[51] Dr. Mahlon DeLong is a professor of neurology at Emory University. In 1990 he contributed to the Albin-DeLong model of the basal ganglia. For the last few decades he successfully performed many DBS operations on patients with movement disorders.

selection of what to describe and quote is contaminated with my own impressions and understanding. Neither could the physician disappear. His behavioral observations, diagnoses, and medical interventions partly define each patient's syndrome. His prescribed treatments are designed to ameliorate the signs and symptoms. Unfortunately, the treatment never cures and rarely removes the signs and symptoms. The most one can hope for is amelioration with minimal adverse side effects.

I report the medications prescribed to each person and what I know about their effects. I do not detail the doses and their schedules. Similarly, while I identify the targets of the DBS, I give only their names, not their more precise coordinates; neither do I describe the electrode frequencies and impedance, or their maker. Prescribing the medications and tuning the electrodes are a combination of science and art. They are determined by a trial-and-error iterative process, in which the response of the patient to the treatment determines the future course of the treatment. The details are important and indispensible for optimal intervention. However, my intention here is not to present a collection of medical records.

The human mosaic introduced here is as rich as any emerging from 16 individuals. Some speak at greater length than others. Some have deeper insights into their life in general and their disorder in particular. Some have advanced education; others struggle with basic reading and math. Each meets the challenges of TS in a unique way, with a unique blend of rage, accepting smiles, loneliness, depression, and despair. The reader will like some more than others, will trust some more than others, and will find some more interesting than others -- just as in any social situation. Not all that I

learned from them could be presented here; some is left for later. And some of what is here will be repeated with additional discussions later, where relevant.

Steven

(Finkelstein et al., 2007-09-28):

Steven is a handsome 19-year-old man who lives with his parents in an affluent suburb of a large city. He recently dropped out of out-of-state college due to his severe TS signs, which worsened when he moved away from home and had to adapt to college life. After returning home, he started to work with children who have special needs. He likes his job and is loved by the children. He continues to take classes in a local two-year college and perform well. In his free time he enjoys screen-writing, for which he received some encouraging feedback. Writing screen-scripts for Hollywood is his dream.

Steven's first tics appeared two weeks after his tonsils were removed, shortly before grammar school. His diagnosis followed shortly and was for his mother like "a death sentence." His first tics included neck-cracking, which is "the only tic that remains. All the other tics changed over time." In the interview, in addition to his neck cracking, he cracks his knuckles. In his Tic Symptoms Self Report (TSSR), he indicates facial tics, head jerks, and copropraxia; but he scores these tics as infrequent and not forceful.

By contrast, all his vocal tics are frequent and forceful. They include grunting, throat clearing, coughing, sniffing, hooting, coprolalia, and echolalia. But his greatest challenge is screaming. He has some control over it in that the worst never happens in public, only at home: "No one, no friends, no people have ever seen me to the highest

degree [of screaming] as my Mom or my Dad." While his room is on the upper level, his mother sleeps in the basement with ear plugs and two pillows over her head to block the sound. For college, his parents built him a sound-proof booth where he could scream unheard. This became his haven. And even though it had no windows or other ventilation, he spent most of his time there: "I could go there as often as I wanted and scream. No one heard it. It was for me… I never accustomed to this environment [the college]… I had to find some privacy… That's when the sound proof room came in."

Besides screaming, Steven also has coprolalia, which he calls "coprolalia contradiction… because it contradicts everything I want to say. Because I, if I look at someone and insult him, which had happened before, it's the opposite of what I think of that person." Steven succeeded to replace his "fuck, shit, nigger, fucking slut," and "My cousin is a slut" at her confirmation party, with a grunt: "I got rid of it [the coprolalia] by turning it all into grunts, all of it. But that …can all come back." The possibility of the coprolalia coming back is scary. Steven remembers with embarrassment his tutor who came to the house to help him with math: "Since the fifth grade she would come to the house and help me out… It wasn't actually ethnic slurs… It was, it was, she would come over and I would like," too embarrassed to describe the situation, he pauses and then continues, "She completely understood; she was *so* nice about it, and it never bothered her, never at all; but I would say some, like erotic things about her… things like I would never say. Coprolalia contradiction. I just don't say those things." And even though he succeeded in replacing his coprolalia with a grunt, the coprolalia is "always there… No matter how hard I try to hide it, it's always there, ya it is."

Exploring when "the feeling" is less intense and the tics diminish, Steven says, "When I am active… interacting with the kids… enjoying myself." He also tics much less when he is intimate with a woman. Working-out reduces his motor tics, but the vocal tics persist: "I used to work out," but when lifting weights I had to take one hand off the weight and grunt." The tics also recede with "anything creative. [This] gives me a lot of relief," he says, alluding to his screen-writing.

When the tics worsen, "time goes so much slowly. When you have Tourette time is … very slow." The obsessive compulsive disorder (OCD) contributes much to the tics, sometimes drives them: "Before a test, I obsess about it, the tics increase." And he further describes: "[I] obsess over trivial things; obsess about the writing… When you have OCD… you get very, very impatient."

The most florid of Steven's tics -- his screaming -- is also linked to his OCD, in that it has to be a "certain noise." It is often preceded by "a bubble. This invisible bubble in my throat that is always there [and that] has to come out. It builds and it builds and it builds. One time I was stuck on a tic for 4 hours. I was trying to get certain noise out… It wouldn't come out so… I was on the floor trying to get it out and eventually I did. I was so relieved." The screaming is a tic that occupies much of Steven's attention: "I always wanted to see if I could scream under the water... It did not work…" And since under the water "the urge is still there," Steven avoids water, where he cannot tic.

Some of Steven's medications[52] address his OCD, as this seems to be the main drive of his tics. The tricyclic antidepressant[53] Anafranil had to be replaced by the

[52] The medications names are commercial. The generic name is in parenthesis
[53] The tricyclic antidepressants are characterized by their three-ring chemical structure. They block the reuptake of the neurotransmitters norepinephrine and serotonin, thus increase their concentration in the synaptic cleft.

selective serotonin reuptake inhibitor (SSRI)[54] Lexparo, to avoid the side-effect of erectile dysfunction. In his interview, Steven says that the antidepressant monoamine oxidase inhibitor (MOAI)[55] Parnate "prohibited [him] from ticcing in public," but his medical records that were available to me and go as far back as January 2007 do not include Parnate. Steven's tics are addressed with the atypical antipsychotics[56] Abilify (aripiprazole) and Risperdal (risperidone). To improve his sleep he takes the α_2 adregenic agonist[57] Tenex (guanfacine) , which helps. For his anxiety, he takes the Benzo tranquilizer Klonopin (clonazepam), which helps too. With his medications, Steven sleeps well and his anxiety has lessened. Maybe his ability to transform his coprolalia to grunts is thanks to his medications too. But his OCD still drives his life and he continues to scream.

Steven has friends from high school but currently they are all away, in college. When they come home for a break, they get together, and have a good time. Recently he developed a new friendship with a young man who is legally blind. The two of them enjoy each other's company. Steven dates over the Internet. But the face-to-face meetings after the exchange of emails is problematic, especially if the relation takes a serious turn and the girl's parents get involved. He expresses one wish before the interview is over -- that people would be able to look at *him*, beyond his Tourette syndrome.

[54] SSRI – selective serotonin reuptake inhibitors used to treat antidepression and OCD by blocking the presynaptic membrane transporter for the 5-HT.
[55] Monoamine oxidase is an enzyme responsible for metabolic breakdown of catecholamines and serotonin.
[56] Atypical antipsychotic drugs are the second generation of antipsychotics, aka neuroleptics. Unlike the classic neuroleptics, they generally do not cause significant extrapyramidal side-effects of involuntary movements that are associated with the basal ganglia.
[57] α_2 adregenic agonists activate autoreceptors and post-synaptic receptors of the α_2 family.

Lionel

(Finkelstein et al., 2007-11-07):

Lionel, a painter of idyllic landscapes and reflective portrays, looks 15 years younger than his 65 years. Three times divorced, he has two sons, 43 and 38, with no TS. Neither does his brother have TS. But his niece's child might have it, and so does his first paternal cousin. His father also had (undiagnosed) light form of the disorder.

During the interview Lionel blinks often with his right eye, repeatedly clears his throat, and sniffs. In addition, in his TSSR questionnaire he reports frequent and forceful motor tics that include head jerks, shoulder jerks, tensing parts of his body, touching parts of his body, having copropraxia, touching objects, and tapping his fingers. He hurts himself and he has great difficulty in starting actions. To a lesser degree he has stomach jerks, and arm movements. He also kicks his legs and touches other people.

In addition to his sniffing and throat clearing during the interview, his vocal tics include frequent and forceful grunting, coughing, uttering single syllables, uttering single words, coprolalia, and echolalia. To a lesser degree he has involuntary whistling, hooting, barking, gulping, humming, palilalia,[58] and insults.

In his Y-BOCS Compulsion Checklist, Lionel lists a set of compulsive behaviors that include checking items, checking that he did not harm self or others, checking that nothing terrible would happen, checking for mistakes, counting repeatedly to 5, saving things like bits of paper and strings, having superstitions, rehearsing mental rituals, having the need to touch and tap, and having the need to do things "until they feel right."

[58] Palilalia is involuntarily repeating of one's own words or sentences.

He has had all these compulsions in the past and in the present. But some past compulsions, like repeating rituals and need for symmetry, have disappeared.

Lionel does not take any medications; partly because the anti-epileptic drug Keppra *(levetiracetam)* did not improve his tics and "made [him] sleepy," and partly because he distrusts psychiatrists and neurologists. Before diagnosis he had seen many of them, and "if they knew what they were doing they should have been able to diagnose me." At the age of 48 he read a magazine article describing TS. "This story was me." The phone number in the article took him to the Tourette Syndrome Association (TSA) that referred him to a local physician who confirmed his self-diagnosis. The relief was great. "The Tourette dominated my life… To be told that you are not insane…. I thought I was the only person in the world like that." But along with the relief came much anger toward doctors who had failed to diagnose him correctly, and teachers who had mistreated and humiliated him.

Lionel thinks that his tics started when he was five. He used to point with his right index finger, holding it alarmingly close to his right eye -- aimlessly according to his parents, mimicking according to him. At moments of great excitement or exertion, as in sports, he often had seizures, and he learned to recognize their coming, "I could feel it coming… Something in the stomach like moving and when this feeling come to my head I knew that I was going to collapse. I would have maybe 20 seconds." He would lie down to avoid injury. During a seizure he could not breathe or speak, and at times turned blue, but he didn't pass out. The seizure would last up to two minutes. People would bring him to the doctor thinking he had fainted. "Nobody asked me what had happened." The doctors told his parents that he was nervous, had "bad nerves." When his fifth grade

teacher noticed his blinking and thought that he had difficulty reading what was on the board he explained to her that he had "bad nerves." With the years, the seizures decreased in frequency and stopped at the age of 15. Since, Lionel had only two, the last one 30 years ago.

He can suppress the tics to some extent. But "you can suppress them so long, and they come back with a vengeance." Self mutilation -- biting intensely on his lips and putting needles into his skin – served as another way to reduce the tics, but he had to go to higher and higher pain thresholds to quiet down the tics with the pain, "as with drug addiction." The tics diminish when he is "really focus[ed] hard on something." Besides, over the years there is the wane-and-wax pattern of the tics. A few years ago his tics waned, almost completely disappeared for a few months; but they came back. They waxed most between the ages of 8 and 11 to a degree that he thought he "would not be able to go on."

This is also when his coprolalia started, at the age of 8. He would try to hide his involuntary foul expletives by "muttering the things under my breath." But then, he would scream them out when he came home or went out to the woods. His coprolalia started as a ritualistic sequence of "shit, damn, darn, hell, fuck;" and "they were not meant to offend anybody… I just had to spit it out." The coprolalia caused him to question his own sanity. Prior to it he had other vocal tics including groaning and meaningless words. His coprolalia was initially set off by hearing or reading the letters R or T, or the number 4. When he was asked by a teacher to add 5+4 he would distract himself from the 4 to 5 and answer 10, knowing that this was wrong. To avoid being caught with a 4, he would count to 5 over and over again spending most of his day

counting. This has greatly improved, and since highschool no longer affected his behavior. But his coprolalia persists. It can be triggered by “certain blue color dress... It can be a certain tone, even the feel of a certain fabric.”

Currently his tics --facial motor tics, occasional vocal tics, and occasional coprolalia -- are not too bad. But they are just “the tip of the iceberg.” Obsessive and intrusive thoughts greatly disturb Lionel. “It’s like being in this room with too many TV’s with too many channels being wide open all the time, and I try to have a conversation with someone when all this crap is going on… I am constantly seeing things ... I can’t push the stuff away.” The obsessive and “evil” thoughts can be triggered by a movie after which he assumes the role of the villain, and harms those closest to him. He experiences “*some*thing *foreign*… inside me and it was twisting; it was trying to torment me; it was trying to torture me and make me think things I didn’t want to think.” As with his other tics, Lionel has found a coping strategy. After realizing that “the stronger I fight the stronger it gets… [I] pretend it’s a bad movie and… [I have] to sit through it and just watch it.” His approach decreases the control of the “demon” and carries him through these torturous hours.

Ted

(Finkelstein et al., 2007-08-06):

Ted is 41 years old, tall, well built, and strong. His sense of humor is contagious. He was born to an affluent Jewish family, who expected as much from him as from his brother, who does not have TS, and loved him at his most difficult moments. His marriage follows a similar pattern, as his wife is “strict” and “provides structure and a lot of strength.” Their boy is a toddler.

Ted is working and is "well received" in his job with children with special needs. He attributes his success partly to being seen as a man who "gives them [the children] a mentor. The parents look at me where I am now and see where their kids can go." But prior to his current employment he had lost a job that he held for 15 years in a camera shop when he uttered "nigger" to a customer.

Ted's TS started at around the age of nine, but he was first diagnosed only three years later. The diagnosis brought great relief, "because I knew I was not crazy." He is not the only one in his family with TS. While his brother does not have TS, his brother's son does. And the brother suffers from obsessive compulsive (OC) behavior.

Ted has his share of OC behavior too. Often, when he goes to work, "every third car – I have to touch it." He has obsessive, repeating thoughts of death, and fearful thoughts of his own funeral and of his wife leaving him. In his Y-BOCS Compulsions Checklist, Ted lists repeating rituals, counting, need for symmetry, difficulty to throw things away, compulsive need to talk, need to tap, need to do things until it feels right, and picking his skin. While he does not harm himself anymore, he has acquired new compulsions of excessive showering and excessive cleaning of items. Dr. Juncos ranks all his compulsions as mild.

His motor tics include punching holes in the walls of the house and regularly pushing the windshield out of the car -- what his mother calls "the most expensive tic." Social situations trigger most of Ted's tics, "people are stimulating to me." His coprolalia is frequent and highly relevant to the situation. Black people evoke "nigger," fat women evoke "you are so fucking fat," a gay friend evokes, "want to suck your dick," large breasts evoke "you got big tits," and the gathering of the congregation in the synagogue

during the Jewish High Holidays evoked the most inappropriate utterance of "I love Jesus." At his better moments Ted succeeds covering up the embarrassing utterance by replacing "nigger" with "Nick," or "bitch" with "witch." During the interview he holds a pencil between his teeth to avoid any utterance. While Ted experiences certain physical relief after his coprolalic utterance, his dominant emotion is of embarrassment, "I love people. I'm not prejudiced… It's almost like the opposite." Ted's copropraxia is also triggered by social stimulations. During the interview, he responds to some stressful questions by raising his middle finger at me. As with his attempt to cover up "nigger" with "Nick," he sometimes succeeds in raising his thumb rather than his middle finger, and thus reduces the social insult.

Ted's tics can be set off by describing them. This happens often during the interview. In his TSSR he lists his tics, including some that are not manifested in the interview: He ranks his finger tapping, touching other people and objects, and copropraxia as frequent and forceful. Many other motor tics are infrequent and not forceful like facial tics, head and shoulder jerks, arm and leg movements, tensing parts of the body, touching parts of the body, smelling, and spitting. Among his vocal tics he lists throat clearing and hooting as infrequent. The list of frequent and forceful includes grunting, coughing, sniffing, barking, saying single syllables and words, and coprolalia as well as echolalia and insults. Dr. Juncos ranked the number, frequency, intensity, and complexity of Ted's tics as moderate-to-marked, and the interference between them as mild.

The vocal and coprolalic tics, Ted says, are faster and more sudden than the motor tics. He compares the urge that precedes them to a pre-sneezing experience. Unlike some

of his TS friends, he cannot hold off a tic until he comes home; he cannot hold off a curse and then come home and scream it out to release the built-up pressure. It is therefore noteworthy that while "I would love to take my mother's glass vase and go like this" (he gestures the dropping of the vase on the floor), the vase is still on the dining table in one piece.

In the mid 1980's, quite in the beginning of the biological revolution in understanding TS, Ted was 20 years old. He was treated with the dopamine-receptor blocker Haldol (haloperidol). And here is how he describes half a year of his life with this medication:

> I started to hit my face and biting nails… I locked myself in the room because I didn't [want] people to find me. Finally they found me and they rushed me to Beth Israel Hospital and… sedated me and put me in a straight jacket… My parents were across the country (sniffs) in Czechoslovakia and they came right back and they put me in Mass General and it got worse and worse 'cause they were raising up the Haldol, and I got to a point where they had to take me out a lot of time on hand cuffs. I would push my teeth on the bed [and pulled them out with my hands]… My father is a dentist: it's absolutely… almost impossible to pull out your teeth. The strength I got was awful. My eyes -- I'd beat them so bad I had to have a cataract surgery on both eyes and I had the beginning of glaucoma… It was crazy… Part of the time that I was in Mass General I thought to myself, 'if I could get out of these hand cuffs and jump out the window, I would.' That's how much pain I was in. Can you imagine your lips and face being open and still sticking fingers. It's like cutting your hand and putting salt in it, salt in it, salt in

> it… I could think straight. I had a normal mind. I was beating the shit out of myself. I was tired. I just wanted to close my eyes and not wake up anymore… My heart got real low. I [gained] 130 lb. It was like awful, awful. My face was all scarred up and my lips,… I had no teeth… And my Mom stayed the whole time and my Dad came up every week... You would never wish to see your child or anyone go through what I went. I was hurting the whole body was hurting. It was almost like, and I don't believe in the devil or anything, it was almost like my body was taken over by evil spirit. But it wasn't of course, it wasn't. Something was in me trying to hurt me… 'Oh Mom help me!'

When taken off the Haldol he got better almost immediately.

Now Ted's tics are treated with the blocker of dopaminergic receptors Orap (pimozide); the MAOI Parnate (tranylcympromine) is for his depression; the over-the-counter (OTC) antihistamine Unisom, taken at night, induces sleep; and herbs and vitamins are supplemented for his general well being. In addition Ted self-medicates with marijuana daily. "If I take a joint of 3 or 4 inch of it, it goes away for a few hours…It's illegal and there are ramifications… [But] it's the only thing that takes the tics away… I don't consider myself a drug user. I only self medicate." Sugar and caffeine affect him badly, but "I like it sometimes. I like it in the morning time. I'll sacrifice a few tics for a good Coca Cola."

Kyle

(Finkelstein et al., 2007-09-21):

Kyle, 25, blinks frequently and utters coprolalic words as well as various sounds under his breath. His "fuck, fuck" is so soft and melodic that it takes me a while to recognize it for what it is. When I share my impression with him, Kyle explains, "I say lots of words but I try to keep it under my breath because people would think I'm crazy." This is not Kyle's only characteristic of his expletives. "I would have to get the cuss word just right before it would stop. And I had to say it over and over and over, and it has to sound *just right* to me. And I keep doing it." When I ask him to further elaborate, he explains with both his hands on his throat, "It's like I say fuck fuck fuck and I have to get just the perfect sound before I would stop… I keep doing it until it feels right."

He then lists out his coprolaliac vocabulary, "I will say [the] 4-letter word, and G-D, and bitch, and M-F," but no racial slurs "I live in a black neighborhood. I don't think that will be good." Atypically, his coprolalia is not necessarily situational. It can be triggered purely from within: "Sometimes it all comes out. I feel like cursing somebody out, who's not even there."

His internal world often brings him terror. "[It] happens every once in a while. I'll be going to sleep. I'll be like somewhere in and out of sleep, and my whole body can't move. And then I think I can't breathe, and it takes all the energy I have to get out of it. And I feel like I'm getting jolted shocked. So he [my partner, William] notices that, because I'll try to cry for help. And he notices it coming out of my breath. And he'll shake me and I'll wake up. So I'm scared to go back to sleep after that, because it's so scary. I don't know what it is. It happens." To fall asleep he needs some calmness and

"sometimes [the tics] won't stop enough for me to go to sleep." His close friend, Monica, with whom he roommated for a few months, is with him. In her interview, she says she never saw him sleeping, "he is always awake." He would be the one to wake her up, as early as he could. She speculates that it might be his anxiety and therefore "he always needs to be with someone."

When he listens to music, Kyle's tics lessen or disappear altogether. He thinks that this is because music puts him "in focus." He enjoys "loud, crazy [music] that changes and has a lot of beats and a lot of bass." With such music, "I can get into it; just go with the flow of the music. Because this is not something I have to do. It's doing it for me."

This passive experience with music differs from his activity of cooking, which also ameliorates Kyle's tics. His partner has a small catering company and, "[cooking] is one of my favorite things to do…We cook all the time. We don't go out to eat. We cook *all* the time." And despite his tics, when cooking, he exerts some control: "I know if I am not careful [with the knife] I get cut." Cooking "takes my mind off things." It provides a happy environment, "I can listen to music, and dance around, and cook." He cooked even before meeting William. Monica says that he had cooked for her and her sister "all the time" and that he is a "very good cook." She also observes that he is skillful with knives and his motor tics are not in any ways interfering with his cooking.

His cooking connects Kyle to happy moments in his childhood, with his maternal grandmother who "pretty much raised [him]." Today, however, their relationships are stressful. She does not accept "that he is gay," says Monica. And in ranking his tics 1-10,

Monica adds that while he is 5 when he is with her and her sister, he is 10 with his grandmother.

His childhood was painful. His biological father disappeared, and when Kyle was three years old, his mother remarried. “I had a really bad childhood with my stepfather. I can’t stand him and he can’t stand me.” His stepfather came before the tics, and when they emerged, his stepfather “thought that [Kyle] was just doing it to get on his [stepfather’s] nerves.” But Kyle thinks that his stepfather “is just mean. He doesn’t like me. I think that’s the way he used to have reason to abuse [me, with screaming and beating].” By contrast his two younger half-sisters have accepted him, but they never became friends.

Kyle’s tics emerged around the 2nd or 3rd grade, when he “started to get into trouble for making noises and distracting the class.” His mother tells him that he had a tic in which he would widely open his mouth, but he does not remember his motor tics. In general his awareness of his motor tics is low, unlike with his vocal tics. Them, “I can *hear* them.”

He was diagnosed with Tourette syndrome around the 4th or 5th grade, and until then “they didn’t know what it was.” The diagnosis helped. Prior to being diagnosed, “it was kind of scary ‘cause nobody knew what it was; and nobody that I knew had it; except for me.” And the diagnosis “made it kind of easier [in school] because I didn’t get into trouble… anymore.”

Like most who have Tourette syndrome, when Kyle “get[s] really nervous, that brings up more [tics].” But when I ask him if he tics more around new people he tells me that “No, I just try to control it with new people until they notice it, and then I tell them.”

Despite his openness about having TS and his strategies, which cover most of his coprolalia under his breath, Kyle fails to hold a job. This might be due to his difficulty to focus. “I can’t pay attention. I can’t focus... There is so much stuff going on that I just can’t get anything done… I can’t explain it. It’s like at work. I get at work something to do, and I just couldn’t pay attention to it. It’s weird to explain… I just can’t sit still. I want to go do something else, run to the bathroom, go find something else to do, listen to music. I can’t get nothing done.”

The difficulty to focus is accompanied with difficulties to read and write: “I don’t read. I don’t bother to read. I can’t pay attention to it. I really don’t bother to read… too much going on for me to pay attention to and bring it all in… I can’t remember stuff.” He remembers what he reads only when “it’s something really interesting.” When he has to read he “ [has] to force [himself] to write it down and make sure and double check.” And this is an effort. The writing presents a motor difficulty. He says, “ I can write really good in the beginning, but if I write a lot it just gets chicken scrabble and my hand hurts. I don’t bother with it usually.” Typing is easier for him: “I’d rather type… I have to look at the keys but I can … that’s not effort. I can do it. It’s not a problem... It just takes me a little while but I get it done.”

His poor memory troubles him too: “People will tell me stuff sometimes. I have to ask them to repeat themselves a lot.” And then he adds, “I can’t remember what I did yesterday half of the day. I have to really sit down and think about it… most of the time it’s out of order but I remember what I did. And sometimes I don’t remember things *at all*.”

In his TSSR self-report, Kyle ranked his motor tics of eye blinking, facial tics, head jerks, kicking legs, tensing parts of the body, and obscene gestures as very frequent and very forceful. Arm and hand movements, touching parts of his body, difficulty in starting actions, and finger tapping are infrequent and not forceful. His vocal tics include grunting, throat clearing, coughing, sniffing, hooting, uttering single words, and coprolalia. They are frequent and forceful. Whistling, humming, and echolalia are infrequent and not forceful. He also has obsessive compulsive behavior that includes repeating rituals, insisting on asking questions until getting an answer, doing certain things until they feel right, blinking rituals, and "cleaning the house until it's perfect." He used to have a counting ritual but not any more. Dr. Juncos ranked his total tic severity at the maximum; most of his obsessive compulsive behavior as severe; and his obsessive thoughts and the anxiety related to them as extreme and disabling.

With his gentle appearance and shy smiles, Kyle tells me that he has many friends. And indeed Monica speaks about him with great fondness. But he ends the interview with, "I always heard that it [Tourette syndrome] goes away when you get older… And it hadn't."

Dylan

(Finkelstein et al., 2007-08-01):

The 29-year-old Dylan was first diagnosed with Tourette syndrome when he was 7 years old, about a year after the onset of the disorder. While no one in the family prior to him was diagnosed with TS, presently his cousin – the son of his father's brother – is diagnosed with TS as well.

Dylan's first signs were the motor tics. At the age of 14 he developed vocal tics, and at 15 his coprolalia appeared. At 10th grade he had to leave school. His behavior was too disruptive to the teacher and the other kids. He was directed to a school for the mentally and physically severely disabled. With his high intelligence he did not fit there, suffered greatly, and left. Under his father's supervision he completed his GED, "I saw that he got it, at least that" his father says.

Dylan had two brain surgeries of deep brain stimulation (DBS). First he had a bilateral DBS in the anteromedial part of the globus pallidus internus (GPi) region of the basal ganglia. It greatly reduced his self mutilation and intense tics. However, the self mutilation, like all his other signs, was only ameliorated, not cured. He continued to scratch his skull, at the site of the operation. This led to infection and the electrodes had to be removed. After healing, a new bilateral DBS was performed in December 2006. This time in the posteroventral GPi.

"You say it took courage," Dylan's father says to me about the decision to operate. "I don't know what courage, because once we knew what it was and what it could do we were eager to do it and there was some apprehension of course because it's a brain surgery and, you know, anything can go wrong but it really [got] to the point where we couldn't imagine it getting any worse. Really there is no way that I could imagine it being any worse."

Before the surgery, his motor tics at home were severe, especially in small, confined spaces. His father tells me that in an elevator, Dylan would bang the walls of the cage. He could not be confined to a closed place or a seat and would therefore not go to church or the movies. Driving the 4 hours from home to the neurologist in Atlanta,

Dylan's father had to handcuff and footcuff him since otherwise he would disrupt his father's driving and might cause an accident. The bathroom was especially a challenging space. When looking into the mirror, Dylan would start shaking, having a tremor. "I had to grab him from behind," says his father, "or he would go into the wall." While taking a bath, "he would go into the faucets; bashed his head repeatedly into a corner in the wall." Dylan's repeated motor tics turned often into self-mutilating behavior. He was hitting his head against the wall, burning himself with cigarettes, rubbing his eyes, and gouging in them until they got "blood red."

Dylan could not hold a job. His day was consumed with his tics. The only relief came with sleep. He did not and does not tic during sleep. Sports also provide some relief, Dylan says: "As a child [I was a] pretty good baseball player; pretty good athlete in spite of the tics; swirled but when the ball came, I would stop whirling. When I had to stop and catch the ball, I stopped to catch the ball; always just at the right time." With dancing feet he tells me about his golf playing, "I am a good golfer. I am left handed… pretty accurate, pretty good… [The tics] slow down… I concentrate." He can have an hour or more with almost no tics, "even before the DBS."

There is a Dylan before the DBS and after the DBS. While he has not been cured by the surgery there is a significant amelioration of his signs and symptoms (50% according to Dr. Juncos). He still has head jerks, copropraxia, coprolalia, and grunting. They take turns, "Some days my coprolalia is worse. Some days my motor is worse." Prior to the DBS they both were bad, "very bad." His coprolalia included "fuck, shit, bitch, whore, spick, pussy, tits, nigger, I want to have sex with you, [and] goddamn," always uttered in a manner relevant to the specific social situation. "Spick" was reserved

for his Hispanic physician, “nigger” for his Black friends or Black passers by, and “I want to have sex with you” to the woman who exercised on the tread mill next to his – for this he was kicked out of the gym. The only way Dylan, whose behavior is polite and gentle, agrees to share his coprolalia vocabulary with me is in writing.

His curses are most intense around new people. “Is it a little bit of a test?” I ask him. “Yes! Somebody that I don’t know… I’ll tic more towards them…” Moreover, if there are a few new people, the one that Dylan likes the least gets the most. When among strangers, his coprolalia might have dangerous consequences, and Dylan is “scared” if his coprolalia is directed at a “big Black guy.” His friends, though, many are Black, like him and have accepted him. The closest of them intuitively discharges the racial tension, and when Dylan, who is Caucasian calls him “nigger,” he says to him, “ I am a nigger and you are a nigger.” They both laugh and it brings an end to the tension and the “nigger.”

While his copropraxia before the DBS included inappropriate touching of himself and others in public, now this happens only rarely. He still raises his middle finger, but often succeeds in masking it as a gesture of moving hair off the forehead. His coprolalia is muffled down too. His motor tics of whirling and bending have greatly diminished in both intensity and frequency. The tics that stayed are gentler and less disruptive. In his TSSR Dylan reports the additional motor tics of head jerks, and movements of the arms, hands, and legs. His vocal tics include grunting, coprolalia, and palilalia. His OCD is manifested in compulsively checking the clock, and when alone, he keeps thinking of turning the lights off.

His self-mutilation and all his intense tics have been greatly reduced. Unfortunately, in the second DBS Dylan acquired a slight stuttering. He sometimes loses

control over the temporal order of his words, "I am getting ahead of myself." In spite of this DBS-acquired deficiency, he finds his current condition much more "controlled" and says, "The DBS has done wonders for me." He can now sit in the passenger seat next to the driver, he has a girlfriend, he can go to the mall, and he holds a job. None of these was possible prior to the DBS.

Dylan continues to take medications and is under medical supervision. Many drugs have been tried, "He has been on… just about every other SSRI in the book," reports Dr. Juncos in Dylan's medical record. After the DBS, the dosage of his medications has been reduced gradually. But there are some side effects. Dylan takes the antidepressants Anafranil (clomipramine) and trazodone. His physician attributes his acquired erectile dysfunction to the trazodone, and recommends lowering its dose, and replacing it for better sleep with the non-Benzo[59] sleeping pill Sonata (zaleplon). Attempts to treat Dylan with other medications were met with drug-resistance; maybe because he keeps returning to cocaine. Dr. Juncos has conditioned the continuation of his treatment on stopping the cocaine, which Dylan succeeded to accomplish twice, with the help of a rehabilitation facility. He is now off cocaine and back to his routine and bi-annual visits to his neurologist, who hopes that Dylan would find new friends who do not use drugs.

Henry

(Finkelstein et al., 2007-07-23):

Henry, 59, welcomes the interview as an opportunity to interact with people and even show off a bit. Looking at the cameraman, he asks, "Ready? Am I on the air?" and

[59] The Benzodiazepine (Benzo for short) are anxiolytic (reduce anxiety) drugs and muscle relaxants that do not reduce alertness.

only when answered in the affirmative does he start barking and shrieking, running towards the camera. Then he closes his introduction by assuring me, “I’m alright. Not crazy. It’s just my tics.” To stop this continuous chatter he addresses himself, “I’m tired of you boy. Down boy, down!”

Henry often speaks to himself. Whenever he is unhappy with his repeating words, or losing concentration, or “going on tangents,” he turns in his chair, and as though facing someone else, he addresses himself as an adult addressing a child, “Be quiet! I am tired of you. Grow up! Grow up!” And at one extreme dialogue, he continues, “Disconnect from your mother! Mother please cut the umbilical cord for me.” How involuntary these dialogues are, is hard to say, especially since immediately before our interview he was examined by a nurse and did not tic at all.

Henry is articulate, animated, and gestures richly. He intonates his phrases clearly, even beautifully. Occasionally he shrieks to a level that is painful to the listener, but then quickly resorts to controlled speech that sometime turns into a whisper. To answer questions, he shuts his eyes. Only then do his answers cohere. When he opens his eyes, he gets distracted and loses his line of thought. But before answering questions, he has his own agenda. Henry shows me the product of his OCD, a densely written notebook with repeated phrases that seem like their only function is to fill in the pages and leave no white spaces. Similarly, his speech has no spaces -- a continuous stream that can be stopped only from the outside or in a dialogue in which he scolds himself. Indeed, Henry describes himself as producing endless streams of words, “I write and I write and it takes me hours and hours and hours.” This he attributes to his OCD. The SSRI Paxil

(paroxetine) ameliorated his OCD, but he stopped taking it, "too expensive," and his condition has worsened again.

There is an interesting relation between Henry's tics and his OCD. When he writes compulsively, he cannot tic. "You can't jerk when you write. You have to be still. So energy starts to build up, then builds up... and it builds up and builds up, and across over a certain point I can't put it down… I miss most of my lunches because of this… It's a driving force." Finally, when the urge to tic overcomes the compulsion to "have the last word" in writing, the tics emerge and then they are severe.

Henry has a need for repetitious rituals and "usually I tic before I start something. I tic before I start. That's why I don't want to start; because I have to go through all these tics before I get started. I got to go through these rituals and these tics and preparations like foreplay." He also has the tendency to repeat his own words, "I verbalize and repeat myself and talk over and over and over and over, over, over, over, over, over; and when I tend to write things then I repeat things on paper over, and over, and over, and over, and over, and over, and over, and over, and over." This monotonous repetition is often demonstrated motorically too, in a lulling or abrupt and sharp swaying in his seat back and forth, back and forth.

His tics can be severe, sometimes he has seizures. "Maybe I will end up being on the floor." The seizures, he insists, are tics and he never completely loses control. "If I'm on the floor it looks like I have an epileptic fit. I'm still in control." He describes a premonition rather than an urge prior to a seizure. He describes it in the present tense "Here it comes… like I am going to be delivered" even though it's clearly not about to happen, since it "happened maybe 5 times in my life," and the last one was "long, long,

long ago." This is how the premonition feels: "My head swells up; it's filled up. I sense the presence of something. I can almost smell something, almost taste. It is rising from deep within my soul, deep within my heart, coming to the surface." Then he mentions how at the time of Gilles de la Tourette people practiced exorcism, but "I am not demon-possessed." To close the topic he turns to his internal, yet performed dialogue, "Be quiet. I am tired of you."

When we explore the nature of his urge, he links it to his OCD. To my question whether the urge is a thought or a physical sensation, Henry responds: "It's all. *Everything.*" With a throat-cutting motion, followed with lifting his hands from his temples to the top of his head, he continues, "It's from here [the throat] up. Everything is from here (pauses) up. The urge comes from here," he says with both his hands on the top of his head. "It is like a malignant tumor that has attached itself to my brain. The brain and intelligence have diminished and the malignancy got bigger… I'm putting most of my energies into not yielding to the urge to tic. And I'm using the rest of my brain to try and get to work saying to you. Three things are going on at once. I can't release. The urge is there right now. I'm thinking about the urge more than about what I'm trying to tell you… and right now I have the urge to do many tics but I'm not. I'm not free." Sometimes he has a compulsive need to touch – objects and people. But it is not of any obscene nature.

Henry caused himself much damage through self-mutilation. At the age of 11 he lost his right eye and now has prosthesis. "I still have the tendency, not severe, to poke pencils and pens at my good eye." He also ruptured his appendix with a tic, in which his right elbow repeatedly hit his belly. Recently he adopted a new strategy, "I'm trying to

switch over from the physical tics to the verbal tics because my body just about had it." As he mentions the verbal tics, they get evoked. A new stammering emerges. "When I was a little boy like I t-t-told you the last time, I would say SS, you know, ch-ch-ch ef – f-f-f-f-f-f ch-ch-ch-ch." Then he mentions a "4-letter word", which he later utters sometimes.

Henry was diagnosed with TS when he was 30 years old. Since early age he has suffered from tics, self-mutilation, OCD, and dark thoughts. "As a boy I would have these thoughts." His darkest thoughts are about the people closest to him and that he "wish[es] the best to."

For the last 30 years, he lives with his parents. He is on disability, not employed and does not "feel good about this at all." His sister lives in New York and his brother, like Henry and their paretns, in Georgia. Twice Henry tried to move in with his brother and his family, but it failed. His mother is 80 years old and undergoes chemotherapy for her cancer. "She needs a miracle." To take her to her chemotherapy treatments, Henry gets up every morning at 5:30 am. Obsessing over getting up on time, his sleep is poor. His mother is his "best friend," yet their relations are as though he were still a little child. "Mom, you know me. Don't ask me to do anything like that, to take the trash out… You are my mother. You should know how I am. Don't say 'get the trash out, get the mail.' Please. I have enough on my mind." His 84-year-old father is non-ambulatory and would soon need "to go some place." While Henry is very angry at him for ongoing criticism, he gets softer when he mentions the old man's fragility.

In his self report Henry ranks his facial tics, touching parts of his body, finger tapping, and difficulty in starting actions as very frequent and very forceful. His head

jerks, stomach jerks, tensing parts of the body, and touching other people are infrequent and not forceful. But the list of "self abusive behavior" includes knee clapping, arm clapping, toe stomps, and hitting his belly with his elbow. And he is "very concerned" about the self-mutilating behavior.

His vocal tics include grunting, throat clearing, coughing, sniffing, hooting, and coprolalia, which are neither too frequent nor forceful. But repeating his own words -- "barking out loud," "laughing out loud," and "screaming out loud," to "relieve self of torment" -- is very frequent and very forceful. His compulsions are many and include cleaning, checking, counting, and arranging compulsions. In addition he has the need to confess and to make excessive lists.

Dr. Juncos recommended reinitiating the Paxil therapy, which seemed to have helped Henry in the past. In addition, Dr. Juncos recommends that he would take the tricyclic antidepressant Elavil (amitriptyline) for sleep; and for anxiety the Benzo tranquilizer Ativan (lorazepam) and clonazepam.

Totally consumed by his tics, compulsions, and self mutilation, Henry studied only one

year in a community college. He has two regrets: "I wish that I can help people with their problems, and disconnect from mine." And he laments, "All of my life I've been waiting to get better to do things. I'm almost 60… and I'm, I'm not down on myself, but I haven't read. My intellect is not --- I couldn't talk sports to you, couldn't talk politics to you, because I haven't read to gain the knowledge that I need. And that makes me feel a little bit sad. But I'm trying to turn it around now, and read more, more stimulation."

Stuart

(Finkelstein et al., 2007-10-26):

Stuart, 29, is studying clinical psychology. After completing his masters degree, he hopes to practice as a cognitive assessor. For his studies he needs to read a lot. He tends to procrastinate and often lacks motivation but he reads, and the tics do not interfere with his reading.

His first tics, mainly neck jerking, appeared in elementary school. But he was diagnosed with Tourette syndrome only at the age of 17, after a school counselor sent him to a psychiatrist: "I told her [the counselor] that sometimes… when I am sitting in a group and there is a person that I do not like, I start thinking 'bitch'… That's what made her suspect. She sent me to [a] psychiatrist." Stuart was glad to be diagnosed. He "like[s] having names for things. … because you can come with a treatment plan."

Most of his motor tics are facial and head jerks. But, as he reports in the TSSR, he also suffers to a lesser degree from eyeblinking, shoulder jerks, arm movements, hand movements, tensing parts of his body, touching parts of his body, picking at things, and copropraxia. Stuart distinguishes between jerks and tics. "The tics -- I can emulate, I can show you what a tic is. But muscle jerk -- it is completely involuntary. He refers to his akathisia – uncontrollable motor restlessness, jerking of his leg. He speculates that this myoclonic tic might result from his medication. Dr. Juncos agrees and attributes it to the Abilify.

Stuart's vocal tics include throat clearing, sniffing, whistling, gulping, humming, and insults. They are collectively ranked by Dr. Juncos as "mild." His most disturbing is his coprolalia. Embarrassed to share his coprolaliac vocabulary, Stuart writes it down:

"Good God damn," "Jesus fucker," and "Jesus fucking Christ." Despite the religious contents of these expressions he thinks that there is no danger that he would utter any of them in church, "No, I don't think so." In addition, his coprolalia includes little songs, which he makes up and sings.

Is his behavior completely involuntary? "It's not *completely* involuntary… [The tic] can be suppressed to some degree." For example, "with my father I sing this song. I can see that it aggravates him." Stuart sings it to me. The song is to the tune of "I'm a little teapot": "I'm a little faggot, yes I am, I eat shit as fast as I can." He laughs at the thought of aggravating his father with his homosexuality and adding to the complexity of the family dynamics.

Stuart is the only one in the family with TS. He is the youngest of five siblings and "there is 15-year gap between me and the next one. My oldest sister is 21 years older than myself." There is very little in common between him and his siblings, which he attributes to their age differences. When he was a child, they thought "I was making it up." He "[does] not remember much of [his] childhood." But even today, after all these years, with diagnosis and treatments, his mother "doesn't believe Tourette syndrome exists." He speaks of good relations with his parents. When he needs financial support they help. And he discusses with them some topics, including sexual issues. "I did talk to my Mom a little bit last night because maybe I would work with [Dr. Juncos] on some of these issues."

He does not specify whether by "these issues" he means his homosexuality or his obsession with pornography. "I have like a sexual addiction, pornography." He labels it as obsessive compulsive (OC) behavior. "I have had periods in my life when I'd

masturbate 5-6 time daily, but this would be for very short periods of time, 2-3 days. Then [there is] the impulse to buy pornography on the internet."

His OC is not limited to sex. "My finances are terrible. I have a budget; I always overdraw… I have to borrow from my parents. That impulsiveness… signing to websites, buying books that I really don't need, or buying a fancy dinner that I can't afford." In general, "when I think of something or I want to do something I just can't rid of it until I can do whatever it is, whether it's to go to a doctor about being ill, or reading an article about something … or buying something. I don't feel peaceful until I actually do that." At times he is also a compulsive eater. And while he tries to transform his OC behavior into healthy patterns of exercising, he fails because "the motivation is not there."

His OC is with him at night too, in the form of bruxism -- he grinds his teeth at night, and "I've lost a tooth surface from [it]." In addition he reports in his Y-BOCS Compulsions Checklist that he has a need for symmetry, he needs to touch things, and certain things need to be done "until it feels rights." It is also difficult for him to throw things away. Dr. Juncos ranks his OC as "severe," and expresses concern about Stuart's "rage attacks," which Stuart did not mention during the interview.

Some of Stuart's OC behavior has disappeared. It is impossible to determine whether this is thanks to medications, waning pattern, or other processes. He used to compulsively check his pants zipper; he had repeating rituals of rereading, erasing, and rewriting; and he had superstitious behaviors. These are all gone.

Stuart compares and contrasts his OC and his tics. "I think they are two different experiences. They [the impulsions and compulsions] can take time unto fruition… The tics [are] pretty quick." But there are also similarities, especially between coprolalia and

his pornography-related OC. With closed eyes he says, "Well, there's definitely an urge there. I think about sex until something happens. It's part of the OC spectrum." And as for his motor tics, "it's [a] relief to do them because you want to do them, and then when you do them it's kind like it's over for a little bit."

He also compares the experiences of motor tics and coprolalia. Unlike with motor tics, "I do not curse spontaneously… The proclivity to curse or sing silly songs … is there." And this proclivity is not limited to sexual utterances. It also includes racial slurs. When driving, he would say "stupid nigger," and would then feel certain relief. In 1999, when Stuart was 19, his brother was murdered by a Black man. In an emotionally-flat tone he says, "[This] kind of engrained some racial negative stereotypes towards Black people." It is difficult for him to sort out his feelings, or even describe the process of his racial slurs. Instead he shares an episode:

> Usually it's a trigger… One time I can think specifically of: I [was] with a friend and we were driving on a lane in a parking lot, very crowded parking lot… there is no parking spaces; and we found a little parking space; and we try to pull in; a Black person just swarms in. I say the 'n' word and then I was so angry; and we got inside the restaurant; we met a couple of other friends; so there were the four of us; I say this [word] several times. They got extremely offended by it 'cause one of them was a professor at Morehouse… He is a White Jew. I was going at the time to a synagogue for gay and lesbian people.

When I ask whether he felt embarrassment, relief, or anger, he answers, "Some anger maybe; a little embarrassment. It strained the friendship a little bit."

Medications ameliorate his coprolalia, "When I haven't taken medicine it got a little bit worse." Dr. Juncos, who saw Stuart for consultation, recommends stopping altogether or at least tapering down the antidepressant Wellbutrin (bupropion), which

causes some irritability. For his OCD, Dr. Juncos agrees with Stuart's physician to continue the transition from the SSRI Zoloft (sertaline) to the SSRI Lexapro (escitalopram). Decreasing the Abilify should help with the akathisia and might decrease the tics as well. Hopefully the mood stabilizer Topamax[60] would prove more effective than the currently prescribed Trileptal (oxcarbazepine), and would also affect the tics.

Some relief comes other ways too. When I ask him to shut his eyes for a while, Stuart looks completely calm, and has no tics, "it feels peaceful and relaxed." He then elaborates, "With the visual world you're seeing reality. Things are there." And by contrast, "When you listen to music you listen to something artificial, you can escape with the music... That's why the audio world can be comforting. You… are tuning out the reality of the visual." Stuart concentrates better with closed eyes and has better access to memory. And when he concentrates, he tics less. But "The most relaxing thing for me is to rock and rock in a chair. This [has] always been comforting and soothing to me. I probably tic less when I'm sitting in the chair rocking."

Stress increases his tics; at "finals week it's crazy." Also, "when I think of the tics they become more frequent, and more severe." Therefore, "when I come to Dr. Juncos… I tic a little bit more than I usually do. For two reasons: One, I'm thinking about it. And two, I don't try to stop them because I want him to see the manifestation of them." The second argument implies some control over the tics. And indeed Stuart hides his tics with some successful suppression or camouflage. For example, his clinical instructor and some of his fellow students tell him that they do not notice his tics.

When we discuss his copropraxia, our conversation evokes the raising of his middle finger. He softens the tic by modifying it into a less jerky, longer, and smoother

[60] Topamax is an anti-epileptic drug that is also used as mood stabilizer.

gesture. It is hard to recognize it as a finger-raising. Often when the copropraxia emerges, "I go out the door because my partner's inside and it can make him mad if I do that."

Stuart's relations with his partner are stressful, and continue mainly because of economical difficulties of his partner. He has a few friends from school, not a whole lot. "Sometime I go to church but not all the time. I've been going to sexaholic anonymous; each week. I missed a couple of weeks here and there but I do that. I have some friends online... I never had a lot of friends." A psychologist, to whom he was referred by his neurologist, thinks "that I [have] schizo-typo personality traits. I never really had a lot of friends… I think I am a lot better now than I used to be, but historically I had troubles in the social arena."

Elliot

(Finkelstein et al., 2007-08-13):

"My mom noticed… barking noises since I was in the crib," says the recently divorced 37-year-old Elliot, a proud father of 3-year-old and six-year-old sons. His children live with their mother, but Elliot, as well as his father and mother see them often.

His own parents divorced when he was 3 years old and his father moved to a distant city. He and his father live now in the same city and his mother is the one who lives far away. But Elliot has remained close with both his parents, and they continue to be supportive -- emotionally and when necessary, financially. Even though no one else in the family has been diagnosed with TS, Elliot's father, whom I interviewed too, often

eyeblinks and coughs, and occasionally tenses his body and lifts himself off his seat. He also shares with me that his own father stammered.

Elliot remembers "being 4, 5 years old… making noises." He also remembers that even at that early age he already noticed that he "had no choice," he had to let these noises out. Along with his vocal tics his motor tics evolved too and got worse and worse. At the time of his high school graduation his legs twitched so badly that he needed a wheel chair.

He was diagnosed with TS in 1977, at the age of 7, by the famous Dr. Arthur Shapiro. Elliot returned for consultation in 1980 but, "frankly, he [Dr. Shapiro] was concerned then with us buying his book." Elliot was suspended from third grade. The teachers took his tics as a behavioral problem. And for a reason that we did not discuss, his parents did not share the information about his TS with the school. Later, when they did, it helped "because it wasn't my fault" and the school apologized and cleared his records. For his parents it was a "major relief," his father says, "to know that it was physical rather than something mental" This because "you have a chance." After breaking the silence about his TS, Elliot told one new teacher about it. But otherwise he did not speak about his TS with anybody. And while the teacher was understanding, "the kids were murderous… physically and emotionally." He was put in a learning disability (LD) class, despite an "incredibly high IQ," according to his father. In high school, learning from his past experience, Elliot spoke about his condition more openly.

He is highly vulnerable to visual stimuli: "Environment is a big part of my tics… If I can only shut my eyes I can stop everything." Then he points to the light over his head and continues, "just that light above me, it's, it's, it's hitting this part of my head

(pointing to the center of his forehead) and I want to move it around." If he had control over his environment, if he could "design a house that [would] be perfect for Tourette's," it [would] have a "room that's all white or all black with nothing in it." And indeed when he shuts his eyes to make his point, his tics stop. When I ask about auditory stimulations, he answers, "No, sounds don't bother me." Typically he is free from tics "three times: sleeping, concentrating, [and] having sex. The sex is concentration, I think."

In his medical records, Elliot is not diagnosed for coprolalia but he confesses his need to use the "f word," which he can often control. He is happy to share his insights about the topic. The saying of the words is a reflection of anxiety. It provides a primitive satisfaction similar to that from orgasm or a physical tic. The satisfaction is *physical*; it is not a reward from sensing the disturbance or annoyance that it may provoke in others.

Not all his tics are preceded by a premonitory urge, but many are. In our exploration of the premonitory urge, Elliot describes in real time a tactile stimulation that generates an urge to tic: "I guess some people with Tourette say it just happens… but that's not the way it is for me. I can tell you right now… I can feel my neck right now because my shirt is stimulating my neck. I feel my shirt on my neck and I want to shrug my shoulders… it's this stimulation of touching."

Is there any relation between the location of the urge and the tic? "Ya, I have the urge in the location where the tic is. Like right now it's in my shoulder… I can feel the location... I can feel the other parts of my body getting involved in that. They get set off and… you cannot just settle with that one and be done with it." And he further explains why body parts other than the one where the urge first takes place are involved too, "You can't jump on the bed on one corner without the other corner feeling."

As for whether the urge is satisfied by its following tic he says, "It depends… There is a level of some tics that are more satisfied than others. But aftertime, all of a sudden, it all comes out again." Then he adds, "It's like a mosquito bite, you scratch it and then you're satisfied for a few seconds but then, you know, all of a sudden you need to do it again… And if you don't it drives you crazy." Nonetheless, he does not "have the explosion of tics that some people have" after suppressing a tic. But his ability to suppress is limited and "there is a moment that you *have* to go."

Elliot's use of language is advanced, which is somewhat surprising since he "hate[s] to read." In college he faked reading based on the first new pages; and he thinks he has never read a book cover-to-cover. He does read some articles in the newspapers and some news websites like CNN.com.

When browsing the Internet he often navigates to eBay, where he spends his money compulsively, with almost no control. In his Y-BOCS Compulsions Checklist he reports checking compulsions, repeating compulsions, ordering compulsions, and mental rituals. He thinks that some of his tics are driven by these compulsions.

He claps his knees against each other until they get blue. He links it to his obsessive need for symmetry, especially in relation to the tactile world. At some point in the interview, he blows strongly on the back of his left hand and then looks at the back of both hands saying, "I was blowing [on my left hand, and] felt the air from my nose [on] this hand, so I have to equalize on the other side and blow on it." Then he adds with a smile, "It's very physical." And he continues, describing other events that demonstrate his need for left-right symmetry: "When I was in school and someone bumped into me in the playground [on my] left side… I fell out of synch. I felt like I *had* to equal it out."

Dr. Juncos observes that Elliot's greatest challenge with his OC is neither his obsessions, nor his compulsions, which are quite mild, but rather his low resistance to his compulsions. In the Yale-Brown Obsessive Compulsive Scale Dr. Juncos describes him as "completely and willingly yields to all compulsions," and having "little control, very strong drive to perform [compulsive] behavior [that] must be carried to completion, [and] can only delay with difficulty."

Elliot's father complains about "cruel" emails that Elliot sends to his parents. When he shares their contents with me, they look like eruptions of dark thoughts. Also, Elliot has the habit of calling up his parents with announcements like, "Hi, I am leaving my wife," "Hi, I just saw an accident," "Hi, I have just lost my job." Obsessed with harmful events?

Even though his first tics as a child were vocal, his current tics are mainly motor. He still has "breathing tics" that he ranks as very frequent and very forceful, but he rates the rest of his vocal tics as infrequent and not forceful. They include throat clearing, sniffing, humming, and palilalia – repeating his own words. By contrast, many of his motor tics are frequent and forceful or even very frequent and very forceful. They include eyeblinking, facial tics, head jerks, neck and shoulder jerks, hand movements, tensing parts of the body, and touching parts of the body. His stomach jerks, arm movements, kicking legs, and touching other people are less severe --infrequent and unforceful. Like Elliot, Dr. Juncos ranks Elliot's motor tics as more severe than his vocal tics. They are multiple, almost always present, and complex. The vocal are less in number and in frequency. They might, however, be more intense than the motor. In summary, Elliot's tics are mildly-to-moderately impairing.

Elliot thinks that the shift in emphasis from vocal to motor tics, is just a "change," not necessarily an effect of his medications. But since he takes Orap, with possible side effect of akathisia (motor restlessness), it is hard to be sure. Despite the possible adverse side effects of the Orap, Dr. Juncos is reluctant to switch to another atypical antipsychotic "given that they [the other antipsychotics] are even more associated with aggravation of diabetes and hyperlipidemia," that Elliot is diagnosed with.

The penalties of TS and possible attention deficit disorder (ADD) caused Elliot to take almost a whole decade to graduate from college. And in spite of having a college degree and being bright, he cannot build a career, and he keeps serving at low-status and low-paying jobs. Currently he holds a job and also has a girlfriend. His greatest source of pride and joy are his boys. Occasionally he is worried that they would have TS, but mostly he enjoys them. The eldest started school a day before our interview, and Elliot was the happiest, most proud father. On such days, even the Tourette is less dark.

Chuck

(Finkelstein et al., 2007-08-24):

Chuck is in his early 40's, and newly married to a woman whom he met on the Internet. He remembers starting to utter "raspy-froggy" voices at a summer camp before his fourth grade. This unusual sound earned him the title of the "froggy man," and much attention, which he enjoyed. But then school started and with it, "You get to go to school and sit in the chair; you have to listen and follow directions; and I couldn't do it." The froggy voice, the throat-clearing, and the "eepie sound" were not funny any more. They got on the "nerves of everybody."

Soon, motor tics joined. Chuck describes a middle-school videotape with vocal and motor tics, "the whole gambit… the sniffing, the teeth, the head jerking, and the arms." But "the one that always really stayed with me is the vocal, the vocal tics, the noise." While the vocal tics stayed with him, their nature changed, and sniffing and barking replaced the frog-sounds. His tics do not include whole words but sometimes he utters syllables, "dra, or cha, or wa, bab, bever." Chuck observes that the balance between his motor and vocal tics differs from that of most who are diagnosed with TS. For him, the vocal came first; it is the more persistent, and the more prominent.

On his TSSR Chuck reports his motor tics – facial, head jerks, kicking, touching parts of the body, and picking at things – as infrequent and not forceful. But his only vocal tic – saying single syllables – is frequent and forceful, and often he has the need for it to be "just right."

He was diagnosed with TS at the 6th grade, about two years after the onset of the first signs. It took his mother, who felt ashamed and guilty, about a year before she could openly speak about her son having Tourette syndrome. By contrast, for Chuck, the diagnosis was "the best thing ever. It finally had a name… I was doing all those weird things and I'd get in trouble for it, yelled at… When I found that [it] had a name I went 'oh! That's what it is' So that helped me out a lot."

Ranking his Tourette's severity, he observes, "I'm definitely more than average. I get the vocal tics. Am I the worst? No. There are people with coprolalia," which he does not have. As for whether he knows that a tic is coming Chuck says, "I don't think that I notice that it's dra or cha. It just sometimes comes out. But do I know it's coming?... I'd

say 99% of the time I know it's coming. I have a little warning." Often, it's more than a warning; it's an urge.

He compares the urge to a mosquito bite. His tic, typically a sound, brings relief. "I make that noise, I feel better." But not for too long, "and then it comes back." Further exploring the nature of the urge, he rubs his chest and says, "Right here that's where it [the urge] is for me." Tapping his leg, he speaks about how a motor tic is triggered when something is put close to his leg. "It's [the urge] here." Then he continues to describe the 'just right' feeling that applies both to the motor and the vocal. "I got to hit it the right way. You got to hit it until it feels right. Kind of like the noise. You got to make the noises till it feels right."

Chuck's attitude to his tics evolved with time. "Now I'm at a point where I don't think about it every time, 'here's an urge, ok it feels better' or 'here's another one.' … I'm at a point where I know it's happening. I don't try to stop it, it just does its thing, that's it." But when I press him on whether he tried to suppress his tics during his first date with his wife, he says, "No. No. I mean, well, Ya." For a moment, the perfectly-positive shield cracks a bit.

Chuck is not treated by a neurologist, but he sees a psychiatrist and takes Tenex: "Well, I'm on medication. You still see what I see [referring to his phonic tics], I mean, you still see what I do. Was it worse before it? I am on vacation [from teaching children] for 5 years now. Was it worse before it? Ya, maybe. Is it better now while I'm on it? Maybe. Just now, I'm fine. So if it's the medication, I give the medication credit. I don't know. But you see with the life issues and jobs, you see that all plays a role. But what I think did happen when I got on this medication was -- I kind of think of it as a threshold,

where I have to get to this threshold (gestures with a horizontal hand motion to mark the threshold). Maybe 5 years ago I had it (gesturing) right here, had to be louder, I had it more frequently, the pitch was higher. When I got the new medication I felt like the threshold got lower. I still do it but I don't feel like I have to do it as much, or as tense, or get that whatever it is as high… Even on my worst days they're not as bad as my worst days were before, when I was off the medication."

With or without medications Chuck's tics get worse at stressful times, and better when he is in a comfortable situation and around people he knows. The tics reduce also "when I am really focused on something." He focuses when he writes, uses a keyboard or a mouse, or plays softball or racquetball. He also does not tic when he works hard, "I'm doing, I'm busy, and I'm work[ing], work[ing] with the kids… Sometimes I tell [my wife] that I don't have time to make my noises." Recently I had the opportunity to see Chuck in a weekend retreat for children with TS and their families. To close the 3-day retreat he oversaw a Family Olympiad with group races and games. With good humor, he managed the event, and for an entire hour ticced very little.

After graduating from college, Chuck insisted on becoming a teacher. And indeed, his ideas and techniques of teaching are inspiring. Equipped with the confidence that he has much to offer and that even though he has "disability or some weakness, [he] can still be successful," he went through 25 job interviews. Repeatedly he was rejected with, "We are not going to hire you. We can't hire you. We never had a teacher who has Tourette syndrome." He persisted. "I knew that the person who was going to hire me would reap the benefits. And we both did, I think. We've both had this opportunity and been working for 12 years now… So it's definitely the right decision and … once people

get to know you, it's not an issue, including parents." He is a successful second-grade teacher, and he is now a teachers' teacher. He applies his experience of being different to his understanding of life in general. "[If] I am teaching every single kid in that classroom the same exact way, then I'm not really doing justice. Because everyone learns in different ways." And he follows the philosophy of "We got to use every strategy as a teacher."

With a co-author, Chuck published a book about his TS and his teaching experience. He is as inspiring in his book as when he speaks about his teaching philosophy. His teaching is successful even though writing and reading are challenging for him. "Can I read? Yes. Do I read? Yes! Do I like to read? No! Because the tics get in the way." In graduate school he had to read much, but "I would never make excuses… [It] might take you one hour to read a chapter, might take me three, [but] it will be done." However, "Do I pleasure-read? No! [I] don't enjoy it." He explains the source of the difficulty, "When you start reading, you start ticcing, and half about the page you forget what you read. You start over again, you forget what you read. You do it again. After a few times of doing that, you forget what book you are reading. You look at the front and you say, 'Ah, yeah, I'm reading that book.' So no! There's not much enjoyment from reading for me." To make up for it, Chuck kept excellent class attendance, always sat in the front, and heavily relied on his auditory perception.`

While having some remarkable personal achievements, his attention is only partial; and so is his understanding. My questions are rarely answered to the point; more by association. To retain new knowledge he relies on the physical world, on first-hand experiences. For his exposure to experiences, he is grateful to his parents. "I was a hands-

on learner. I experienced it through life. My Mom, my Dad, my brother -- we experienced it, it's life. I mean, we weren't sheltered. It wasn't like 'stay at home.' It was more like, 'What d'you want to get involved with? You can do sports, you can do soccer, do baseball, basketball, you're going to be in camp, you're going to get out there.'" His learning included much traveling, "You want to learn about art, then you go to Paris, you go to Italy, and you go in those places and you learn." Then he repeats and summarizes -- his learning and achievements were through places, people, and doing. "Let's do something. Let's not just sit back." And when we close the interview I share my feeling with him, "You make me want to sit in your classroom."

Neil

(Finkelstein et al., 2007-08-22):

Neil, at his early 50's, is a psychiatrist who specializes in movement disorders including Tourette syndrome. He "admire[s] humor." Not necessarily in the form of being a clown or telling jokes but in the ability to respond quickly and wittily to a situation. This is how he describes himself and others with Tourette syndrome: "I'm … *lousy* at telling a joke... I would bet … that most of your Tourette patients… probably can't tell jokes that well, because we dwell too much and we tell too many details, worry too much about the correct word to say. But we can come up with something (snapping his finger) that's germane to the moment."

But humor has two faces. While it bridges to others, it separates as well, "I… think that humor can be a way of building walls… not getting connected. And I probably do that too much." And he further explains, "If you think of someone who makes you laugh all the time… you are not getting to know *them*, you're getting to know their

humor… It's not necessarily somebody you would trust with your inner thoughts." He proceeds to speak of others, whose humor is central in their human interactions: "Comedians, for example, are notoriously shy off camera, off stage, I think in general. And they don't necessarily make a lot of close friends … they get along really well with people but they don't necessarily make the strong bond connections, and I'm a little bit like that." He closes the topic with, "If I can change myself, that's a thing I would change."

His humor often challenges social conventions, especially when socially drinking, but otherwise as well.

> The worst was when I was new, bran new... practitioner, [and the hospital] have me speaking about something… I [started] the lecture… with what we -- all amateurs – do, start with a joke… I was telling the audience one reason I like [to speak about] movement disorders is because I have my own movement disorder. I have Tourette syndrome. I used to hate it but one day I asked my wife, 'honey… how can you stand living with me?' I am telling this to a bunch of nuns and other nurses, and she says, 'honey it's ok as long as you don't start saying fuck you bitch fuck you bitch.' As soon as… I got into the bitch part I said, 'I can't believe I just did that' and half the audience was in hysterics; the other half was staring at me like 'I cannot believe this man is up there. How dare you?'

And he summarizes, "I probably blurt out things that are mostly true, in settings that they shouldn't be said." He links this socially inappropriate behavior to impulsivity. "I used to be *very* impulsive... I would say things that are really witty but inappropriate."

His tics started when he was a child, maybe around the age of 8. He did not notice his excessive eye-blinking until "one adult [was] blinking back at me. I thought that was not a right thing to do (he smiles). I thought he was kidding with me… I blinked back at him and he was sort of mimicking me and I did not know I was doing that. " He also

sniffed a lot and his mother thought it was a form of allergy. He thinks the other kids and the teachers noticed but "they never mentioned it to me and my mother. You know that was back in the 60's, and people then knew much less about Tourette syndrome." When speaking more about his childhood, Neil's tics – sniffs, eyeblinks, taps his right hand on his leg, protrudes his tongue, and occasionally stammers -- although he describes a "gregarious and witty" child that had friends at least until high school. Since his father, an employee of the Federal government, had to move a lot, he always had only a small core of friends, but that's all he "cared about."

His tics are unilateral on his right. He uses his left for gesturing. In his TSSR he ranks his eyeblinking and facial tics as frequent and forceful. But his head jerks, stomach jerks, shoulder jerks, arm movements, finger movements, kicking the leg, touching parts of the body, and picking at things are infrequent and not forceful. He lists only two vocal tics, sniffing and squeezing air under his right upper lip. They are both frequent and forceful.

Among his compulsions he marks checking that he did not make mistake – a compulsion that he discussed when we discussed his writing. He also has counting compulsions and compulsive need to tell. Dr. Juncos ranks the interference of his compulsion with his work as mild, but the distress associated with his compulsive behavior as moderate; his resistance against a compulsion is moderate, and he has little control over his compulsive behavior. While his tics – both motor and vocal – are frequent and moderately intense, their interference with Neil's behavior is minimal.

At the age of 29, in medical school, he self-diagnosed his TS: "Then even neurologists knew little about TS." In his residency in neurology he received little help

and support. “During my residency I did talk to [Dr. X] who told me that I should just be calmed down and not worry about it and they will probably go away, which is odd because I was an adult by that point. I wouldn’t think that this would be likely… So [Dr. X gave me] good advice but not the right advice.”

In exploring the nature of his premonitory urge, Neil observes that he does not have an urge prior to his frequent eye blinking. But when he has the urge for other tics and tries to suppress it, it is irritating, like when a person, after touching “poison ivy, [is] told not to scratch.” He thinks it’s a good analogy, “You have an itch, you scratch it, it feels better, for a few seconds or maybe a few minutes and then it starts itching again. “Trying to suppress it, “you can do it but there is that thing that just grabs you and the feeling that wells up deep within.” He then brings another analogy to “an air gun. You compress the air a little bit and… pumping it then pushing it … But it lasts less than a second and in between I feel pretty good.” In some situations, despite the urge, he successfully suppresses his tics but tics more intensely later. “For example, I am not really suppressing now but I am a little bit and… when I leave here I probably will have more. And it’s not that it’s really conscious, I am concentrating more here. Probably after the concentrating, talking, I’ll be ticcing more often.”

His tics are more frequent when he is tired, “The worst time of the day for me is late afternoon after about 5.” But, unlike most other with Tourette syndrome, Neil says, “stress is not big part of it.” Sitting *quietly* in a conversation or in front of the TV exacerbates the tics. But in general “there are some days that I have no clue.” He thinks that he does not tic when he sleeps. His wife told me the same. When he concentrates --

preparing a PowerPoint presentation or dictating a medical report of a patient -- he tics less. His tics always diminish during physical exercise.

In social situations "I think I am trying to hide it. And I am hiding it by suppressing it or I'll rub my eye. That's sort of things." When he drinks socially the first two drinks bring relaxation and comfort. "[But] if I have [more] drinks sometimes it exacerbates [the tics]." He does not think that caffeine has any effect on his tics. He does not smoke marijuana "but I've heard that maybe it can be beneficial." He is not exposed to any other drugs and he does not take medications.

The only time that he did try a medical intervention was bad: "The worst thing I ever did was the Botox. It's draconian. I think I still have a residual of it." Since he tics only on the right, he injected Botox[61] to the right side of his face. He simultaneously froze and weakened this area but the tics persisted; they switched sides and moved to the left. "The weakness of the Botox probably lasted longer than three months; probably about four or five." And resolved to give up other medical interventions, he ends telling about this episode with a laugh, "You see that's a great story."

Even though Neil reads and says it is not hard for him, he often falls asleep while reading. In the past, when he had longer distances to commute, he used to listen to audio books. Books bring him pleasure, "I like reading." And he enjoys sharing his reading with his wife, who "adores reading."

Writing is more complex since it interacts with his obsessive-compulsive tendencies: "I don't like writing…. People say I am a good writer… [But] it's so hard for me. Probably some of it is that's obsessive… It just feels like I got to find the *right* word, and it's probably not *the* right word... When I write handwriting I'm scratching a lot. And

[61] Botox (botulinum toxin type A) relaxes muscles by blocking the release of acetylcholine.

so a lot of my notes are… illegible… Some of it is because I start a thought, writing a thought, and I change my mind. It's more my impulsive nature."

Neil shares with me some of his medical observations as a neurologist, especially about coprolalia. "My experience with coprolalia… is that it grows out of a cultural background. Often children from very religious families will start saying 'Fuck Jesus Christ' or you know that sort of thing, the very things they are not supposed to say… Of course I have patients that have complex vocal tics and say single words or phrases that are not coprolalia but it's interesting in that that it is forbidden fruit. Most people don't say it, but when they do, it tends to be towards the vulgar."

We speak some of his children, both teen agers. His daughter is diagnosed with OCD and his son with transient tic disorder. He hopes things will not get worse, "I am concerned whether they'll develop the phenotype." As for the family dynamics, "I'm pretty transparent about it…. It's beyond my control. It's something I'd rather not have. You know I'm pretty mild and I'm grateful for that but if it were worse I would probably feel differently. I'd rather not have it but I don't think it affects my relationships with anybody from my family. They are very good about that."

Danielle

(Finkelstein et al., 2007-08-17):

Danielle, 31, a plump woman with gentle, smiling face is 17-week pregnant with her second child. She blinks constantly and with no awareness of it, "like... talking or breathing." She is also unaware of her frequent facial tics until her muscles get sore: "With the facial ones, I don't even realize… until the muscles start being so sore." And

then she adds with a smile, “or somebody will start saying ‘you feeling OK?’ and then I know they’re saying ‘Your tics are getting bad.’”

Since early age, Danielle suffered anxiety. “I have so much anxiety and so much worry. Growing up, my parents called me worry-worry.” For example, when asked to remember something, she “would stay up all night long. [I would] go over it over and over in my head so I wouldn’t forget.” She is still very anxious. In addition she describes herself as “very compulsive.” For example, she stocks up produce: “If I use something up I have to have five spares in the closet,” for possible hard times. In her Y-BOCS Compulsions Checklist she reports compulsions in the past and the present. Some past compulsions disappeared but she still suffers from washing, checking, repeating, counting, and ordering compulsions. Dr. Juncos rates most of her obsessions and compulsion as moderate, but her resistance to them is lacking and she has only very little control over her compulsive behavior. She is driven by a need to carry her compulsions to completion, and can delay them only with great difficulty.

Her tics started at 4th grade "with a constant eye blinking." Barking followed. Then the facial tics came and the lip smacking. She was diagnosed with Tourette syndrome in 1990, at the age of 15. "It was nice to finally know what was wrong with me." Her signs continued to escalate: "Once I turned about 20… my tics have gotten worse over the years." The facial tics spread to other parts of her body -- arms and legs -- and finally to seizures that she calls "body tics."

Danielle was tested for epilepsy but the electroencephalogram (EEG) showed no abnormality. However, since the EEG was not taken during a seizure, it is non-informative, and the way Danielle describes her seizures leaves the question of their

nature open. “They [the body tics] almost mimicked the seizure. But I was conscious. I mean I couldn’t respond but I could hear what’s going on… My eyes for a while would roll up, kind of like to the back of my head, and I couldn’t see.” There were times when she had seizures up to 8-10 times a day, especially during her college years. But it is much better now.

Danielle thinks that the anti-epileptic Topamax (topiramate) has helped her greatly. Her last seizure was in December, nine months before our interview. She had to stop the Topamax during her pregnancy, but fortunately, “the pregnancy, it’s amazing how it changes my tics. This is the best my tics have been in a long time.”

She still experiences premonitory strange sensations, but seizures do not follow. “[It’s] a weird feeling… that [something] would happen.” It can be “just a little tingling,” or it can be “like aura... Sometimes I would almost feel like I was outside my body… Just this weird feeling before… Everything was going kind of slow. My face started tingling… Sometimes I’d feel… just funny… kind of numb-ish… Something was about to happen; and a lot of times my eyes would start kind of twitching… Lots of times I would fall.”

She learned to listen to these warnings: “If I knew [a body tic] was coming I would go ahead and lie down somewhere on the floor or wherever. Because there have been a few times where I fell down the stairs.” The seizures brought relief: “I felt so much more refreshed even though I was exhausted for a while.” But feeling exhausted is not limited to her seizures. The constant tics use up Danielle’s energies, and she needs a lot of rest. The rest is also required for her frequent migraines that occur 3-4 times a week.

At junior high school Danielle attempted suicide with an overdose of pills and with a razor blade. After these attempts, the razor blade was used often, "not that I'd do it real deep, just enough to get blood and almost that was the satisfaction. And then I would hide it so nobody could see it." Trying to understand how the razor blade provided comfort, she adds with tearful eyes, "I don't remember. Not really. I just remember feeling better; almost like that took the pain or did something for me… I don't really remember. Just remember doing it a lot." Her suicidal thoughts continued even when she did not act on them.

Two or three years ago a change for the worse took place. She was in constant pain and was administered morphine to calm the pain down. She also experienced pains in her hands that led to a few hand surgeries. Concomitantly she stopped taking care of herself. But all this reversed for the better after a flu. She stopped all her 23 medications. "I had some withdrawals because it was so sudden but I felt so much better after that… I felt tremendously better after that. " And then for a period of almost four months her tics disappeared. Alas, they returned.

She continues to have tics. In her TSSR she reports very frequent and forceful eyeblinking and facial tics; frequent and forceful head and shoulder jerks, arm and hand movements, and touching objects. Some of her vocal tics are also frequent and forceful – grunting, coughing, and repeating her own and others' words. But in spite of their severity, her vocal tics are not socially disruptive. For example, in church "I will do just the lip smacking and kind of noises but never any loud noise that is disruptive." Indeed, Dr. Juncos rates her vocal tics as much less severe than her motor ones.

Her memory is not good and her comprehension is limited to the literal. “I memorize exactly the way I write it.” She reads the Bible with her boy daily, but cannot repeat the Jonah story that they read last week every night. She wonders whether the medications play a role in this condition.

Currently the choice of her medications is dictated by the safety of her fetus. The antipsychotic Haldol, the SSRI Zoloft, and α-methyl-dopa are prescribed for her third trimester. After her pregnancy, Dr. Juncos recommends to resume the Topamax.

In spite of the waning and waxing of her symptoms -- her rollercoaster trip of suffering -- Danielle seems to have reached some peace. Maybe due to her current pregnancy, which “is the best my tics have been in a long time.” Or due to a combination of the right medications, marriage to a loving husband, the joy of her 7-year-old son, and having the conditions required for the much rest that she needs.

Nick

(Finkelstein et al., 2007-12-07):

In his interview, Nick, 55, displays some facial tics and head twitching. No vocal tics, but he says that he often sniffs. First, his Tourette seems mild. He reports in his TSSR a long list of motor tics that include eyeblinking, facial tics, head and shoulder jerks, arm and leg movements, tensing parts of his body, touching objects , and finger tapping; but describes them all as infrequent and not forceful. And with the exception of his frequent and forceful sniffing, his vocal tics – grunting, throat clearing, saying single words, coprolalia, palilalia, and echolalia – are also infrequent and not forceful. *Dr. Ju*ncos ranks the tics more severely than Nick does -- moderate in frequency and intensity.

"The real problem that messes up things for me are compulsive and impulsive things like the OCD," says Nick. And indeed, this is what he is mainly treated for. His OCD is treated with Anafranil. The beta-blocker Indernal (propranolol) is prescribed for his anxiety and migraines. And the statin Pravachol (pravastatin) lowers his cholesterol.

Nick's first tic was his head twitching. He was diagnosed in the early 1980s, in his late 20's, after his mother, with whom he still lived at the time, had read an article about Tourette syndrome in the newspaper. He and his mother suspected that he had TS, and it was confirmed in the local hospital. Haldol was prescribed to him in small dose but to no effect. The very process, though, helped him realize "that the tics… are not the problem," that it is his obsessive compulsive disorder (OCD), and the self mutilation, which might be linked to it.

When his compulsions take over, he violates social etiquette: "If I am guilty of one thing it's probably interrupting people. When I think of something, I got to say it *then*." This behavior cost him a job when he was "compelled or impelled to say just one more thing" in what turned to be his last conversation with his boss. Many periods of his life were spent without employment. Currently he works, "doing security, which is ok. I don't mind it. It helps me to focus a little bit, always observing things. Gives me false sense of being important… It's tolerable. I don't love it but I don't hate it either."

His compulsive behavior can be more serious than just saying the wrong things at the wrong time. It can be physical. "If I get mad… I've done stuff to release it sometimes… I've done stuff to release it… I used to break stuff." Breaking does not always need to be motivated by anger: "Sometimes I reach out and touch stuff (playing

with his pant's cloth between middle finger and the thumb of his right hand). It's crazy… I just sometimes squeeze it and sometimes I hold it too tight and I'll break it."

His "stupid" behavior has led him to playing with knives and to self injury: "I once cut myself in the lip… This was back in July 94… Sometimes I bite glass." His self mutilation can take other forms as well: "Sometimes even on a good day, just getting up, I use to put my fingers in fans and stuff like that…. Get the urge to do stuff. I know I shouldn't do it but I do it sometimes."

The Anafranil seems to help, "I'm getting better about that by controlling my temper." Being angry, "I'll just get out and walk around." Concentration and focusing help too. Yet he observes that the very focusing can be a form of obsession, "over focus." For recreation, he plays war games with friends and he believes he does not tic during a game since he needs to concentrate. His compulsions recede too.

Nick describes himself as a good observer of details but immediately adds, "Maybe I observe too much." His observation skills attracted him to photography. He took courses but somehow, "like in catch 22," never carried his desire to photograph further; never translated it into an action. He considers his current job in security as a good use of his observation skills. He only wishes he could apply his photography, though he does nothing to achieve it. The gap between his passion to photography and his lack of advancing himself in it is noteworthy: "[I] would like to do something with my photography. I like photorealism... but I haven't explored it. I probably should. .. Nothing seems to have happened with it. I need to go back to school and catch up on digital. Get photos; send them out in the field." Yet he does not.

Initially it is easy to be misled by Nick's laughter about his own weaknesses, and mistake his comments as stoic self observation. But as the interview proceeds the impressions change. His laughter covers self-deprecation. It seems like a mask, which serves to hide inescapable suffering and loneliness. Nick hopes that a job on a ship in Alaska, for which he applied, will come through and provide him with a supportive structure, good money, and opportunities for photography.

Claire

(Finkelstein et al., 2007-12-21):

Claire, 45, beautiful and chic, was born in Germany, not far from the Rhine. At the age of 22, on the train back from her work as a cosmetics salesperson, she met her husband -- an American who served as a civil servant in the military. They moved to England, after a few months to Italy, and after a few years to the United States.

Claire describes herself before adolescence: "I was a good student. I didn't have any problems in school, or… had Tourette's, or anything. I was normal, no problem. I was normal during my school time." But at the age of 15, when walking, Claire started to skip. This is the earliest that she remembers of a Tourette behavior. She attributes the triggering of her skipping to a "mean boss" that she had when selling cosmetics at the age of 16, even though her skipping started earlier, at the age of 15. Claire takes pride in her advancement at work and describes herself without TS: " [I] always sold cosmetics in a department store… where I lived. And [I was] normal; no skipping, nothing, no any signs of [TS], nothing. I started from the back [of the store] to the front."

She aligns the trajectory of her TS and its increasing severity with the trajectory of the changes in her life -- first moving to England, then to Italy, and finally to the

United States. But reflecting on the possibility of a causal connection, she says, “I don’t think so. I don’t think so.” In Italy “it started getting worse,” with a new behavior, “kind of screaming… [and] scratching myself.” The self mutilation got worse: “I got mad one time at my husband and I had a glass door… and I cut my artery here,” she points to her right inside wrist.

After returning from the hospital, she adopted a new life-style, which helped: “We started running -- 3 miles, 4 miles, almost every day -- and I felt good.” But she was not cured and continued to skip involuntarily. Her self mutilation stopped but her violence did not; it turned against her husband: “I was really mean. I was mean towards my husband. I scratched him up.” Life was isolated, “I didn’t have a car and friends; I couldn’t walk… into town. This is near the mountains about 40 minutes away from the village.” But despite all this, there is a nostalgic tone when she adds, “we liked it.”

The move to the States marked new changes. While her violence against her husband stopped, Claire started to kick the walls; often, so that they “need to be repaired, and I call somebody to come and fix it… [All our] money [is] going into sheet rock.” She developed fallings and screaming. Getting off her chair, she demonstrates a fall, at the end of which she is lying horizontal on the floor, facing down: “On the floor, like this, like this… and I lay there until I, see I’m out of breath almost, until I feel (stops for a breath) the right moment to get up, to get up.” She attempts to get up from the floor, but falls back on her knees, and continues, “and I fall like this and (getting up) hold myself.”

Then she describes the pattern of her screaming “three, four times, six times, until it feels right.” While getting off the floor from a demonstration that set off her real body tics, she adds, “Then I get up and I start all over again.” And returning to her seat, she

says, "It's always, constantly, constantly." Her frequent falling "don't hurt;" neither when she presses her fingers into her inflamed bursas, nor during the fall itself: "I don't fall on the bone I fall down here (pointing at the bursa) I fall here." But she does not credit the sparing of her bones to self control.

Claire's falling is often followed with piercing screams. "Sometimes I fell down on the floor (demonstrates) and then I just scream (screams)... Sometimes 2 minutes long: keeps, stops, keeps-on; ritual I guess is the word. And sometimes I can't breathe and then afterwards when I'm done I feel that my heart is gone little fast and I start sweating; start getting hot sweating."

Her screaming in worst in private: "When I am home alone it's more." But it's bad enough in public to keep Claire from going out. She does not shop; she limits her gym visits to hours with only few visitors; and she participates only in gym classes with small attendance. She used to be a body builder. This is a significant loss for her. Spending most of her time alone, she likes to sit in coffee shops, but has "been thrown out from several coffee places because of [my] problem."

In addition to her screaming and falling, Claire has coprolalia and copropraxia. Her "Nigger!" need not be triggered by the presence of a Black person. And she adds, "I have a lot of Black friend[s]." In response to my question whether it matters if she likes the person or not, she first screams and then answers, "O gosh! No it don't matter. It don't matter." She thinks her coprolalia is more intense in front of strangers. "I don't feel comfortable being with other people, and my tics coming on. I scream, I say 'Nigger!' or I don't want to be around other people."

She thinks that her other tics are also more frequent and intense around people. According to her husband, even her screaming is more intense among people: “We cannot go out. At the moment we enter a new place she brings her biggest screams.” He attributes it to their relations, “I think she is testing me.”

Many of Claire’s tics are set off when she describes them; so is her copropraxia. “When I, in the morning, get up (scratches her private parts and places her leg on her hand to stop it); when I get up in the morning, I touch myself (keeps touching herself). Or sometimes I try [it] on other people or other things.”

We discuss situations in which Claire tics less. “When I sleep,” she says. Even when awake there are situations in which she tics less. “I lift weights. Then I really don’t have it, my symptoms.” She also tics less cooking, which she hardly does any more. But cleaning her house, which is “spotless,” and washing the dishes bring no relief.

Does the visual world affect her? I ask her to shut her eyes for a short while. When she opens them I ask, “Does it feel more comfortable?” and she answers, “Yes, it does, it does,” and agrees with my suggestion that visual stimuli might trigger the tics. As for acoustic stimuli she says, “It [the noise] has nothing really to do with it [the TS].”

It seems her internal emotions drive Claire’s self mutilation and violence: “In Italy it was mostly when I was getting mad. When I got mad, or I got jealous, or got mad with my husband… It was not mostly the tic; [it] was mostly the other thing [the screaming and self-mutilation].” And in her attempt to understand, she resorts to “I was saying that… the devil is in me.” Later in the interview she returns to this image, “like when I see that movie *The Exorcist*… I ask, [who] put me… in the same situation,” even though she mocks the image and describes herself as non-religious.

Claire and her husband have tried to understand what affects her tics. “My husband always seems to say that coffee is the problem… I drink maybe 5-6 cups a day. Sometimes decaf, all depends. I don’t think it has anything really to do with it.” Her opinion is formed less by self observation than by a “psychiatrist [who] told me I could drink coffee, and it really don’t have anything to do with it.” She does not drink alcohol and never tried recreational drugs.

The intensity and frequency of her signs worsened in the States, “The worst things started since we moved to the States. [It] started about 10 years ago maybe… and since 5 years [ago it] increases more and more; got worse.” She speaks of Italy with yearning: “I liked Italy. I liked Italy more better than here in the States. We did more things; we had more friends in Italy and more to do than here. I said I want to go to the States, and now I don’t; I don’t like it here anymore. I want to go back to Italy. I had girlfriends, you know, it was just different.”

Recently Claire attempted again self-mutilation. “Two months ago I started a little bit [scratching] on my neck.” But “not like it was in England or in Italy, not like that… It seems like I was more possessed with doing it in Italy than here.” Then I ask her if she feels she has more control here. Her answer is, “It seems like; it seems like.” Her self-mutilation has stopped, or at least greatly diminished.

We speak about the urge. She struggles to articulate the process. While she knows that “something comes out,” she typically does not “really know what’s going to happen. But when it is self-scratching, sometimes to bleeding, she does know what is coming. However, she never attempts to suppress it: “The urge comes on and I have to let it out. It don’t matter if I’m here, there… or anywhere.” However, after more probing Claire

describes situations in which she tries to suppress the screaming. “I tried [to stop the screaming]. It’s hard, difficult to hold it in.” And she succeeds only “seldom, seldom.” When she does suppress the screaming, “I [later] let it out. When I get out the store then [I scream]… more.” When her urge is to fall, Claire also knows what is coming. Her premonitory bodily sensations are “all over,” and they start “from here” she points at her head. She explains it all with “the OCD. I can’t stop it. It has to be just right. Until it’s the moment I feel comfortable and then I stop.” And she adds, “I feel better afterwards.”

Her medical history is as complex as her behavioral history. Her parents already took her to a neurologist at a young age, but she remembers very little of it. In her early 20’s she tried hypnosis and Anafranil to ameliorate her obsessive compulsive behavior (OCB). She stopped them both. Since, she has also been diagnosed with attention deficit disorder (ADD) and anxiety disorder. And now, under the care of Dr. Juncos, she takes the neuroleptic Risperdal, the anti-epileptic Topamax, the SSRI Zoloft, and the Benzo tranquilizers Valium (diazepam) and Xanax (alprazolam). She and her husband are desperate and feel that their only hope for change is deep brain stimulation (DBS), which Dr. Juncos is trying to arrange for her[62].

There is OC behavior in Claire’s family: “My mother -- she got some… She is very clean... and she has a kind of depression also… She is just very clean, spotless.” Later she speaks of her maternal grandmother and emphasizes the possessive nature of her love: “My grandmother, my mother’s mother, she was kind of little bit [out of] order. She was crazy, when I was a child, about me. She wants to keep me, and she was kind of

[62] DBS for TS is still controversial and not officially approved by the FDA. As such, this expensive operation is not covered by medical insurance, and if not paid for by the patient, needs to be conducted within a study.

nuts, so to speak… She wants to keep me. She don't want to give me away… I am the only one. She wants to keep me. She don't want to give me away."

Away from her grandparents, parents, brother and sister, and two nieces, Claire takes her two dogs for walks. They "are used to" her falling and screaming and do not respond to them. The are "the only normally friends, so to say, I have." Her husband, though, describes her behavior with the dogs as abusive and thinks that the dogs are afraid of her and try to avoid her.

Donna

(Finkelstein et al., 2008-02-08):

Donna, 43, has an identical twin, whom she describes as "my love." Her sister is in a mental hospital, and Donna is bothered by the "horrible" quality of her treatment. She often returns to speaking about her twin, especially when she wants to avoid the topic of the conversation.

By navigating the conversation to her preferred topics, Donna gains some control. To achieve it, she employs her sense of humor and then complains about her small apartment, her lack of money, and her loss of gist for life. If stressed for an answer, she screams "fire," her most prominent vocal tic. This is how she describes it: "I can't stop it… It's like I sneeze… I have never been in a fire. And then sometimes I'll say [it] if I hear it on the news. I don't watch the news, pyromaniac or fire or gasoline. I have said that for years. I don't know why." In addition to "fire" it can be "fuck you all," or "fuck you all the urine," or "fuck you motherfucker." Her screaming is the most disruptive of her tics: "Living and working in an apartment, staying in an apartment with people above

you… Knowing that they are up there is making me nervous… [it's] like putting me in hell."

Donna describes her tics as frequent and forceful. Her most severe motor tics include eyeblinking, facial tics, kicking her legs, tensing parts of her body, touching other people, touching objects, finger tapping, picking at things, and obscene gestures. Touching parts of her own body and self hurting are infrequent and not forceful. Her vocal tics are typically more severe, and she describes them as very frequent and very forceful. They include screaming, saying a single word, coprolalia, repeating her own words, and repeating others' speech. Less severe, still frequent and forceful are her grunting, throat clearing, coughing, sniffing, and insulting others. In his YGTSS, Dr. Juncos ranked her impairment resulting from her tics as severe with association to self esteem, family life, social life, and holding a job.

Only some of her tics are displayed in her interview. She is restless, occasionally stands up, expresses her need to go to the rest room, but stays in, and finally sits down. Much of her behavior seems to be triggered from within.

To further explore what triggers her tics, we discuss visual stimuli. "What about the visual world? " I ask her. And she responds with, "I can't touch the mall. I never go into a mall." When I ask her to sit quietly and shut her eyes, she does it for over a minute without any ticcing, and when she opens her eyes she says, "It's relaxing." She also does not tic when she sleeps. But typically, even at home by herself, she tics "from morning to night."

Constantly troubled by the themes of her loneliness, small apartment, and lack of money, Donna often interrupts the conversation. When necessary, she takes license to use

a tic-like interruption so she can gear the conversation back to her obsessive thought. In one such occurrence, when I commented that "this was not a tic," she responds with, "I don't like to be told that but it's probably true." Her strategy of gaining control started early. Already at the 4th grade, "my mom and I were so close and I didn't want to leave her, and at that time I would fake sick." Her Dad was harder to convince since he, "did not have patience for us." For him, she had to perform, "like… in the academy awards." Fourth grade was also when Donna started to tic. She and her twin sister were moved to "special classes." Only when they were 16 years old, were they diagnosed with Tourette syndrome in "the most prestigious place to go for Tourette syndrome."

Donna realizes that her major challenge, more than her screaming, and more than her motor tics, is her obsessive compulsive behavior. "[As a child] I thought there was always something wrong." Her speech is interrupted with the scream "horrible all the urine or dirty urine*"* and then continues, "and I was already OCD at the grade school, and I thought I had cancer or I thought I was going blind… My compulsivity [was] the most that scared me, the OCD."

In the Yale-Brown Obsessive-Compulsive Scale, Dr. Juncos marks her at the top of the scale: An hour rarely passes without several obsessions; severe distress associated with her obsessive thoughts; extreme interference of her obsessions with her daily life; she willingly yields to all her obsessions, and has no control over them. Her compulsions are only slightly better: The frequency of her compulsions is moderate; its interference with her daily life is severe; so is her distress over it; like with her obsessions, she completely and willingly yields to her compulsions, and has little control over them.

Being a captive of dark thoughts and sense of helplessness, Donna is lonely. Already as kids, she and her twin "didn't have many friends because of that disease… It's a very isolated, horrible, crippling disease." By contrast, when she lived in the Mid West and had a boyfriend, her life was good despite the relationship being "tumultuous." She speaks of this time with longing and pride. "When I was dating my boyfriend, who looked like Tom Selleck, I was very calm around him. He worked at human service where I worked … I should have brought a picture. I was quiet with him." Her screaming and restlessness were not prominent then, "we went to movies, three-hours movies... and went to dinner when I met his family. He was very calming. I hardly had any noises around him at all."

But now, "I have so much pain, I cry every night. I'm always alone. I'm sick of being alone. I don't have a life." Her life was not always that bleak, and she used to enjoy physical activities at which she is good. "I'm a good tennis player… I play very well. I am a good ice skater. I'm a good horse-back rider. I like horses. In [the Mid West, I was] snow mobiling. Not my forte but [it] was exhilarating." She now dreams of better life when she will use her free air ticket to return to the Mid West. She also dreams of writing a book describing her life with Tourette syndrome, but meanwhile she is feeling lost:

> I have to try to see what's going to come next because there is just so much that a human being can take… I'm not saying that's what I'm going to do. I won't kill my mother or my twin sister. I'm saying that would kill them… It just that I can't be strong anymore. I've been the strong one all these years and it's sliding down. I can hardy drive anymore… I'm living somewhere I don't like… I don't have a job. I can take that type writer or the computer, not break it (laughs) and type and do work. Somebody should hire me and be lucky to have me as employee and

> take a tax credit for it or whatever, but they don't. I try and I try and it just it doesn't happen.

There is a great discrepancy between how Donna sees herself and how she is seen by others. She laments that, "It's a curse to me to have this [the TS] because it doesn't show the real me." But her perception of the "real me" is not always realistic. For example, while she dreams about writing a book, the page that she writes for me is hardly intelligible.

Neither does she read, "I do not read any more. I read magazines but mot much. I can't concentrate. I used to read books… My time is spent pretty useless." Like many others with OCD, Donna finds reading challenging and is poor with math and money. "[I am] horrible… budgeting." And later she adds, "I'm not good with math." Nevertheless, she maintains some independence. "I buy my food stamps, [and] I do my rent." Her money is insufficient and "Mom has to supplement but she doesn't have any more."

Being "sedentary," Donna developed osteoporosis. She also suffers from Crohn's disease[63] that causes her to throw up a lot. To these ailments she responds with increased drinking, which "intensifies [the tics]." She also takes her medications capriciously, sporadically, and arbitrarily. She stopped taking the neuroleptic Abilify, the antidepressant Cymbalta (dulexitine), and the mood stabilizer Topamax because "it did not work." However, she tried them only for 2 weeks and at very low doses. So their real impact is impossible to assess. She admitted herself to a psychiatric department where they thought that perhaps she might be abusing Ativan. But according to Dr. Juncos, "She has never exhibited drug-seeking behavior… It is the only medication that has consistently helped her and she uses it primarily for sleep."

[63] Crohn's disease is an inflammatory bowel disease.

Based on her past experiences with the medications, Dr. Juncos recommends that Donna resumes the Abilify and Cymbalta; add the antidepressant Effexor (venlafaxine); and meanwhile hold off the Topamax. She resists any possible solution, including a job that I had arranged for her working with horses. There is no way out of the vicious circle in which she is imprisoned. She realizes it, "I don't think I am letting anything helping me," but excuses herself with "it's too late," and adding later, "I want to die. I can't go on with it."

Louis

(Finkelstein et al., 2008-05-30):

Louis, 39, has recently moved to Atlanta from out-of-state. The local TS association recommended Dr. Juncos, and this is how he joined this study. He is happy to contribute since "it's nice to see that they are focusing some more on adults." Louis is a registered nurse with a masters degree in public health. Paul, his partner of 20 years, is a school counselor. Their 5-month separation, when Louis moved to Atlanta for a better job, was stressful; but they are now together again and on their way to financial and emotional recovery.

Louis was diagnosed with TS in 1988 at the age of 20. At that time there was already much in the media about the syndrome and "I knew that's what I had." As for many others, the diagnosis brought him relief, "I guess I always thought that I was a freak, or something was *really* wrong with me... 'cause kids made fun of me… I always made good grades but I got made fun of a lot, ridiculed a lot… a freak or weirdo… It was so nice to know that everything that I did, there was actually a diagnosis for… that other people did the same thing… and I think when I was diagnosed, it was sort of like a

weight that was totally lifted because I knew that I wasn't abnormal; that I wasn't all by myself."

His tics started early, "I've had tics ever since I can remember." He used to blink his eyes a lot, and he sniffed. He also remembers "twisting" his hair, and "knot and pull them out." But even though his trichotillomania left him with "some bear spots in [his] head," and "hurt sometimes," he "never really had self mutilation," he says. His father, who divorced his mother and remarried, treated his hair-pulling like a bad habit; maybe because he "never listened to my own [problems]." His paternal grandmother suggested to Louis that "[his] dad… had Tourette's." And, Louis says, "he still might have it." Neither his older sister nor younger brother have TS.

His eye-blinking greatly diminished when he went to college. His trichotillomania has disappeared too. But he still sniffs occasionally. Whether this is the waning of his TS in a waxing-and-waning pattern, is not clear. One tic persists – a wrist jerk. "The thing that I've had the longest is the wrist jerking. Seems like I've always have that. It's one thing that never decreased." This is a tic that impairs his ability to perform some nursing tasks. "I used to do clinical nursing back when I was working in the hospital, but it got to where I was feeling that it was hard for me to do because my hand and my wrist jerk… A lot of nurses would not go with me because I fling syringes up out of my hand." And this is why now most of his responsibilities are administrative.

In his TSSR he ranks his shoulder jerks and arm and hand movements as frequent and forceful. His eyeblinking, facial tics, and head and stomach jerks as infrequent. This is also how he ranks his self hurting and tooth clinching.

His vocal tics are more severe than the motor and disruptive, especially at work. Some of them he ranks as very frequent and very forceful – saying single words, coprolalia, palilalia, and echolalia. But to a lesser extent he also displays grunting, throat clearing, coughing, sniffing, and humming. His coprolalia emerged only about 2 years ago. Louis attributes their emergence to unusual stress in his life. "Some of it was the stress of moving here by myself because [Paul] was not here for the first five months. He had to finish his contract with the school district... That plus the manager I have with my first job was a total… just a huge life change. But my mother passed away about 3 years ago. I was very close to her. Just everything, I guess, combined. It has probably a lot to do with why I developed more vocal-type tics."

His vocal tics include "jibble, shibble, jibble juice, gerbil juice, and jerley jibble." But he also utters "shithole" and "shithead." He elaborates about his "Kwanzaa" and "Kwanzaa juice" explaining what brought the word to his attention. "The reason… why I started to say that Kwanzaa word, because I have a friend that worked with a lady from Nigeria. She was telling me that the holiday was not an African holiday." But despite the racial charge of the word, there need not be an African person present to evoke Louis's "Kwanzaa." He further adds, "Not that I'm trying to be racist or something, but it just comes out sometimes." Nevertheless, when I ask about possible racist feelings that might accompany his utterance, he answers, "Mmm, maybe," and then hurries to align himself with his African friend, who feels apart from African Americans. "In Algeria she has a lot of problems with African Americans. Here there's a big divide… and when she and I had this conversation [about the authenticity of Kwanzaa] I felt more passionate about that it was not really a holiday. That's why I say it more."

His tics are most frequent and intense at the safety and comfort of his home: "A lot of times in the evening, when I come home is when I let loose," and when he is with Paul, by himself, or when he was with his mother. When we further explore Louis's public behavior, he says, "Most of the time [I succeed in suppressing the tics in public], but usually in the mall or somewhere where there's lot of noise going on, I may slip words sometimes. When I know it won't be heard, [when] there's a lot of background noise going on."

His current job provides some relief. Working less with patients and more with papers, brings Louis some peace, even when other people are involved. "At work I don't do it because it's quieter and I'm not distracted. I sit at a desk most of the time… If I'm with a group of people or anytime when I'm with a group of people… I don't do it. I'm able to suppress it. Lot of time… I don't even think about it."

His insights about the premonitory urge are limited to his wrist-jerking tic, "With the wrist… it feels like I have to do it." Later he adds, "I guess I'm thinking about it [the wrist jerking] and doing it in the same time. But when I get home in the evening, when I walk through the door, it's like (shapes his hands as though holding a ball and exhales loudly)--- Because I'm trying to suppress it the whole day, it's like it's going to come out. And I feel like I *have* to do it." It seems that Louis's ability to suppress most of his tics in public is concomitant with the limited presence of an urge to tic when he is in public. The urge emerges mostly in the privacy of his home. And then, Louis feels that he should let the tic out or "I'm not going to feel better." And indeed, after "letting it out… I feel better."

In recent tests, his testosterone level was found to be low and he gets injections every two or three weeks to raise it. Dr. Juncos, however, is concerned that it might aggravate the tics, as is reported in the literature of exogenously administered testosterone. For his tics he is treated with Tenex, with no adverse side effects; but also with no amelioration. His dose was increased, produced some sedation and no benefits. The dose then was reduced to its original level, and will continue to be gradually reduced to zero, if possible, as his tics are stabled. As for stimulants, he does not know how caffeine affects him since "I've never gone off caffeine to really know." He observes that when drinking alcohol, which he does socially, "my tics are better…. It's easier to control them at that point." And when he smoked, before he quit nine years ago, his "tics were worse."

His OCD is described by Dr. Juncos as "minimal, not impairing." Louis describes it as: "I don't really feel like I have the OCD very bad. The biggest thing with the OCD is that I have to finish before I can start with something else." During the interview, he repeats each idea many times. His unit-of-repetition is not an individual word but rather a whole sentence or even a short paragraph. Often the stream of words, expressing the same idea over and over, stops only with an interruption of a new question.

In his Y-BOCS Compulsions Checklist he reports of past compulsions of ordering and saving. But they disappeared. Instead he has now checking compulsions and a ritualized eating behavior in which he "must finish one item on his plate before going to the next one." He attributes his uncontrollable over-eating, which caused him to get overweight, to his OCD too.

Louis is spared of many severe signs of TS. His mathematical abilities are good; he reads many pages at his work and comprehends what he reads; and he handles money responsibly. But even though he has a partner and they have, he tells me, many friends, he expresses a sense of loneliness and wishes there was an adult TS support group in Atlanta – a place where people would be able to share difficulties and solutions and enjoy the comfort of being like others.

Daniel

(Finkelstein et al., 2008-06-04):

Daniel, 18, underwent two procedures of deep brain stimulation (DBS): In his first operation, which was performed in November 2006, the electrodes were placed bilaterally at the antromedial globus pallidus internus of the basal ganglia (amGPi), and ameliorated his self-mutilation and coprolalia. However, there was no effect on his severe motor tics; thus the second DBS in August 2007. In it, electrodes were placed bilaterally in the centromedial thalamus. This DBS greatly ameliorated his tics and further ameliorated his coprolalia. Currently Daniel has 4 electrodes in his brain but the intention is to relieve him of at least two of them through a gradual change of the electrode tuning.

Prior to his first DBS, Daniel got, according to Dr. Juncos, "industrial" doses of Haldol (haloperidol), as well as "higher-than-optimal" dosage of Risperidal for his OCD, and the mood stabilizer clonidine. His most frequent motor tics were twirling and forcefully touching the floor by bending over. This caused him a chronic lower back pain. He experienced anxiety; was involved in self-mutilation like hair-pulling (trichotillomania); and compulsively needed to touch objects and be involved in ritual repetitions of writing.

The onset of his TS, what Daniel's mother referred to as its "surfacing," happened at the age of 7, with eye blinking and mild tics that seemed like bad habits. She noticed, though, from birth that "he was different." For example, he would not fall asleep unless he tightly pressed his finger-nails under hers.

Unlike with all the other participants in the study, Daniel's mother was present in his interview, very protective, maybe over-protective. She observed that a bad turn happened at the third grade with severe chicken pox, after which "he has never been the same." Her theory is that there must have been a genetic disposition (one of her brothers is schizophrenic and the other tends to have rage attacks), and that the chicken pox boosted it to a severe TS. Daniel's two teen-age younger brothers are healthy, physically active, and academically excelling.

Even before his first DBS, Daniel enjoyed some good moments. When he concentrated (for example, reading instructions for a new video game) he did not tic. According to his mother, he typically does not tic when he sleeps. When he was younger, if he ticced in his sleep, his parents woke him up and asked about his dreams. He dreamt that he was ticcing. When the urge to tic is great and insuppressible, Daniel goes to his room, where he has the license to hit his bed, scream, or curse until he is relieved.

The cursing is the greatest embarrassment for the whole family, and they never use the word "coprolalia," only "vocal tics." The vocabulary of Daniel's coprolalia is defined by his environment. Since third grade, he has been home-schooled in a Christian fundamentalist family. With only very little exposure to other kids, he had also minimal exposure to cursing. The worst he knew was the family-forbidden "shut up" and "doodoo." These words made up his coprolalia. As he grew up and started to meet other

kids, the vocabulary has expanded. However, neither Daniel nor his mother would tell me or write down for me what it is. Like with his other tics, Daniel's coprolalia can be controlled for a short time but then it is uttered with high intensity and frequency.

After the first DBS, the whole family was greatly relieved. The "vocal tics" disappeared. The only one that has stayed is, according to his 1/8 Cherokee mother, the "Indian tic." When Daniel gets excited, typically when he is happy, he gently hits his lips with his hand to generate a loud "Whhooooooooo" call. It is tolerated since it does not offend the family's values; tolerated but annoying.

After the second DBS, Dr. Juncos evaluates "that he has had significant improvement… He is no longer doing the hitting, the forceful touching of the floor by bending over. He is not doing the twirling, the self-mutilation, the anxiety; all these things have remained improved. He still does a little bit of the kicking of the legs." He does not twirl anymore. Occasionally he kicks with his foot, but his walking is upright and quite stable and with no bending over.

Accordingly, his medications, their doses, and schedules have been revised. These revisions, as well as the tuning of his electrodes, are an ongoing process, which depends on Daniel's response to his treatment. Currently he takes nine medications: The mood stabilizer clonidine; the antipsychotics Risperdal and Seroquel; the neuroepinephrine reuptake blocker Straterra for his attention deficit; the anti-epileptic Keppra and Lamictal (lamotrigine); the antidepressant bupropion; the Bezo tranquilizer clonazepam; and the neuroleptics haloperidol.

Daniel is still sleepier than healthy people, most likely in part because of his medications. For example, following a visit to the museum, he slept a whole day before

he could return to his usual routine. He gets up late, close to noon time. But, as Dr. Juncos points, he performs chores at home; reads more than before, since he has less tics that interrupt his reading; and he studies mathematics and science with the help of his mother, who used to be a teacher, his father the chemist, and his two younger brothers.

SUMMARY

The 16 people who are introduced above and their relatives gave me their time and confidence, and are my Tourette's teachers. While there is much that is common among them, they also vary significantly, each with his or her idiosyncratic phenotype. To minimize my interference with their voices, I have not shared my own observations in this chapter. In the following chapters I will study their stories closely, analyze their behaviors, classify their tics, study their success and failure in developing coping strategies, reflect on the involuntary nature of their disorder, and attempt to capture the complexity of their syndromes.

TIC CLASSIFICATION: OBSERVABLE BEHAVIORS

Classification is really a process of reasoning…When… we think of the objects compared, we classify; when we think of the relations betwixt them, we reason… thus: In investigating cases of nervous disease, we are really classifying them or their symptoms. From all points of view classification is of vast importance.

J. Hughlings Jackson in the "On the scientific and empirical investigation of epilepsies" *Medical Press and Circular* 1876.

CHAPTER CONTENTS

IN THIS CHAPTER I classify tics. First I reflect on the importance of such a practice. Then, with the background of the literature and studies of others, I focus on the population of my study and its phenotypes. I classify in three ways: by tic-modality, by tic-complexity, and by tic-triggers. The modality and complexity are observables and are discussed here. The classification by triggers, informed by subjective reports of the participants, is left for the next chapter.

ON CLASSIFICATION

Studying categorization and the construction of concepts has been central in cognitive psychology for more than half a century (For example see Medin, 1989; Rosch, 2002). In literature, Jorge Luis Borges describes the imaginary whimsical animal taxonomy of the "Ancient Chinese Encyclopedia":

> On those remote pages it is written that animals are divided into (a) those that belong to the Emperor, (b) embalmed ones, (c) those that are trained, (d) suckling pigs, (e) mermaids, (f) fabulous ones, (g) stray dogs, (h) those that are included in this classification, (i) those that tremble as if they were mad, (j) innumerable ones, (k) those drawn with a very fine camel's hair brush, (l) others, (m) those that have just broken a flower vase, (n) those that resemble flies from a distance (cited in Lakoff, 1987/1992:92).

This classification is very different from ours and evokes a world organized and constructed totally unlike our own.

An example brought by the evolutionist Stephen Jay Gould makes the point (1983): Biological taxonomy can be phylogenic -- by clades, which are defined by common ancestory; or it can be phenetical -- by overall similarity, usually in morphology. The two do not necessarily converge: Phenetically, apes -- gorillas,

chimpanzees, and orangutans -- share looks and behaviors distinct from those of humans. Phylogenetically, however, humans are grouped with chimps and gorillas and are "between" them and the orangutans. Does the class of apes exist or not? The answer is, It depends.

While classification supports understanding and communication, it puts us in danger of reification. "Apes" is not an entity and should not be reified. When we speak phenetically, the concept of ape is useful, a good shorthand. When we speak phylogenetically, there is no room for this concept. All classifications are dynamic processes. So are the classifications of syndromes. And like all classifications, they are ever-changing and never final.

Tic classification

Classification and naming are intimately linked. This is reflected in the detailed and influential tic classification of Meige and Feindel, *Les Tics et leur Traitement* . They observed that "[w]ant of precision in words leads inevitably to confusion of ideas and endless misunderstanding. In this respect the word tic is a great culprit" (Meige & Feindel, 1907/1990:xiv).

Leckman, King, and Cohen (1999) classify tics anatomically. Meige and Feindel, however, consider anatomical classification as appropriate for spasms, in which "neighboring muscles are apt to participate as well" but not for tics. They go further and propose to use the proximity of the muscles involved as a diagnostic criteria differentiating spasms from tics. They observe that when tics involve more than one muscle group, the association of the groups "is rarely anatomical." They therefore

consider it "advisable to name a tic... from the *functional* act of which it is" (142, my italics).

I propose to explore in this study three classifications of tics: (i) by modality, since some of its refinements are still debated, and yet essential for diagnosis; (ii) by complexity, since greater refinement than is currently applied would add new insights about the disorder; and (iii) by what triggers or suppresses the tics, since this is not studied much and may add to the understanding of TS and to possible interventions. These three categorizations can be thought of in terms of input-output; or, in biological terms, sensory-motor. Triggering or suppressing a tic is its input. The modalities of the tics and their complexities are their output.

It is tempting to follow the input-output metaphor and start with the input, then describe the output. But such a chronological presentation can mislead as though we know the etiological causality of tics. To avoid such a causal association I start with the output. The modality and complexity of a tic are *observable* and can be considered and analyzed even without consulting the patient. I will discuss them in this chapter. The triggers of tics require patient reports. Some are symptoms, others are hypothesized based on observations. I will discuss them in the next chapter.

My list of tics is far from being exhaustive. As Meige and Feindel acknowledge, it is impossible to "have collated every known case observed up to the present" (1907/1990:142). My discussion is limited to tics of the participants in my study. Some were demonstrated in the interviews, others were reported by the participants.

The participants

For completion, I briefly repeat here the description of the participants. Sixteen adults, who have met the DSM-IV criteria for Tourette syndrome (TS) participated in this study. Fourteen of them brought another close person -- referred to as *partner* – who was an immediate relative or a close friend. Out of the sixteen adults with TS (ATS), thirteen were male. The participant ages ranged between 18- 65, and averaged 39 years. They represented a spectrum of severity from simple motor and vocal tics to coprolalia, copropraxia, and self mutilation. All but two were being treated with medications. Two had undergone deep brain stimulation (DBS).

In studying this population and attempting to derive hypotheses that generalize to the larger TS population, it is important to remember Shapiro's warning of *ascertainment bias:* "[T]he described [pathology] might have characterized a particular patient but was unrelated to Tourette syndrome. The logical fallacy was to generalize the results from a single patient to all patients" (cited in H. I. Kushner, 1999:118). With this possible bias in mind, the high-severity phenotypes in my study might represent a sizable but only a portion of the adult TS population. For the rest of this dissertation, I will reserve the label ATS for this population that is represented by my participants, but whose size and percentage in the TS population are yet unknown.

THE MODALITY OF TICS

For the diagnosis of Tourette syndrome (TS), the DSM-IV-TR requires both motor and vocal tics. Such a distinction seems easy, almost trivial. But a closer look reveals some complexities.

Motor tics

Motor tics, performed by the motor system, can be simple or complex, as we will see in the classification of tics by their complexity. But regarding their motor nature, they can belong to one of three types: (i) Clonic tics -- brief, rapid, jerk-like, and typically last less than 100 msec. Examples of clonic tics are excessive eye blinking, nose twitching, and head and neck jerks. As noticed by Meige and Feindel (1907/1990), they are the easiest to recognize. (ii) Dystonic tics -- characterized by their twisting or gyrating motions. They are typically longer than the clonic, lasting more than 300 msec. Shoulder rotation and oculogyric – circular movements of the eyeballs -- are examples of dystonic tics. (iii) Tonic tics -- prolonged, lasting more than 500 msec. They include muscular contraction against resistance with marked increase in muscle tone. Abdominal and limb tensing are examples of tonic tics (Jankovic, 1997; Jankovic & Kwak, 2005).

Distribution of motor tics – Descriptive statistics

The Tic Severity Self Report (TSSR) questionnaire lists 18 motor tics[64]. Table 6.1 presents the motor tic distribution among the 16 participants. The severity values are: (1) infrequent and not forceful, (2) frequent and forceful, and (3) very frequent and very forceful.

I found only one study detailing tic distribution by modality. This study had 666 participants with "lifetime tics," and was conducted by Shapiro and colleagues (Shapiro, Shapiro, Bruun, & Sweet, 1988:127-143). Some differences between their study and mine should be mentioned: (i) the ages of the Shapiro et al participants ranges between 4

[64] For the questionnaire, see Appendix I

and 69 years with a median of 14. As such it presents a population different from my adult-only population; (ii) since the onset ages of the participants in the Shapiro et al

jID	Patient	1. blink	2. facial	3. head	4. stomach	5. shoulder	6. arm	7. fng_hnd	8. leg	9. tens_bdy	10. tch_bdy	11. tch_oth	12. tch_obj	13. str_actn	14. hrt_self	15. fing_tap	16. pick_thg	17. copro_x	18. mot_mis	mot_max	mot_tot	mot_cnt
1	SRA		1	2				1					1					1	3	3	9	6
2	LRA	2	2	2	1	2	1	3	1	3	3	1	1	3	3	3	3	2		3	36	17
3	TQA		1	1		1	1	3	1	1	1	2	2				2	3	1	3	20	13
4	KGA	3	3	3			1	1	2	3	1			1		1		3	1	3	23	12
5	DQA			1			1	1	1			1	1					1		1	7	7
6	HBA		2	1		1	2		1	1	2	1	3	3	3		2		3	3	25	13
7	SZA	1	2	2		1	1	1		1	1						1	1		2	12	10
8	EEA	2	2	3	1	2	1	2	1	3	2	1							3	3	23	12
9	CBA		1	1					1		1						1			1	5	5
10	NFA	2	2	1	1	1	1	1	1	1	1						1			2	13	12
11	DLA	3	3	2		2	2	2	2	1			2			1	1		2	3	23	12
12	NYA	1	1	1		1	1		1	1			1		1		1			1	10	10
13	CFA	2	2						3	2	3	2	1				2	2		3	19	9
14	DGA	2	2	1	1	1	1	1	2	2	1	2	2	2	1	2	2	2	2	2	29	18
15	LLA	1	1	1	1	2	2	2		2	1					1			1	2	15	11
16	DGB						1	1	1	1	1			1		1				1	7	7
	Count																			36	27	17
		10	14	14	5	10	13	12	13	13	12	7	9	5	4	6	10	8	8		6	4
N=16	**%**	63	88	88	31	63	81	75	81	81	75	44	56	31	25	38	63	50	50			
N=666	**% Sha-piro**	80	36	47	19	55	19		7		13	11										

Table 6.1: **The motor tics** in the all-adult study (N=16) compared to Shapiro's study (N=658). The tics tabulated here are: (1) eyeblinking, (2) other facial tics, (3) head jerks, (4) stomach jerks, (5) shoulder jerks, (6) arm movements, (7) finger or hand movements, (8) kicking leg movements, (9) testing parts of the body, (10) touching pars of the body, (11) touching other people, (12) touching objects, (13) trouble starting actions, (14) hurting self, (15) finger or hand tapping, (16) picking at things – clothing, etc., (17) obscene gestures – copropraxia, and (18) other motor tics. The last three columns are summaries: mot_max is the maximum tic-severity of each probands (1 is the mildest, 3 is the most severe); mot_tot is the combined total tic severity of each probands; and mot_cnt is the number of kinds of tics of each probands. For more details see the text.

study ranged between 2 and 57, those whose onset was after the age of 18 would not meet today's DSM-IV diagnostic criteria for TS. In my study, all the participants meet DSM-IV criteria for TS; (iii) Shapiro and colleagues used the Movement Disorder Questionnaire (MDQ), which groups the tics both by modality and complexity. The TSSR, used in my study, groups the tics only by their modality – motor or vocal; (iv) not all the items in the TSSR questionnaire have counterparts in the Shapiro et al study; (v) and finally, the Shapiro et al study has 666 participants, unlike my small, pilot study. With the above qualifications in mind, my study suggests an ATS severity much greater than that of the general TS population as represented in the Shapiro's study.

Vocal and phonic tics

It seems obvious that motor tics involve movements and vocal tics involve sounds. However, the recently added term "phonic" to describe some tics points beyond a simple distinction between movements and sounds. Regrettably, many use "phonic" and "vocal" interchangeably, but they are different. Jankovic and Kwak (2005) define phonic tics as "vocal utterances that result from contractions of the nasopharyngeal and the oral passageways, [and] are yet another type of *motor tic*." (2005:174, my italics). Swerdlow echoes this understanding: "Many vocal tics are better described as 'phonic,' and result from *motor* event (ie, a contracting diaphragm moving air through the upper airways)." (2005:329, my italics). Swerdlow goes farther: "Eliminate the distinction between vocal and motor tics and TS is pared down to a disorder of motor tics." (2005:329).

The experiences of the participants add to the complexity. When asked in his interview about the relations between vocal and motor tics, Steven answers, "The grunt is like a mixture" (Finkelstein et al., 2007-09-28), describing a tic that is simultaneously

vocal and motor. Following Jankovic and Swerdlow, I will refer to any non-verbal sound (e.g., barking) as phonic and to any verbal expression -- a word, a sentence, or a phrase -- as vocal. When this distinction is immaterial to the discussion, I will use vocal as more general including both the verbal and the non-verbal.

In public the TS behavior is always disruptive, but more so for vocal than for motor tics; they are harder to ignore and cannot be contained in a small area. In *Front of the Class*, Brad Cohen, a second grade teacher who suffers from TS, tells his readers: "With TS it is very difficult for me to go to a movie because my barking noises distract people who, like me, have paid their hard-earned money to watch the movie." (Cohen & Wyscocky, 2005:35). He also tells of an incidence that happened to him in a restaurant, "One day I went to lunch… with two colleagues from school… We hadn't been there very long before the manager came over to inform us that we would have to leave if I continued making noises." (200). When Steven went away to college, his parents built him a soundproof booth so he would be able to scream without being disruptive to the other students. "I had to find some privacy… That's when the soundproof room came in." (Finkelstein et al., 2007-09-28). For Donna, even being in her apartment is a challenge, "living and working in an apartment, staying in an apartment with people above you… knowing that they are up there is making me nervous… [knowing that they are hearing me is] like putting me in hell" (Finkelstein et al., 2008-02-08). Because of her screaming Claire has been asked to leave "several coffee places" (Finkelstein et al., 2007-12-21).

Distribution of vocal tics – Descriptive statistics

Based on the Tic Severity Self Report (TSSR) with its list of 16 vocal tics, Table 6.2 presents the vocal tic distribution among the 16 participants. The severity values are:

(1) infrequent and not forceful, (2) frequent and forceful, and (3) very frequent and very forceful.

As with motor tics, the comparison between my study and the TS life-time population study of Shapiro and colleagues (1988) is not complete since the lists are

ID	Patient	19. grunt	20. thrt_clr	21. cough	22. sniff	23. whistl	24. hoot	25. bark	26. gulp	27. hum	28. syll	29. word	30. copro_l	31. rpt_own	32. rpt_oth	33. insult	34. voc_mis	voc-max	voc_tot	voc_cnt
1	SRA	2	2	2	3		2				1		1	2	2			3	6	4
2	LRA	3	3	2	3	1	1	1	1	1	3	3	3	1	3	1		3	30	14
3	TQA	2	1	2	2		1	2			3	3	3		2	2		3	18	11
4	KGA	2	2	3	3	1	3				1	3	3			1	3	3	14	9
5	DQA	1										1	1					1	6	6
6	HBA	1	1	1	2			2	2		1	2	1	3	2		3	3	22	11
7	SZA		1		1	1			1	1			1			1		1	7	7
8	EEA		1		1					1				1			3	3	16	9
9	CBA										2							1	3	3
10	NFA				2												2	1	8	9
11	DLA	2	1	2	1					1				3	2			2	15	9
12	NYA	1	1		2								1	1	1			1	7	7
13	CFA	3	2	2	2			1			2	2	2	2	1	1	1	3	15	7
14	DGA		2	2	2			1				3	3	3	2	2	2	2	24	15
15	LLA	1	2	1	2						2	3	3	3	2		2	2	12	8
16	DGB																	1	7	7
	Count	10	12	9	13	3	4	5	3	4	8	8	11	9	9	6	7	16	16	16
N=16	**%**	63	75	56	81	19	25	31	19	25	50	50	69	56	56	38	44			
N= 666	**% Sha-piro**	46	57	25	33	9	33	19		18			32	17	17					

Table 6.2: **The vocal tics** in the all-adult study (N=16) compared to Shapiro's study (N=658). The tics tabulated here are: (19) grunting, (20) throat clearing, (21) coughing, (22) sniffing, (23) whistling, (24) bird noises – hooting, (25) animal noises – barking, (26) gulping, (27) humming, (28) saying single syllable, (29) saying single words, (30) obscene words – coprolalia, (31) repeating own words/sentences, (32) repeating other's speech, (33) insults – lack of inhibition, and (34) other vocal tics.
For more details see the text.

different. For example, the “gulp” in my list is not in Shapiro, while Shapiro’s list has some items (e.g., hissing) that my list lacks. But for most vocal tics, the percentage of various vocal tics among ATS in the pilot study is greater than of the lifetime vocal tics population. The 69% of coprolalia in my study exceeds Shapiro’s greatly. But this is by design. Since exploring coprolalia has been one of the goals of my study, the selection criteria ensure that more than 50% of the participants have coprolalia.

A close consideration of motor and vocal tics reveals that there is more to the vocal-motor distinction than meets the eye. And since the DSM-IV requires the presence of both for TS diagnosis, there are applicable consequences to their understanding.

Do modalities mix?

A statistical study of the temporal analysis of the video recordings of the interviews conveys that motor and vocal tics seldom co-occur. It is not completely surprising: Two vocal tics cannot co-occur due to biological constraints; two motor tics are also unlikely to co-occur. But mixing vocal and motor modalities is common in normal communication of speech and gesture (McNeill, 1996). The non-mixing of vocal and motor tics may support Jankovic & Kwak and Swerdlow, who consider phonic tics as motor. Indeed, if the phonic is just another form of motor, we should not expect the phonic and the motor to co-occur. If, however, we consider coprolalia as vocal and close to speech, the rarity of motor-vocal co-occurrence is interesting and requires new interpretation.

In chi-square tests[65], applied within-subject to 14 out of the 16 participants[66], I compared the co-occurrence of vocal and motor tics to (i) random prediction of co-occurrence of motor and vocal behaviors, and to (ii) mixing gesture (motor) and speech (vocal). Each participant defines a population (see table 6.3) of seconds with either only one modality of behavior (motor *or* vocal) or two modalities of behavior (motor *and* vocal). Two chi-square tests are applied to each such population[67].

		Rand co	Rand pure	Tic/rand	Tic co	Tic pure	Tic/lang	Lang co	Lang pure
1	Steven	20	512	17.54	1	531	393	157	105
2	Lionel	59	190	57.77	3	246	62.04	78	240
3	Ted	28	87	25.91	2	113	97.94	116	85
4	Kyle	113	358	101.8	9	462	38.62	16	85
5	Dylan	37	115	20.08	9	143	21.19	27	75
6	Henry	77	396	77.69	2	471	472.5	123	27
7	Stuart	31	364	32.27	0	395	192.8	55	71
8	Elliot	54	757	21.56	16	795	322.6	137	187
9	Chuck	84	269	95.34	0	353	85.67	57	202
10	Neil	10	40	11.11	0	50	63.63	164	103
12	Nick	1	368	1.001	0	369	114.6	80	213
13	Claire	30	13	0.0049	30	13	0.08782	139	54
15	Louis	7	20	8.043	0	27	50.12	193	87
16	Daniel	0	26	?	0	26	3	5	32

Table 6.3: Uncorrected two chi-square comparing (i) tic vs. random behavior, and (ii) tic vs. linguistic behavior. The results are in the yellow columns. For $p<.05$ the chi square critical value for one degree of freedom is 3.84. For $p<.01$ the critical value is 6.64. Only the values in red are below these critical value. For all the others the tic modality mixture is significantly different from either random or linguistic distribution. **Abbreviations**: *co* co-occurence, *lang* language, *pure* only one modality, *rand* random,.

As presented in table 6.3, for 11 participants there is a significant difference between their vocal-motor tic-modality co-occurrence, and their speech-gesture co-occurrence; and a significant difference between their tic-modality co-occurrence, and their predicted

[65] I used (Dean & Sullivan, 2009) for the chi square tests.
[66] The almost constant eye blinking of Danielle (#11) has made the high-resolution temporal coding of her tics too challenging. Donna (#14) was difficult to code due to her frequent getting off her chair and walking in the room.
[67] The technical details of the study are described in chapter 4, "Research goals and methodologies."

random mixed-modality behavior. The outliers are Nick, Danielle, and Claire. Claire has especially severe phenotype with forbidden gesture, falling tics, and self mutilation; and is about to be scheduled for a DBS. This might explain her difference from the rest. Nick's modality-mixture when he tics does not differ significantly from a random prediction. Daniel's tic-modality co-occurrence does not differ significantly from his linguistic behavior. The "0"'s in his data cannot be calculated in the chi square 2x2 table, since the chi square would have a "0" in its denominator.

Of course, this is a pilot study with a small sample and therefore with too low power to take these results as more than suggestive. Moreover, interpreting these results is difficult. No understanding of tic temporal behavior has been achieved yet. Data about the frequency of the electrodes of DBS and the effect of their frequency tuning on behavior has not been successfully interpreted. It seems that all a person can have is one tic at a time. As though the neurocognitive resources involved in ticcing can be expressed in only one tic. However, the meaning is still elusive and requires further studies.

To the best of my knowledge only one other study attempted a temporal analysis of video recordings of TS behavior. Indeed, the coding is labor-intensive and the results are still hard to interpret. In a 1998 study, Bradley Peterson and James Leckman gathered data from videotaping of 22 medication-free TS participants. The authors assessed the intervals between temporally adjacent tics. Applying statistical and graphical methods to analyze and present their data, they found (in mathematics-free language) that (i) the longer a behavior is videotaped, the more frequent the tics get; this is true in their study

for the motor tics alone, the phonic[68] tics alone, and their combination; (ii) the temporal pattern of tics at any time depends on the temporal patterns that precedes it; (iii) tics tend to come in bouts; and (iv) the temporal pattern of the tics and their bouts is fractal; that is, there is a pattern that repeats itself regardless of scale[69]. In other words, if we look (not too closely) at a graph of tics over a time period with 50 tics in it, or 250, or 500 tics they look very similar in shape and complexity (Peterson & Leckman, 1998). The authors conclude from their findings that the short-term bout pattern and the long-term waxing-and-waning pattern might reveal the same underlying tic dynamics. The pattern of bouts has been long recognized (For example see Leckman et al., 1999; Meige & Feindel, 1907/1990), but to the best of my knowledge, the fractal nature of the tics, to which the authors refer as "suggestive though not conclusive" (1337), has not been replicated or followed up.

THE COMPLEXITY OF TICS

The most common classifications of tics consider both modality and complexity. For motor tics, many distinguish between simple and complex tics according to the muscle groups that participate in the tic (For example see Jankovic & Kwak, 2005; Leckman et al., 1999); for vocal tics, the simple-complex distinction is often assigned to sounds vs. words (Erenberg, 2005). Erenberg distinguishes simple motor, complex motor,

[68] The authors do not dwell on the meaning of the word "phonic," and it is not obvious from their paper how they mean it. Since the diagnosis of TS requires vocal tics, it is reasonable to assume that their "phonic" is "vocal" including meaningless sounds as well as verbal expressions.

[69] A fractal dimension measures the roughness of an irregular shape, like a coastline. A natural process with a constant fractal dimension demonstrates self-similarity. Aside from random details it looks the same regardlessofo magnification. Common examples besides that of a coastline are of a snowflake and a fern leaf.

simple vocal (phonic), complex vocal (phonic), dystonic, and sensory (156). It is convenient to maintain the modality-complexity distinction for simple tics.

But for complex tics I propose classification across modalities. In addition I propose a refinement of the classification of complex tics into three subcategories with ascending severity: (i) elaborate, (ii) symbolic, and (iii) violent tics. Two Venn diagrams comparing the existing and proposed categorizations are in figure 6.1[70].

Simple tics

Simple tics are *sudden* and *brief*. They are limited to one or only few muscle groups. This muscular pattern is often considered as a reliable way to recognize them. Simple *motor tics* include: eye blinking, nose twitching, lip pouting, and rapid jerking of any part of the body. The most common are eye tics and neck tics (Shapiro et al., 1988: Chapter 5). Simple *vocal (*or *phonic) tics* include: throat clearing, coughing, sniffing, screeching, barking, grunting, gurgling, clacking, hissing, sucking, and other sounds. (Leckman et al., 1999).

Simple tics are sometimes so simple that the ticcer may be totally unaware of them. A vivid description is given by Adam Ward Seligman, who remembers himself at the age of seven, after the sudden death of his father, in a summer camp: “I came into the bunkhouse and found another camper sprawled on my bunk. I asked him to move. He began making faces at me – blinking his eyes, shrugging his shoulders and smacking his lips. ‘Why are you doing that?’ I demanded. ‘That’s what you do – ALL THE TIME!’ he

[70] In citing the literature, the novelty of my proposed categories presents a challenge. To which of my categories the complex tics in the literature fit? Context has been my guide. For example, when an author writes about complex tics but focuses only on, say, symbolic tics, the context guide me to interpret the “complex” as “symbolic.”

said… I looked in a mirror. He was right." (Seligman, 1992:34). A similar story was told to me by Neil in his interview, "I do remember one adult blinking back at me. I thought that this was not a right thing to do, (smiling). I thought he was kidding with me… I blinked back at him and he was sort of mimicking me and I did not know I was doing that." (Finkelstein et al., 2007-08-22). Danielle said, "It's almost like with the facial ones I don't even realize… until the muscles start being so sore or somebody will start saying 'you feeling OK?' and then I know they're saying 'Your tics are getting bad'" (Finkelstein et al., 2007-08-17). In an objective electroencephalography (EEG)[71] study of 6 TS patients, similar observations were made. The simple tics, which were not preceded by a premonitory urge, were often un-noticed by the ticcer (Obeso, Rothwell, & Marsden, 1981).

Elaborate tics

Elaborate tics are described by Leckman, King, and Cohen as longer than simple tics. They are stereotyped and look more purposive than simple tics (Leckman et al., 1999). Indeed, in coding the 16 video-recorded interviews, I sometimes found it difficult to decide whether a certain motion was a tic or a gesture.

Unlike simple tics, elaborate tics are generated by complex temporal orchestration of simple tics or by recruitment of a few muscular groups. Below I describe ritual tics and falling tics. There are many more kinds of elaborate tics, but I focus on these two because they are severe and were performed or described in the interviews.

[71] In EEG, the spontaneous electrical activity produced by the brain is recorded from multiple electrodes placed on the scalp.

Ritual tics

Some elaborate tics are stereotyped series of simple tics. Many of Steven‘s tics are performed in series. Often he takes three seconds for knee clapping or bending. Then he follows with a sniff, typically in the same intensity as the preceding tics, and this brings the elaborate tic to its completion. (Finkelstein et al., 2007-09-28). The ritual suggests that part of the elaborate tic depends on obsessive-compulsion disorder (OCD) mechanism. Ted commented on the difficulty in distinguishing ritualistic tics from OCD: “You know a lot of times… when I walk to work…Every third car -- I have to touch it. If you call it OCD I don’t know but washing hands it’s not” (Finkelstein et al., 2007-08-06).

Ted’s distinction between pure OCD, which often includes hand washing, and his TS+OCD is supported by the literature. In their literature review of the OCD spectrum, Shytle and Wilkinson cite studies showing that touching and counting compulsion, like Ted’s rituals when he walks in the street, is more likely to be found among people with TS+OCD, while contamination obsessions and compulsions are more common among people with only OCD (Shytle & Wilkinson, 2007).

A home-video of Daniel shows him before his deep brain stimulation (DBS) in his home kitchen. A ritual of two consecutive tics punctuates his moving around the kitchen: He either bends twice, or whirls twice, or whirls once and then bends once. If the second tic is bending, it’s preceded with a small and soft jump (Juncos, DeLong, & Finkelstein, June 2008). Another indication that the sequence of tics assembles to a ritual is that typically all the tics in any instance of the ritual are performed with the same intensity as the others at this instance. This has been observed not only in Daniel’s home-video but also in many of the DVD’s.

Falling tics and epilepsy-like tics

OCD seems to contribute not only to ritual tics but to other tics as well. In her interview Claire describes and demonstrates: "Sometimes I fell down on the floor (demos) and then I just scream (screaming)… Sometimes 2 minutes long… keep on [the] ritual… And sometimes I can't breathe and then afterwards when I'm done, I feel that my heart is gone little fast and I start sweating…and I fall like this and (getting up putting her hands on the desk demos falling on knees), and 3-4 times, six times until it feels right" (Finkelstein et al., 2007-12-21). This falling tic appears to combine a motor part, responsible for the falling, and an OCD mechanism, driving the repetition and the "feels right" experience.

Other falling tics resemble epilepsy. They are severe and mysterious. I discuss their observable signs here and their possible triggers later. Danielle calls them "body tics" and this is how she describes them: "I was conscious. I mean I couldn't respond but I could hear what's going on and since they [the doctors] never been able to get anything about it [referring to her few EEGs] they're pretty sure they're almost like just a major body tic and for a while I was having, you know, eight to ten a day…My eyes for a while would roll up kind of like to the back of my head and I couldn't see" (Finkelstein et al., 2007-08-17). Danielle's body tics are diagnosed as psychogenic non-epileptic seizures (PNES). She underwent a few synchronous video-EEG tests, which are recognized by the psychiatric community as providing "definite diagnoses of PNES." (Reuber & Elger, 2003:205). Based on these, she was diagnosed as non-epileptic.

Lionel was never diagnosed with epilepsy but suffered from "seizures" in his youth. He described them as involuntary, even though he learned to predict their coming.

His premonitory sensations served as an alarm pointing that he should lie on the floor to avoid falling. The seizures themselves, though, unlike many of his other tics, could not be controlled at all (Finkelstein et al., 2007-11-07). Henry, who also has occasional body tics, describes them as under control, "If I'm on the floor you think I'm having an epileptic fit. I'm still in control." But it is not clear that this is indeed the case, and it was difficult to probe him further about the nature of his epileptic-like fits (Finkelstein et al., 2007-07-23).

Whether the body tics described by Danielle, Lionel, and Henry are in the same spectrum with other motor tics is a difficult question. Comorbidity with epilepsy is possible too. Guerrini and colleagues teach us that "The boundary between epilepsy and movement disorders may sometimes be difficult to define… Not only can epilepsy masquerade as movement disorders , but movement disorders may be observed which are not easily differentiated from epileptic seizures; syndromes or diseases may associate both epilepsy and movement disorders occurring in the same patient" (Guerrini, Aicardi, Andermann, & Hallet, 2002:xix). This further adds to the spectrum complexity of TS.

Symbolic tics[72]

Symbolic tics have *meaning*. Coprolalia and copropraxia are the most prominent symbolic tics. According to Gilles de la Tourette they are central to the disorder, and he considered coprolalia mandatory for the diagnosis of *maladie des tics*. The motor analogue of coprolalia is copropraxia. Singer (1997) and Roger Freeman (2007) refer to them collectively as coprophenomena regardless to their modality. And this further highlights their foul nature. My observations of the copro-tics have convinced me that

[72] I use the word "symbolic" in a different manner than Freud and his followers have. I elaborate on it in the chapter "Symbolic tics."

while their foul nature is common, it is not necessary. What characterizes them is the *violation of social conventions*. I will further discuss these tics it in the chapter "Symbolic tics."

Violent tics

Tourette syndrome patients can be violent towards themselves and towards objects, but typically not towards other people.

Violence against objects

Ted's tics include punching holes in the walls of the house. In addition he regularly pushes the windshield out of his car (Finkelstein et al., 2007-08-06). Before his deep brain stimulation, Dylan's tics were very severe, especially in small, confined spaces. In an elevator, he would bang the walls of the cage. In the bathroom when looking into the mirror, Dylan had to be grabbed or he would go into the sheet rock and damage it (Finkelstein et al., 2007-08-01a, 2007-08-01b). Nick used to break things, seemingly not motivated by anger: "Sometimes I reach out and touch stuff (playing with his pant's cloth between middle finger and the thumb of his right hand). It's crazy… I just sometimes squeeze it and sometimes I hold it too tight and I'll break it" (Finkelstein et al., 2007-12-07).

Lowell Handler, the photographer of the documentary *Twitch and Shout*, used to get permission from his mother to completely demolish his room. She understood his violence against his room as "venting and releasing an overflow of energy." Handler further explains this energy as resulting from the "inability to express myself," and "Later, in high school, I resorted to smashing walls and breaking objects. My inability to

understand my condition exploded in these unsubtle and inarticulate ways" (Handler, 2004: xxiv).

Self mutilation

The line between violence against objects and against self -- self injurious behavior (SIB) -- is not always clear. Nick does not stop at breaking things. Sometimes he bites glass and cuts himself in the lip. "Sometimes even on a good day, just getting up. I used to put my fingers in fans and stuff like that…. Get the urge to do stuff. I know I shouldn't do it but I do it sometimes." Attraction to glass is not unique to Nick. For years, Ted has had the urge to take his mother's favorite glass vase and drop it on the floor (Finkelstein et al., 2007-08-06). Claire got mad one time at her husband and cut her artery at the right wrist on a glass door (Finkelstein et al., 2007-12-21) . Danielle also described cutting herself, not with glass, but with razor blade. And "not… real deep, just enough to get blood and almost that was the satisfaction" (Finkelstein et al., 2007-08-17).

Dylan often "bashed his head repeatedly into a corner in the wall." But his SIB did not stop there. After his first deep brain stimulation (DBS), he scratched the surgery area so badly that he infected it, to the point that another DBS had to be performed. Before the surgery he used to burn himself with cigarettes. He also used to rub his eyes and gouge in them until they got "blood red" (Finkelstein et al., 2007-08-01b).

The eyes are frequently a target for self mutilation. At the age of 11, Henry damaged his right eye and has now prosthesis. He further damaged himself, hitting his belly with his right elbow until he ruptured his appendix. Lesser, yet self injurious behaviors, continue for him. Clapping his knees against each other, Henry says, "I had a knee tic. I had a bruise… you would not believe. I am thinking about blood clot…

Anyway, I can't stop… I'm trying to hurt myself. It hurts… it hurts." (Finkelstein et al., 2007-07-23).

The hair is a common target too. Pulling one's own hair is called trichotillomania. Louis remembered himself as a child twisting his hair and pulling them out. It has left some bear spots in his head (Finkelstein et al., 2008-05-30). Before his DBS Daniel used to pull his hair out (Finkelstein et al., 2008-06-04).

Ted picks his skin. And when he was treated with Haldol that sent him to the hospital he hurt himself severely: "Can you imagine your lips and face being open and still sticking fingers. It's like cutting your hand and putting salt in it, salt in it, salt in it" (Finkelstein et al., 2007-08-06).

Danielle attempted suicide a few times, but has given up her suicidal attempts. Nonetheless, she continues to have suicidal thoughts even though she does not act on them, and she continues to inflict injuries on herself (Finkelstein et al., 2007-08-17). Her behavior and thoughts challenge the opinion of the psychiatrist Armando Favazza and the neurologist Robertson (Favazza, 1996; Mary M. Robertson, 2000), who define SIB as being without suicidal drives.

Almost all the participants with SIB attribute their self-mutilation to their OCD. This linkage is in agreement with OCD studies showing that self-mutilation happens when both OCD and TS are present (cited in Shytle & Wilkinson, 2007:49), and in agreement with TS studies summarized by Mary Robertson and Michael Orth that SIB "often have an obsessional quality to them" (2006:55). When discussing pathological self-mutilation as opposed to culturally sanctioned practices, Favazza too considers the behavior to be impulsive (Favazza, 1996).

Interestingly, column 14 of table 6.1 reveals that only 4 participants in my study self-reported SIB, or as it is also called, self mutilation. Lionel and Henry rated their self mutilation as very forceful and very frequent, and Nick and Donna reported of mild and infrequent self mutilation. None of the other participants reported self-mutilation in their TSSR. But Claire, Dylan, Louis, and Daniel, described self-mutilation behavior in their interviews[73].

The complexity of tics -- Summary

I have proposed four levels of tic complexity: simple, elaborate, symbolic, and violent. The complexity crosses modalities and applies to both motor and vocal tics. A s*imple* tic typically depends on only one muscle group. An *elaborate* tic, in contrast to a simple tic, relies on more than one muscle group (e.g., whirling) or is a rapid sequence of simple tics (e.g. a series of: eye blinking-shoulder shrugging-sniffing). A *symbolic* tic is linguistic, either through speech or gesture. Finally, there are *violent* tics. They include violence against objects and self-injurious behavior (SIB). I am not aware of tics that include violence against other people.

As in many classifications, the borders between categories are often fuzzy. Spitting, for instance, is usually classified as simple. However, there is room to consider it symbolic since cross-culturally it is an insult (Flynn, 1977). Moreover, since spitting might physically touch its target, in some situation it might be considered as violent. Context is important. The situation provides the context and its interpretation (Yeh & Barsalou, 2006). Spitting while working alone in the garden might very well be a simple

[73] The discrepancy between the information in the TSSR and the interviews is an example of the weakness of self reports, mentioned previously in the chapter "Research goals and methodologies."

tic, spitting at a friend during a conversation might be symbolic, and spitting at a person who had stepped on one's toe in a public place might be violent. And even though these tics are mostly involuntary, their nature is defined by the situation.

The two Venn diagrams in Figure 6.1 below represent the relations between the current complexity classification and the proposed new one. The proposed classification is in *italics*. *Simple* and *elaborate* tics are distinguished mainly by the muscles that produce them and by their durations. Tics of coprophenomena are *symbolic*, but might have at times a subtle form of violence; thus they cross the border between the symbolic and the violent. Tics of breaking or otherwise damaging objects and SIB are *violent tics*.

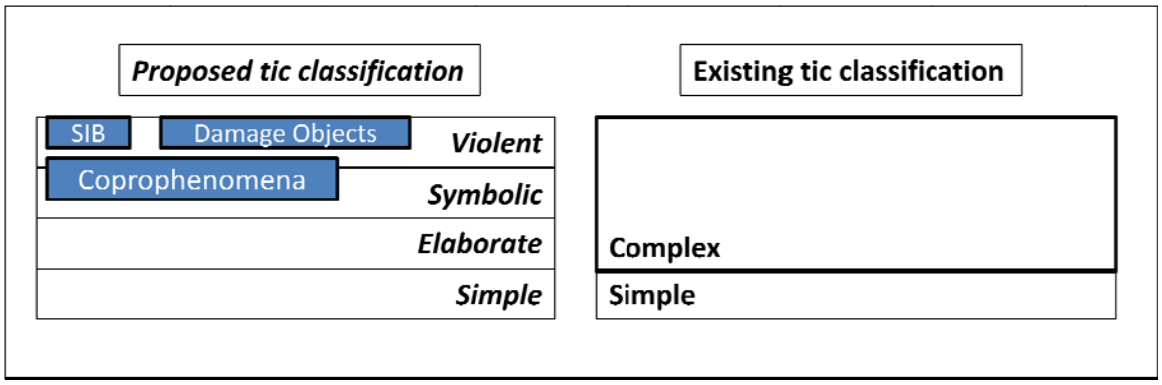

Figure 6.1 Two classifications of tics by complexity.
The right Venn diagram represents the current common classification.
The left Venn diagram (with italicized fonts) represents my classification.
See more in the text.

CONCLUSION

In this chapter I conducted two classifications of the observable behaviors of TS: by modality; and by complexity. When classifying by modality, the clear distinction between motor and vocal gets complicated and phonic tics emerge in the gray zone

between the vocal and the motor. In addition, close temporal analysis of the video-recording shows different relations between motor and vocal tics than between speech and gesture. The rarity of co-occurrence of motor and vocal tics is difficult to interpret but it may suggest the possibility of shared underlying mechanism between these two modalities. When this mechanism is recruited by one tic, it is busy and cannot be recruited by the other.

The classification by complexity is across modalities and is more refined than the current classification of tics to simple and complex. I have proposed simple, elaborate, symbolic, and violent tics. The various kinds of tics and their severity may be related to the spectral nature of the disorder, and to the question of comorbidity, which is relevant to this study since all the participants have turned to have OCD, and some have SIB. The OCD seems to contribute to the severity and maybe to SIB too – a point that I will discuss further in the next chapter.

I did not discuss symbolic tics in this chapter. Instead they are the topic of the chapter "Symbolic tics." I will discuss them after the next chapter, in which tics will be classified by their triggers.

TIC CLASSIFICATION: TRIGGERS OF BEHAVIORS

CHAPTER CONTENTS

IN THIS CHAPTER I continue the task of classification. Here I classify the tics by what triggers them. Unlike for the previous classifications by modality and complexity, the data for classification by triggers have to be provided by the participants. The processes of triggering the tics are not observable; nor are the conditions in which people tic less.

THE TRIGGERS OF TICS

Visual and somatosensory[74] stimuli trigger tics. My interviews suggest that acoustic stimuli do not. The olfactory were not discussed. Social environments trigger tics and affect their frequency and intensity. Internal triggers arise from purely internal processes of which the person may or may not be aware. But they themselves may be triggered by the visual, the somatosensory, or the social.

What triggers tics -- the visual world

Both Meige and Feindel (1907/1990) and Leckman, King and Cohen (1999) agree that eye tics are demonstrated by most people with TS, and that often eye blinking is the first tic to appear at the onset of the disorder. In addition, the eyes are a common (even though not the only) target for self injurious behavior (SIB): E.g., pressing vigorously on the eyeball (cited in Eapen, Yakeley, & Robertson, 2005), burning the eye lids with cigarettes (Visse-Vandelwalle et al., 2003), and damaging the cornea (Margo, 2002; Schweinitz, 1906). As the eyes are the organ of visual stimuli, it is likely that visual stimuli are disturbing to TS patients and trigger their tics.

[74] I thank Krish Sathian for his suggestion to use "somatosensory," which comprises all of touch (non-noxious), pain, and proprioception (movement sense).

There is more evidence suggesting this possibility. Saccade[75] of most people with TS is different than that of the general population (Munoz & Everling, 2004; Munoz, LeVasseur, & Flanagan, 2002). Reading is often challenging for people with TS. And finally, many do not tic when they sleep or at least they tic less than when awake, even though the sleep of many is restless and disturbed.

Eye tics

Meige and Feindel (1907/1990:146) subdivide eye tics to eyelid tics and eyeball tics. This distinction is found in the literature of epilepsy (For example see Kent, Blake, & Whitehouse, 1998) but with the exception of Meige and Feindel I have not found it in the literature.

Eyelid tics

The eye blinking tic, the palpitation of the upper lid, differs from regular eye blinking only in frequency and abruptness (Meige & Feindel, 1907/1990:146). It can be unilateral or bilateral. Its possible link to visual stimuli and in particular to light, was proposed by the participant Elliot: "Environment is a big part of it…If I could just close my eyes it probably would be much better…(Closes his eyes) there is no lights, there is no this (pointing at the ceiling lights)…that light above me. It's hitting that part of my head (circling with his right index finger around the center of his forehead) and move it around like this (draws a halo-like circle above his head with his right index finger)… Environment is such a big part… It's almost like I have to push them [the pictures in the

[75] Saccadic eye movements are rapid eye movements, which act to redirect the eyes from one object of interest to another. Broadly speaking, the purpose of these movements is to bring images of particular areas of the visual world to fall onto the fovea to afford acute vision.

room] down… certain lights and windows … that's why I say when I close my eyes it's better … I don't see anything that I have to interact with" (S. R. Finkelstein et al., 2007-08-13). Elliot ranked his own blinking at 2 on a 0-3 scale with 3 being the most severe.

A similar ranking of 2 was self reported by Lionel. A painter, Lionel's interaction with the visual world is rich and subtle. When I asked him whether there was anything in the visual world that triggered his tics, he said that his coprolalia seems to be triggered by the visual world. The only visual specificity that he could provide related to colors, "All I know is the ritualistic kind coprolalia; that things that I would see would trigger that. And sometimes it would be certain colors. I might see a hundred different blues and it wouldn't bother me and then… a certain blue color dress or there is something, you know, they would trigger it [the coprolalia]" (S. R. Finkelstein et al., 2007-11-07).

Danielle ranked her blinking at 3, and blinked bilaterally almost without stopping throughout her interview. "When I was in fourth grade and I started like with constant eye blinking… and blew my hair out of my eyes… they thought then because of my constant eye blinking that I needed, ah, glasses… I started off with a constant eye blinking" (S. R. Finkelstein, PhD, Juncos, Poh, & Kushner, August 17 2007: 00).

Neil ranked his blinking tics at 2. His blinking is mostly unilateral, on the right. And while he remembers sniffing rather than eye blinking as his first tic "because I remember that my mother thought I had allergies" (S. R. Finkelstein et al., 2007-08-22a), it is possible that his eye blinking started earlier and was not noticed by his mother. Already Gilles de la Tourette observed that "[t]he attention of parents is soon drawn to the fact [that their child has a series of tics], but they seldom give much heed at first" (cited in Meige & Feindel, 1907/1990:224).

Ten of the sixteen participants in my study (62.5%) reported eye blinking in their Tic Symptom Self Report (TSSR). This is below the 69% reported by David Comings (Comings, 2001:13) and the 80% reported by Shapiro in a study of 666 patients (Shapiro, Shapiro, Bruun, & Sweet, 1988:351), but showing a similar tendency. Even higher percents were reported by Frankel and Cummings who found 28 out of 29 TS patients having eyelid tics or reporting having them in the past (1984).

Eyeball tics

The eyeball tics "usually consist of brief, rapid, conjugating upward and/or side-to-side ocular movements, sometimes presenting as eye rolling." (Fernandez-Alvarez, 2001:334). In their study of 29 TS patients, Frankel and Cummings observed 2 patients with eye-rolling, and 2 other reported having eye-rolling in their past (1984).

Danielle's eyeball tics come with her "body tics." She experiences them as different from other tics. "For a while I was having, you know, 8 to 10 a day…My eyes for a while would roll up kind of like to the back of my head and I couldn't see" (S. R. Finkelstein et al., 2007-08-17b).

Lionel reported eye-rolling that accompanied his falling too. But this was earlier, at his childhood (S. R. Finkelstein et al., 2007-11-07). Henry had a few eye rolling tics during his interview. And he too linked his eyeball tics to his falling (S. R. Finkelstein et al., 2007-07-23).

Eye injury

For some people with TS the eyes are an irresistible target for touching. "I rub my eyes, which I'd like to do" said Neil (S. R. Finkelstein et al., 2007-08-22a). His eye

rubbing seems quite benign. But this is not always the case. Mary Robertson comments on inflicting severe eye injuries as "curious" (2000:431). Favazza, citing from a 1989 paper by her, describes a TS patient who by the age of 22 had self injurious behavior (SIB) with "severe eye poking" (Favazza, 1996:240). Michael Frankel and Jeffrey L. Cummings report on a TS patient who slapped his eyes with his hands until he ended up with bilateral pterygia[76] and a traumatic cataract (1984). They also report of self destructive behavior -- staring at the sun or other bright lights. Houeto and colleagues, who performed deep brain stimulation (DBS) procedures on five TS patients, report that among their patients was a 36-year-old woman who stuck her finger in her eye, causing herself lesions of the cornea (Houeto et al., 2005).

As was mentioned above, Dylan's father said that before his DBS Dylan "was… goug[ing] in his eyes… rub[bing] his eyes until it gets blood red" (2007-08-01b). Henry, who had told us that at the age of 11 he damaged his right eye and has now a prosthesis, kept bringing his right fist close to his only, left, eye: "My good eye… When I was a little boy I would hit myself here (hitting his left cheek)… Even now… I still have the tendency, not severe (hitting his left cheek)… [to] poke pencils and pens at my good eye." (S. R. Finkelstein et al., 2007-07-23). After being (mis)treated with Haldol, Ted was hospitalized: "My eyes -- I'd beat them so bad I had to have a cataract surgery on both eyes and I had the beginning of glaucoma" (S. R. Finkelstein et al., 2007-08-06).

Saccade

Another eye-related abnormality found in TS patients is observed objectively with appropriate equipment. In the antisaccade task, the subject is instructed to inhibit a reflex

[76] Pterygia is a fleshy mass of thickened conjunctiva that grows over part of the cornea usually from the inner side of the eyeball and causes a disturbance of vision.

eye movement towards a peripheral target light and instead to generate a movement in the equal and opposite direction. In a study of 6 adult male TS patients, Dursun and colleagues have found the patient antisaccade latencies to be significantly longer than that of the controls[77] (Dursun, Burke, & Reveley, 2000). This result is in agreement with a previous study of 10 adult TS patients who showed a significant increase of the latency of antisaccades and highly impaired performance of sequences of memory-guided saccades.(Straube, Mennicken, Riedel, Eggert, & Mülller, 1997). Similar observations were reported in a 2001 study of 10 adult TS patients, who compared to control subjects, showed longer saccadic reaction time and smaller saccadic amplitudes. They also found longer antisaccadic reaction time (LeVasseur, Flanagan, Riopelle, & Munoz, 2001).

If this abnormality in the saccade mechanism exists, it might also explain the common head and neck tics. While moving the head or the neck is much cruder than saccade, maybe such movements attempt to compensate the less-than-perfect alignment of the fauve with the object of attention. Fourteen of the participants in my study, 88%, self reported head tics (see Table 6.1). The TSSR did not ask about neck tics. But the urge for neck tics is marked among the highest among people with TS (James F Leckman et al., 1999).

Reading

Walkup and colleagues discuss the comorbidity of TS and learning disability (LD) (Walkup et al., 1999). Among the markers of LD, reading difficulties are prominent. And indeed, for some of the TS patients that I interviewed reading is challenging. Chuck, despite being a teacher and a teachers' teacher, finds reading

[77] The order of magnitude of antisaccade latency is around 500 msec.

difficult: “Can I read? Yes. Do I read? Yes! Do I like to read? No.” He blames it on the “tics [that] get in the way.” He elaborates: “When you start reading you start ticcing and half about the page you forget what you read. You start over again, you forget what you read. You do it again. After a few times of doing that you forget what book you are reading. You look at the front and you say, a yeah, I’m reading that book” (S. R. Finkelstein et al., 2007-12-21).

There are a few possible explanations, not mutually exclusive, to the reading difficulty: (i) It may be, as suggested by Chuck, that the disruption of the tics does not permit a smooth, normal, reading. (ii) Maybe the disruption is more subtle and caused by abnormal saccade mechanism. (iii) And finally, there may indeed be comorbidity with LD.

Chuck has more to say about his reading: “When I’m reading my issues are more of the tics. It’s physical. If you twitch your head enough you lose where you are so you got to keep your finger there and you twitch and trying to figure it out. You just do the best you can.” But in addition, his strategies around this difficulty address the challenge posed not by tics but more by distracting visual stimulation: “Sometimes you use pointer or maybe you use a paper and you slide it down so you don’t see the rest of the page.” (S. R. Finkelstein et al., 2007-08-24).

Danielle spoke about her reading difficulties, in spite of which she earned an undergraduate degree. Like Chuck, she attributed her reading difficulties to the physical action of reading. But then we discussed it further. When she said to me “I don’t comprehend when I read very well,” I asked her, “Did you try sometimes, when you don’t comprehend, to read out loud?” Her answer was “Yes. Even in college… my

counselor would have made her assistant read all the textbooks I had to read on a tape so I would read and listen at the same time and that would help." (S. R. Finkelstein et al., 2007-08-17b). It is not clear whether only the challenge of the visual environment was in her way. There might have been some level of LD.

Neil, with the highest education among the 16 people in the study, reads "mostly non-fiction." And when I asked him, "Is reading any challenge?" he said, "I fall asleep a lot reading," and later added, "I am a slow reader, fairly slow reader." When reading fails him he "listen[s] to books." (S. R. Finkelstein et al., 2007-08-22a).

The possibility that for some TS patients reading difficulties are a by-product of the motor disruptions produced by tics and abnormal saccade, adds another consideration to the question of the comorbidity between TS and attention deficit hyperactivity disorder (ADHD). LD is often linked to ADHD, which in turn is often comorbid with TS (Walkup et al., 1999). In a 2007 global study of 6805 TS probands, Freeman has found 55.6% ADHD comorbidity and 22.0% LD comorbidity (Freeman, 2007). The possibility that for at least some TS patients, ADHD and LD symptomology is a by-product of TS, not a manifestation of real ADHD and LD, deserves further exploration.

The difficulty in determining whether there is a cognitive LD, motor disruptions of the reading, disruptions from the underlying neurological visual mechanism, or any combination of them, is further complicated by the fact that reading and its comprehension is a learned skill that takes many years of practice to master. It is possible that due to the initial reading difficulty and the frustration, rather than joy, of reading, not enough time is dedicated to acquire reading skills; that reading hours are missing from the educational trajectory of TS children. It is difficult to determine whether reading

deficiency in TS patients is cognitive, mechanical, neurological, or educational; and which combination plays a role for each individual. But it is likely that abnormalities in the visual system contribute to the reading difficulty and in turn to attention and learning deficiencies.

Sleep

During sleep, with hardly any stimuli from the visual world, most of the participants were observed by their partners as either ceasing ticcing altogether or significantly reducing its frequency and intensity. Danielle said, "The only time I am really, they really relax, is when I am resting at home… Lots of time when I rest or finally fall sleep they [the tics] kind of go." And added, "As far as the tics that's the only time it seems really, you know, kinda at ease" (S. R. Finkelstein et al., 2007-08-17b). Dylan, who before his DBS was completely consumed by his tics, does not tic when he sleeps (S. R. Finkelstein et al., 2007-08-01a; S. R. Finkelstein et al., 2007-08-01b). Elliot does not tic when he sleeps (S. R. Finkelstein et al., 2007-08-13). Neil and his wife think that he does not tic when he sleeps (S. R. Finkelstein et al., 2007-08-22a; S. R. Finkelstein et al., 2007-08-22b). Claire tics less when she sleeps (S. R. Finkelstein et al., 2007-12-21). Donna who tics "from morning to night" does not tic when she sleeps (S. R. Finkelstein et al., 2008-02-08). And Daniel, even before his DBS, has not ticced during sleep (S. R. Finkelstein et al., 2008-06-04).

But the disappearance of tics or their reduction during sleep is not universal. Lionel was told by his former wife that during sleep he was restless and made noises. (S. R. Finkelstein et al., 2007-11-07). Kyle is often afraid to go to sleep "[It] happens every once in a while. I'll be going to sleep. I'll be like somewhere in and out of sleep, and my

whole body can't move. And then I think I can't breathe, and it takes all the energy I have to get out of it. And I feel like I'm getting jolted shocked." This is called by Meige and Feindel a "tic of immobility" (1907/1990:124). In their brief discussion they wonder whether this is a tic at all. And I have not found a discussion of this phenomenon elsewhere. To fall asleep, Kyle needs some calmness and "sometimes [the tics] won't stop enough for me to go to sleep" (S. R. Finkelstein et al., 2007-09-21).

That tics cease or reduce during sleep has been reported in the literature (Towbin, Peterson, Cohen, & Leckman, 1999). Erenberg is cited by Shapiro reporting that 86% of TS patients included in his study ceased to tic during their sleep. In Shapiro's study with 394 informants, 97.2% of the informants ceased to tic in their sleep. For 0.8% the tics slightly decreased, and for 2% they decreased markedly (Shapiro et al., 1988:287).

Some literature disagrees (for example Jankovic, 1997). There are also reports like those of Lionel and Kyle. The spectrum of sleep patterns of TS patients is wide. 20–50% of TS patients complained about sleep disturbances that include separation anxiety in the evening, sleep walking, sleep talking, unpleasant dreams, nightmares and difficulties in both falling and staying asleep. In contrast to such reports by patients and their relatives, a few polysomnographic[78] studies did not find similar patterns of frequency, intensity, and variability of sleep disturbances in TS (Reviewed by Mary M. Robertson, 2006).

Interpretation of ticless sleep is difficult since sleep differs from wakefulness in many ways. Still, there is the possibility that the lack of visual stimulus is an important aspect of cease or at least amelioration of tics during sleep.

[78] Using electrodes for EEG reading, polysomnography is a comprehensive recording of the biophysiological changes that occur during sleep.

The visual world -- summary

Whenever Henry answered a question he kept his eyes shut. When he opened them he lost his thread of thought and his coherence (S. R. Finkelstein et al., 2007-07-23), as though the visual world was distracting, interfering with his stream of thoughts. When I asked some of the participants to shut their eyes Elliot, Claire, Donna, and Stuart did not tic and reported a feeling of calmness.

Mr. O., the patient of Meige and Feindel, explained his tics: "A large number of my head and face movements owe their origin to the annoyance caused me by my seeing the tip of my nose or of my moustache from time to time." He also attributed his eye blinking to visual stimulation: "It is for an identical reason [of playing hide-and-seek with my nose] that each moment finds me blinking one eye or the other, or both" (Meige & Feindel, 1907/1990:3-4).

Eye blinking, abnormal saccade, ticless sleep, calmness with closed eyes, need to close the eyes for coherent communication, self injuries to the eye, and difficulty in reading collectively suggest, each in its own way, that visual stimuli trigger tics. Future studies of the visual system in TS patients could be productive and lead to deeper understanding of the disorder.

What triggers (and suppresses) tics – the somatosensory world

The interaction of TS patients with the somatosensory world is complex. Some are sensitive to tactile asymmetry and respond with touching or avoiding touch. At times of great stress, touch, even pain, may be sought after to calm down a tic.

The need for tactile symmetry

Lowell Handler, the photographer of *Twitch and Shouts* describes himself, "I like touching objects as well as people. If I touch something or someone with my left hand, I must touch with my right hand to obtain an equally satisfying feeling" (Handler, 2004:89). Some are over-sensitive to touches and experience a violation of symmetry with the lightest of touches. This may be an aspect of a more general need for symmetry, not only tactile. It can be visual and then it is typically manifested in the compulsion to arrange things. Without distinction between the visual and the tactile, 8 participants in my study (50%) reported such need for symmetry in the past, and 7 (44%) reported such need in the present. Five (31%) had it in the past and still have it in the present (See Table 7.1).

Elliot claps his knees against each other until they are blue, which he links to his obsessive need for symmetry, especially tactile symmetry. To get his point across, he demonstrates: First he blows strongly on the back of his left hand and then looks at the back of both hands and says, "I was blowing [on my left hand, and] felt the air from my nose [on] this hand, so I have to equalize on the other side and blow on it." Then he adds, "When I was in school and someone bumped into me in the playground [on my] left side… I fell out of synch. I felt like I *had* to equal it out" (S. R. Finkelstein et al., 2007-08-13). Ted's need for symmetry is satisfied by creating tactile numerical patterns. Often, on his way to work, "every third car – I have to touch it" (S. R. Finkelstein et al., 2007-08-06).

The irritation of being touched

Touch can be annoying, even when it does not violate the sense of symmetry. Lionel's coprolalia can be triggered by "the feel of a certain fabric" (S. R. Finkelstein et

al., 2007-11-07). Elliot has a neck tic since, "I can feel my neck right now because my shirt is stimulating my neck. I feel my shirt on my neck and I want to shrug my shoulders… it's this stimulation of touching" (S. R. Finkelstein et al., 2007-08-13). Danielle gets irritated when in front of the TV her husband touches her legs with his (S. R. Finkelstein et al., 2007-08-17b).

The need to touch

Lionel responds to stimulating touch with a coprolalic tic; Elliot with a motor tic; and Danielle avoids the touch. Meige and Feindel speak of "cutaneous irritation" that leads to "scratching tics" (Meige & Feindel, 1907/1990:186). The response to cutaneous irritation can itself be of a cutaneous nature. After his first DBS, Dylan could not resist touching the sensitive location of his surgery, infected the area and the electrodes, and needed a second surgery (S. R. Finkelstein et al., 2007-08-01a).

ID	Participant	Past	Current
1	SRA		
2	LRA	1	
3	TQA	1	1
4	KGA		
5	DQA		
6	HBA		1
7	SZA	1	1
8	EEA	1	1
9	CBA	1	1
10	NFA		
11	DLA	1	1
12	NYA		
13	CFA		1
14	DGA	1	
15	LLA	1	
16	DGB		
	COUNT	8	7

Table 7.1 Self Y-BOCS
Reporting the need for symmetry in the past and in the present. "1" stands for "yes."

The need to touch can be indeed in response to cutaneous irritation but not necessarily. Table 6.1 includes self reports about three touch-related behaviors. Twelve probands (75%) self-reported of touching their own bodies; seven (44%) reported of touching other people; and nine (56%) reported of touching objects. Touching one's own body and other people might be a form of copropraxia. But even then, it seems different than raising one's own middle finger. The need to touch has to be satisfied. Often, when the touching is severe and harmful, it cannot be distinguished from SIB.

Nick description of his SIB, which we heard earlier, is rich with details about touching: "Sometimes I reach out and touch stuff (playing with the cloth of his pants between the middle finger and the thumb of his right hand). It's crazy… I just sometimes squeeze it" (S. R. Finkelstein et al., 2007-12-07). Lionel touches his own body, objects, and taps his fingers (S. R. Finkelstein et al., 2007-11-07). Kyle touches his own body and taps his fingers (S. R. Finkelstein et al., 2007-09-21). Henry has a compulsive need to touch people and objects (S. R. Finkelstein et al., 2007-07-23). Stuart touches his own body and picks at things (S. R. Finkelstein et al., 2007-10-26). Chuck touches parts of his body and picks at things (S. R. Finkelstein et al., 2007-08-24). Claire touches herself. "or sometimes… other people or other things" (S. R. Finkelstein et al., 2007-12-21). Donna touches other people as part of her copropraxia. But she also touches objects, taps her fingers, and picks at things (S. R. Finkelstein et al., 2008-02-08). Daniel compulsively touches objects and prior to his DBS completed each bending tic with touching the floor (S. R. Finkelstein et al., 2008-06-04).

The comfort of pain

Self injurious behavior reveals possible relations between pain and tics. Despite giving up suicide, Danielle used the razor blade often, "not… real deep, just enough to get blood I… remember feeling better; almost like that took the pain or did something for me" (S. R. Finkelstein et al., 2007-08-17b). For Lionel, biting his lips and putting needles into his *skin* served as a way to reduce the tics. When he hurt himself the tics diminished. But with the passage of time he had to increase the pain thresholds to quiet down the tics, "as with drug addiction" (S. R. Finkelstein et al., 2007-11-07). Daniel's mother noticed that from infancy "he was different." Especially, he would not fall asleep unless he tightly pressed his finger-nails under hers (S. R. Finkelstein et al., 2008-06-04).

The somatosensory world – summary

The SIB and the attempts to quiet tics by self-inflicted pain raise a few possibilities: (i) The tics recruit some of the pain circuits. Therefore the pain, if high enough, can quiet down the tics; (ii) the pain threshold of some TS patients, especially those with SIB, is higher than that of the general population; and (iii) the pain threshold dynamically increases in a mechanism similar to that of addiction.

As for the other tactile tic-triggers: The need for symmetry is most likely linked to OCD. And the attempts to avoid touch might be driven too by the need for tactile symmetry. Further research of TS and the somatosensory world will further our understanding of the disorder.

What triggers tics – the social world

The most embarrassing and penalizing TS tics are the coprophenomena. They happen primarily in social situations and are triggered by social stimuli. I mention them here for completion, but discuss them in detail in the next chapter "Symbolic tics." They are not the only social tics. Echophenomena – mimicking other's speech or motor behavior happen too. For Charcot and Gilles de la Tourette, echolalia was a diagnostic criterion (Kushner, Luzzatti, & Finger, 1999). While this phenomenon is mentioned frequently in the literature, it has not been studied often. In their study of 666 probands with life-time tics, Shapiro and his colleagues have found 117 (17.6%) to have echolalia (repeating other's speech), 116 (17.4%) to have palilalia (repeating one's own speech), and 56 (8.4%) to have echokinesis (repeating other's movements). In my study the percentage has been much higher and echolalia and palilalia have been each self-reported by 9 participants (56%).

Lowell Handler describes the identical twins Carla and Claudia of Atlanta, both diagnosed with TS: "Both twins had a severe jumping tic, whereby they alternately jumped two feet into the air, crashing down a second later. This resulted in a constant rumbling because when one twin was in the air, the other was getting ready to jump. When the other was airborne, the first one crashed to the floor. And so it went, throughout the visit." (Handler, 2004:111).

The echophenomena might demonstrate an irresistible need to mimic. But it might be a part of the larger set-off phenomenon. Ted prefers to avoid Donna's company since her tics "set off" his own. And during his interview, his descriptions of some of his tics set them off (S. R. Finkelstein et al., 2007-08-06). When Claire demonstrated her falling

tics, she sets off a real falling tic. And, like Ted, just describing some of her tics was sufficient to set them off (S. R. Finkelstein et al., 2007-12-21).

What triggers tics – the darkness in the internal world

External triggers, acting through the sensory system, are not the only ones. The internal world of feelings and thoughts can trigger tics as well. Whether this world by itself is triggered by external stimuli or has its own life varies. Before ending our interview, I asked Lionel whether there was something that had not come up and that was important for my understanding of TS. This is when he spoke of his *o*bsessive and intrusive thoughts: "It's like being in this room with too many TV's with too many channels being wide open all the time, and I try to have a conversation with someone when all this crap is going on… I am constantly seeing things ... I can't push the stuff away." The obsessive and "evil" thoughts can be triggered by a movie after which he assumes the role of the villain, and harms those closest to him. He experiences "something *foreign*… inside… [that] was twisting; it was trying to torment me; it was trying to torture me and make me think things I didn't want to think." (S. R. Finkelstein et al., 2007-11-07). Henry shares: "As a boy I would have these [dark] thoughts." And they were about the people closest to him and that he "wish[es] the best to" (S. R. Finkelstein et al., 2007-07-23). Donna feels a prisoner of her dark thoughts and is helpless in their presence (S. R. Finkelstein et al., 2008-02-08).

Henry and Donna attribute their internal dark thoughts to their OCD. So does Adam Seligman: "In talking to Lowell [Handler] that evening I described the weird intrusive thoughts of violence, suicide, and sex that were interfering with my

concentration at work. He listened closely, then to my amazement told me that he had similar obsessions" (Seligman, 1992:53).

Rarely do people act on these thoughts. Lionel has found a coping strategy. After realizing that "the stronger I fight the stronger it gets… [I] pretend it's a bad movie and… [I have] to sit through it and just watch it." His approach decreases the control of the "demon" and carries him through these torturous hours (S. R. Finkelstein et al., 2007-11-07). When a friend of his calls up before dawn, Lionel keeps her company over the phone and stays with her until her "movie" is over.

The urge

Often a premonitory urge precedes a tic. The tic responds to the urge and relieves the tension that the urge has built up. The relief can be complete or partial, as will become evident from the testimonies of the patients describing their urges. The series of events, stimulus-urge-tic, is not mandatory. Simple tics are often not preceded by an urge.

Adam Seligman, who found out about his ticcing only in a summer camp, when a roommate mimicked him, started only then to experience an urge to tic. He distinguishes between *tics* and *twitches*. "A tic, to me, was a rapid uncontrolled involuntary movement of a small range; an eye blinking, a grimace, a small jerk of the hand" (Seligman, 1992:35). In other words, his urge-less tic is a *simple tic*. By contrast, "A twitch was what my neck did several times a minute; a violent large movement that quite often produced pain and had an internal feeling proceeding it, a sense of energy building up that had to be released by movement" (35). According to Seligman, then, simple tics come without urge. The complex tics release energy, which gets built up and experienced as an urge. A similar distinction is made by Danielle. Reflecting on the question of the urge, she says:

"Hmmm, it's hard [to say] because my tics are so continuous there is almost not a break in between; so it's hard to say that there is a really stop beforehand. I kind of just… almost like a constant thing." Yet, with further reflection she speaks about tics that are preceded by an urge, like her arm tic (S. R. Finkelstein et al., 2007-08-17a).

That simple tics are urgeless was also reported in the EEG study of Obeso and colleagues. In this study of 6 TS male patients (14-38), the EEG events prior to the simple tics were compared within-subject to the EEG changes prior to voluntarily mimicked tics. Voluntary jerks were prefaced by a pre-movement negative potential commencing about 500 ms prior to the muscle electromyography (EMG)[79] discharge. But no such pre-movement potential was evident in the EEG prior to spontaneous simple tics in five of the six patients (Obeso, Rothwell, & Marsden, 1981).

Lionel suggests a different distinction between tics that are preceded by an urge and those that are not. For him, the urge for motor tics is *always there* and just gets intensified and quickly rises before a tic. By contrast, vocal tics and coprolalia need a trigger other than the urge: "I don't feel, physically feel, in my body the coprolalia." (S. R. Finkelstein et al., 2007-11-07). Henry describes an urge that is always there, never gets completely satisfied, and erupts as a tic when it reaches certain "energy level" (S. R. Finkelstein et al., 2007-07-23).

To communicate their experience of the urge, participants describe it with analogies and gestures. Ted compares his premonitory urge to the sensation preceding a sneeze. "I can feel it coming. It's just like that sneeze (pressing with his left thumb and index finger the tip of his nose as to produce the feeling) and you don't want it to come.

[79] In EMG, the electrical potential generated by muscle cells when they contract is recorded. Measured EMG potentials range between less than 50 μV and up to 20 to 30 mV, depending on the muscle under observation.

You do everything you can to stop it (opens up his hand and bends his head towards it touching the tip of his nose to the inside of the hand) but eventually it comes out (separates his hand from his face) and that's what the Tourette's is." (S. R. Finkelstein et al., 2007-08-06). This pre-sneeze analogue is the most prevalent description given by TS patients when they share their premonitory experiences. It is effective since it describes the urge in terms familiar to all. It also alludes to the possibility that TS is on the extreme end of an experiential spectrum common to all. Neil uses another analogue: After touching poison ivy, "you have an itch, you scratch it, it feels better, for a few seconds or maybe a few minutes and then it starts itching again" (S. R. Finkelstein et al., 2007-08-22a). Chuck compares the urge to a mosquito bite, after which one is compelled to scratch (S. R. Finkelstein et al., 2007-08-24).

The psychologist Peter Chadwik, himself afflicted with Tourette syndrome, describes his urge and in addition hypothesizes that it might use the same circuitry as the sensation that we experience before sneezing or itching: "The heavings…began in my tummy and over the course of a second or two rose up within my body past my chest eventually to my mouth, throat and vocal chords as if I was being 'verbally sick.' I…suggested that the impulse might make partial use of the same circuitry as that in the sneeze mechanism, as the heavings..." (cited in Carter, 1999:55).

A search for this circuitry is the theme of an event-related fMRI study conducted by (Bohlhalter et al., 2006). Ten adult (19-49) probands were scanned, and their brain images at 2 seconds prior to the tic-onset and at the tic-onset were analyzed. A network including the anterior cingulate cortex (ACC), the insula, the thalamus, and the parietal operculum (PO), which are activated during an itch, an urge to scratch, and an urge to

respond to pain, was activated during the premonitory urge of TS -- indeed, as was hypothesized by Chadwik and in agreement with the experience of the urge by many TS patients.

Where is this urge experienced? Is it a thought? Is it a physical sensation? For Henry "it's all; everything. It's from here (moving his right hand across his neck, producing a gesture similar to describing a killing, and then raising his two hands on both sides of his face) up. Everything is from here (the neck) up. The urge comes from here (putting his hands on his head, above the temples). Yes it's on my mind now as I speak."(S. R. Finkelstein et al., 2007-07-23). For Claire the premonitory sensations are "all over," and they start "from here" she points at her head. She explained it all with "the OCD. I can't stop it. It has to be just right. Until it's the moment I feel comfortable and then I stop." And she added, "I feel better afterwards."(S. R. Finkelstein et al., 2007-12-21).

For Elliot, with his tactile over-sensitivity, the urge is in the same location where the tic would follow, "I have the urge in the location where the tic is. Like right now it's in my shoulder… I can feel the location... I can feel the other parts of my body getting involved in that. They get set off" (S. R. Finkelstein et al., 2007-08-13). Much like Elliot, Chuck too has the urge where the tic would be, "right here that's where it [the urge] is for me." He further explores the question. Tapping his leg, he describes how a motor tic is triggered when something is put close to his leg. (S. R. Finkelstein et al., 2007-08-24).

Seligman describes the urge before a coprolalic tic: "First it felt like a physical tic; involuntary, uncontrolled, rapid. But soon it built up into a stronger feeling of internal tension, more like the feeling in my neck but less violent" (Seligman, 1992:36). A

participant in the 1991 TS conference told the director of the documentary *Twitch and shout,* "It's enormously uncomfortable before the tic. It feels like I'm itching inside. It's a sensation through all my body from my fingertips to my toes. And it's coming up centered either in my neck for a neck tic or at my vocal cord" (Chiten, Medley, & Russel, 1994).

Not all premonitory urges are alike and those that precede falling tics are different from the need to scratch or sneeze. They are also insuppressible. They only provide a warning that can be used to avoid a serious injury from the fall. Henry calls this sensation "premonition," and this is how he describes it: "My head swells up; it's filled up. I sense the presence of something. I can almost smell something, almost taste. It is rising from deep within my soul, deep within my heart, coming to the surface." And he compared it to what once was considered as being possessed, and added, "I am not demon-possessed" (S. R. Finkelstein et al., 2007-07-23). Lionel says, "I could feel it coming… Something in the stomach like moving and when this feeling came to my head I knew that I was going to collapse. I would have maybe 20 seconds [to lie down to avoid injury]" (S. R. Finkelstein et al., 2007-11-07). Danielle describes her premonitory sensations prior to a falling tic: "[It's] a weird feeling… that [something] would happen… [It can be] just a little tingling, [or it can be] like aura... Sometimes I would almost feel like I was outside my body… Just this weird feeling before… Everything was going kind of slow. My face started tingling… Sometimes I'd feel… just funny… kind of numb-ish… Something was about to happen; and a lot of times my eyes would start kind of twitching… Lots of times I would fall" Like Lionel, "if I knew [a body tic] was coming I would go ahead and lie down somewhere on the floor or wherever. Because there have been a few times where I

fell down the stairs" (S. R. Finkelstein et al., 2007-08-17b). The falling tics, then, seem different from all other tics not only in their observable patterns but also in their premonitory sensations. This raises again the question that was introduced above about whether they are tics or epileptic seizures.

Sensory tics

Tics that are preceded by an urge serve to relieve it. Shapiro raised the question about how voluntary or involuntary they are. He argued, "An essential feature for all tics, in our opinion, is that they are involuntary, unintended, have no psychodynamic purpose." (Shapiro et al., 1988:346). Therefore tics that are *intended* to relieve a disturbing sensation, that have a voluntary aspect, should be recognized as "sensory tic subtype of Tourette's disorder" (34); in contrast with "classical tics."

Shapiro, guided by whether the action is voluntary, "shifts" the tic from the action that intends to relieve the urge to the urge itself, which is involuntary. He calls any somatic sensation, which is relieved by an intentional motor or vocal tic, a *sensory tic*. The tic then is not the motor or the vocal observable behavior but rather its sensory trigger. The motor and vocal are not tics any more but "movements, which utilize voluntary muscles in any part of the body, [and] are intentional." (Shapiro et al., 1988:356).

Joseph Bliss, A patient with a 62-year history of TS, describes his 35 years of self-observations of the subjective events that precede, accompany, and follow his tics. He shares his self-observations: "Each movement is preceded by certain preliminary sensory signals and is in turn followed by sensory impressions at the end of the action. Each movement is a *voluntary* capitulation to a demanding and restless urge accompanied

by an extraordinary subtle sensation that provokes and fuels the urge" (Cited in Pringsheim & Lang, 2005:15, my italics). Like Shapiro, he understands the response to the urge as voluntary. But others consider the response to the urge as a tic, even when they acknowledge that this response, at least in part, is voluntary.

The "just right" sensation

A common behavior, reported both in the TS and the OCD literature is the need to do things until they feel right. A whole section is dedicated in the *Handbook of child and adolescent obsessive-compulsive disorder* to this phenomenon in children with OCD. It often manifests itself in the need to perform perfectly, thus writing a letter or a paper over and over again (Ledley & Pasupuleti, 2007: 339). Henry describes such behavior and attributes it to his OCD, "I write and I write and it takes me hours and hours and hours." And then he shows me a notebook filled in with small letters with hardly any white space between them. This behavior is not ticcing. On the contrary. It suppresses Henry's tics: "You can't jerk when you write. You have to be still. So energy starts to build up, then builds up... and it builds up and builds up, and across over a certain point I can't put it down… I miss most of my lunches because of this… It's a driving force." Finally, when the urge to tic overcomes the compulsion to write, his tics emerge (S. R. Finkelstein et al., 2007-07-23).

But the need to have it just right can lead to tics rather than compete with them. In a different time during the interview Henry speaks about ticcing "[t]ill I do it right" (S. R. Finkelstein et al., 2007-07-23). Mr. O. confesses to Meige and Feindel about "the craving to keep my head in a *correct* position" (Meige & Feindel, 1907/1990:8, my italics). And one of the participants in *Twitch and shout* describes his vocal tics: "When the tic

happens it may not satisfy this pressure [to vocalize]. I have to do it again *till I do it right*" (Chiten et al., 1994, my italics).

The "just right" feeling is observed across all tics – vocal and motor, elaborate and coprolaliac. Steven shares: "One time I was stuck on a tic for 4 hours. I was trying to get certain noise out. I was like (opens up his mouth to demonstrate). It wouldn't come out so I practically, to be honest, I was on the floor trying to get it out and eventually it did. I was so relieved." (Finkelstein et al., 2007-09-28). Kyle describes, "I would have to get the cuss word just right before it would stop. And I had to say it over and over and over, and it has to sound *just right* to me. And I keep doing it" (S. R. Finkelstein et al., 2007-09-21). Chuck has the need for both the vocal and motor tics to feel right, "I got to hit it the right way. You got to hit it until it feels right. Kind of like the noise. You got to make the noises till it feels right" (S. R. Finkelstein et al., 2007-08-24). Claire, who screams a lot, needs to do it "three, four times, six times, until it feels right" (S. R. Finkelstein et al., 2007-12-21).

The need to get things right is extended to the general behavior. Neil discusses his language – speaking and writing. His rehearsals before speaking over the phone intend to prepare him to say things "perfectly" and are so intense that they cause him to almost stutter. "It needs to be perfect." He is thinking about what he will say over and over. As for writing, "I don't like writing…. People say I am a good writer… [But] it's so hard for me. Probably some of it is that's obsessive… It just feels like I got to find the *right* word, and it's probably not *the* right word... When I write handwriting I'm scratching a lot. And so a lot of my notes are… illegible… Some of it is because I start a thought, writing a

thought, and I change my mind. It's more my impulsive nature" (S. R. Finkelstein et al., 2007-08-22a).

The internal experience "to get it right" is common enough that Leckman and colleagues have named it the "'Just right' perception" (1994). In a study with 134 people (9 – 71), 81% of those with both TS and OCD, and 56% of those with only TS reported being aware of a need to perform compulsions until they were "just right." Is "just right" a compulsion? And if yes, what is the meaning of having it and yet not being diagnosed with OCD? After all 56% of those who were diagnosed with only TS reported it? Clearly the borders of the definitions of TS and OCD are blurred.

Urge-tic vs. obsession-compulsion

Shapiro, insisting on only biological etiology for TS, and understanding OCD as psychopathology, rejected any possibility of including OCD it TS. In that he was with agreement with Gilles de la Tourette but not with Charcot, who spoke of "ideational tics" that impose themselves on the ticcer. (H. I. Kushner et al., 1999). The whole chapter 6 of the Shapiro's book is dedicated to an impassioned opposition to including obsessive compulsion disorder (OCD) and obsessive compulsive symptoms (OCS) in TS. This is in spite of being "surprised that so many patients had OCS-like symptoms," (Shapiro et al., 1988:240).

Shapiro distinguishes between OCD and OCS only by severity: "OCD [unlike OCS] requires marked distress and interference with functioning" (1988:233). It seems, even though I could not find any discussion focusing on this nomenclature explicitly, that today, Shapiro's OCS is called by some obsessive-compulsive behavior (OCB) (for example Albin, 2006). Mary Robertson speaks of OCB or sometimes of OCS/OCB in the

sense of Shapiro's OCS. "[T]he obsessive-compulsive symptoms (OCS) and obsessive-compulsive behavior (OCB) encountered in TS may well be describing one and the same phenomenon, but that they are clinically and significantly different from the OCS encountered in pure OCD" (Mary M. Robertson, 2000:430). For her, "pure OCD" is an anxiety disorder related mainly to anxieties about cleanliness. Juncos has also observed that the TS obsessive-compulsive nature is "more primitive" and not related to cleanliness (private communication).

Shytle and Wilkinson speak of "many disorders that have characteristics involving repetitive thoughts and behaviors, like those that occur in people with OCD. These disorders are sometimes called Obsessive-Compulsive Spectrum Disorders (OCSD) because of these similarities" (2007:47). They consider TS an OCDS comorbid with OCD; comorbid rather than belonging to the same spectrum. In that they are like Shapiro. Relying on patient testimonies, Shapiro says, "Patients describe these symptoms as different from their tics because they are intended as voluntary acts, whereas their tics are involuntary." (241). While sensory tics are classified by Shapiro and his colleagues as legitimate TS involuntary tics, the OCS's behavior is not. The involuntary trigger "legitimized" the *sensations,* not the motor or vocal behavior, as sensory tics. The Shapiro group excludes the semi-voluntary *behavior* of the "just right" phenomenon from TS.

In the rest of this discussion I will often speak of OC in manner uncommitted to whether it is OCD, OCS, OCB, or OCDS. While it becomes obvious that study of TS cannot be divorced from the study of obsessions and compulsions, the subtle distinctions within the various OC behaviors have not been resolved within the community. The

insistence of Shapiro of separating the psychopathological from the neurological is no longer advocated (James F. Leckman & Cohen, 1999; Mary M Robertson & Orth, 2006).

The descriptions of the "just right" perception suggest both similarity and difference with tics. While often the urge preceding a tic gets satisfied, completely or partially, by *one* tic (even if for only short period of time), the obsession gets satisfied only in *a series of compulsive steps*. The urge-tic is a onetime process. By contrast, the obsession-compulsion is a multi-step process; as though each compulsion approaches only partial satisfaction of the obsession and needs to be repeated until the goal, a full satisfaction, is achieved. The description in *Twitch and shout* states it most clearly: "When the tic happens it may not satisfy this pressure. I have to do it again till I do it right" (Chiten et al., 1994).

Stuart distinguishes between urges to tic and compulsions based on the temporal patterns. The motor tics -- "it's [a] relief to do them because you want to do them, and then when you do them it's kind like it's over for a little bit." But it seems different from compulsions, "They can take time unto fruition." But he also noticed similarities; especially between his coprolalia and his pornography-related OC: "Well, there's definitely an urge there [in the OC]. I think about sex until something happens. It's part of the OC spectrum" (S. R. Finkelstein et al., 2007-10-26).

Neil's description of his language-related OC is also a step-wise process towards completion of a need. But his process, even though extreme in its intensity and in how involuntary it is, is familiar to all who write. We first write a draft, which is good but not good enough. We then correct it and get closer to the desired stage, but not quite. And so we continue until *it feels right*. The "just right" perception is a common phenomenon

among TS patients and one that demonstrates similarities not only with tics, but with what considered normal as well. The frequency, intensity, and how voluntary the behavior is define the border between the normal and the pathological and separate between pathologies.

Voluntary or involuntary?

The question of whether responding to a tic or suppressing it is voluntary or involuntary needs to be studied along a continuum rather than in a binary manner. Some of it will be addressed in relation to symbolic tics. And I will say more about it in the chapter "Interventions" since the level of volition plays a role in the choice and success of interventions.

WHEN TICCERS DO NOT TIC

There are some activities and situations during which tics cease altogether or at least get reduced. Sleep was discussed above. There are more.

Which stimuli do not trigger tics

I have discussed the effect that visual and somatosensory stimuli have on tics. By contrast, acoustic stimuli seem to not have such an effect. For Stuart the world of sounds provides an escape from the visual "That's why the audio world can be comforting. You… are tuning out the reality of the visual" (S. R. Finkelstein et al., 2007-10-26). When reading fails them, Neil and Danielle listen to audio books (S. R. Finkelstein et al., 2007-08-17b, 2007-08-22a). For Claire, like for most people, too loud stimuli are disturbing. But "otherwise no. It [the noise] has nothing really to do with [the tics]" (S. R.

Finkelstein et al., 2007-12-21). Elliot who elaborates on how the visual world is the main trigger for his tics says, “No, sounds don’t bother me” (S. R. Finkelstein et al., 2007-08-13).

Which activities ameliorate tics

Many have observed that some activities reduce tics. Meige and Feindel cite Charcot’s comment on the reduction of tics during gymnastics (1907/1990:300). Oliver Sacks reports about two TS patients: “[Ray] was free from tics when he swam or sang or worked, evenly and rhythmically, and found ‘a kinetic melody’” (Sacks, 1970/1998). And “Bennett… was pedaling furiously on an exercise bike… He can read, thus calmed without his usual obsession and distractions. But as soon as the rhythmic cycling stop, a flurry of tics… took over” (Sacks, 2004). Sacks’ interpretation, according to which the rhythm stops the tics, was not shared by the participants in my study, even though it was not excluded.

Elliot is tic-free during concentrating and sex. And he further interprets: “The sex is concentration, I think” (S. R. Finkelstein et al., 2007-08-13). Steven observes: “Anything creative gives me a lot of relief” (S. R. Finkelstein et al., 2007-09-28). Ted tics less when he is focused (S. R. Finkelstein et al., 2007-08-06). Chuck’s tics reduce “when I am really focused on something.” He focuses when he writes, uses a keyboard or a mouse, or plays softball or racquetball. He also does not tic when he works .hard, “I’m doing, I’m busy, and I’m work[ing], work[ing] with the kids… Sometimes I tell [my wife] that I don’t have time to make my noises” (S. R. Finkelstein et al., 2007-08-24).

When he listens to music, Kyle’s tics lessen or disappear altogether. He thinks that this is because music puts him “in focus.” He enjoys “loud, crazy [music] that

changes and has a lot of beats and a lot of bass." With such music, "I can get into it; just go with the flow of the music" (S. R. Finkelstein et al., 2007-09-21).

Daniel, even before his DBS, has not ticced during video games (S. R. Finkelstein et al., 2008-06-04). Dylan, who did not stop ticcing before his DBS and could not stay in a regular classroom, describes his sports skills: "As a child [I was a] pretty good baseball player; pretty good athlete in spite of the tics; swirled but when the ball came, I would stop whirling. When I had to stop and catch the ball, I stopped to catch the ball; always just at the right time… I am a good golfer. I am left handed… pretty accurate, pretty good… [The tics] slow down… I concentrate" (S. R. Finkelstein et al., 2007-08-01a).

All the examples given by the participants in the study involve concentration and focusing. But maybe more importantly, they are all activities chosen by the person with high self-motivation.

CONCLUSION

The subjective experiences of tics, as shared by the participants, have led me to identify triggers of tics. Visual stimuli trigger tics. Somatosensory stimuli trigger tics. They are complex and at times lead to conflicting behaviors. They might be driven in part by OCD. Sound seems to not trigger tics. The social world contributes much to tics but has been left for the next chapter "Symbolic tics." The internal world of thoughts and feelings can trigger tics. They may be driven by OCD. The "just right" phenomenon seems much like OCD too. The urge that precedes many tics is an important aspect of ticcing and raises many questions about volition and intention. .

Recognizing conditions that reduce ticcing is one key for self-help. Avoiding the tic triggers, if and when possible, is an obvious strategy to ameliorate tics. But in

addition, many participants report less ticcing during self-motivated activities that require high concentration. I will return to this in the chapter “Interventions.” But before addressing the important issue of intervention, I will discuss the tics with the greatest social implications, symbolic tics, in the next chapter.

SYMBOLIC TICS

I have elsewhere spoken of swearing and of other similar ejaculatory sentences. These seem to border on emotional actions, or, as I have already put it metaphorically, "oaths are phrases which emotion has filched from the intellect to express itself in a more definite way than it could by mere loudness of tone or manner." We scarcely say anything when we swear, although we utter words in relation. The secret of the perfect utterance of these phrases… [is] that they are easily elaborated by long habit, and can thus be brought out by a slight stimulus..

J. Hughlings Jackson in the "Notes on the physiology and pathology of language" *Medical Times and Gazette,* 1866.

CHAPTER CONTENTS

IN THIS CHAPTER I focus on coprophenomena. *Coprophenomena* is the collective name for coprolalia and copropraxia, regardless to modality. The "copro," from the Greek *Kopros*, dung, emphasizes the foul nature of the behaviors. I argue that coprophenomena are not necessarily foul, rather that they are characterized by being *socially and culturally forbidden*[80].

First I discuss the sense in which the coprophenomena are symbolic. Then I describe them statistically, behaviorally, psychologically, and neurologically.. I explore the impact of this involuntary behavior on the patient and his or her social environment, and I consider ways to ameliorate this impact.

"SYMBOLIC"

This is a chapter about symbolic tics. "Symbolic" is widely used in everyday life, and it takes different meanings in the different contexts of philosophy, psychology, and sociology. My use of the word is unlike Freud's and rather like C.S. Peirce's.

Freud's symbols

Freud's notion of symbolism is rooted in his dream interpretation. Dreams according to him are "[i]ndirect representation – the replacement of a dream-thought by an allusion, by something small, a symbolism akin to analogy" (Freud, 1905/1960:105). The symbol substitutes, replaces, an "objectionable element by one that is indifferent and that appears innocent to the censorship" (212). Through the process of psychoanalysis, which is based on free associations, a fragmented, incoherent, "text" gets worked upon to

[80] I distinguish cultural from social. A cultural rule does not depend on the situation. For example, one has to honor one's parents. The social is more situational. For example, American young kids are taught to distinguish between "outdoor voice" and "indoor voice."

become a coherent narrative. This is possible only through the process of "reducing disguises," to use Paul Ricoeur's expression (Riceour, 1970:30). Dream interpretation deciphers the primitive elements of dreams, the disguising symbols, into a cohesive story told in the rational language of analysis (Frieden, 1990). Criticizing Charcot's neurophysiological approach to hysteria and extending his interpretation of dreams to understanding hysteria, Freud had seen hysteria behavior "as though anatomy did not exist or as though it had no knowledge of it" (cited in Harrington, 1987:251). Hysteria for Freud was a *symbol* for repressed emotions.

The disagreement between Charcot and Freud extended from hysteria to *maladie des tics*. Freud considered tics, at least in their beginning, as symbolic behavior; Charcot considered them as a neurological disorder (for a full discussion see H. I. Kushner, 1999)[81]. And while Freud did not deny the likelihood of neurological contribution to tics, the psychoanalytic tic therapy, especially as practiced by Margaret Mahler, depended mainly on dream interpretation. Even though Mahler, like Freud, did not deny neurological susceptibility to tics, she considered their manifestation as due to the psychic rather than the somatic (H. I. Kushner, 1999:110-112).

The psychoanalytic interpretation calls for a "dictionary" that would map the behavior, which is considered symbolic, to the repressed events and impulses, which it symbolizes. In this psychoanalytic dictionary the etiology of neuroses is sexual repression (Freud, 1961). The therapist attempts to help the patient get rid of his or her symbolic tics by converting elements of the patient's unconscious into a cohesive narrative. This conversion relies on understanding the unconscious as constructed from past events and

[81] The case of Freud's patient Frau Emmy von N., whom he diagnosed with hysteria, while others consider her to have suffered from *maladie des tics*, is of special interest in Freud's understanding of tics (H. I. Kushner, 1998). However, here my emphasis is on Freud's concept of symbolism.

sexual impulses. However, often the patient experiences this creative narrative, no matter how cohesive, as a fiction. Somehow the therapist's dictionary does not fit with the patient's subjective experience.

Earlier we met the eleven-year-old Pete, who was described by his psychoanalysts Margaret Mahler and Irma Gross as a patient with a typical example of tic syndrome. Their treatment originally included free associations and dream interpretation and was directed to Pete's sexual problems. Mahler believed that these were his basic problems. Pete, whose tics did not improve, shared his feelings that psychoanalysis was "witchcraft and superstition" with his mother. In confronting his therapists, he said "You told me the tics came because of mixed-up feelings and lots of those feelings have to do with the sex business. Well, I've had the tics since I was about 3 years old. How does that fit in?" (H. I. Kushner, 1999). So while the psychoanalytic narrative made sense to Mahler, it did not seem truthful and meaningful to Pete; nor did it relieve him of his tics and bad language.

Deservedly, Freud has been studied by many with great subtlety and depth. We are greatly indebted to him for bringing the unconscious to our consciousness and for a refined study of the conscious, preconscious, and unconscious (Freud, 1939/1967). However, in my tic classification, *symbolic* is in the sense of the American philosopher Charles Sander Peirce (1839-1914).

C.S. Peirce's symbols

C.S. Peirce presents three kinds of referential associations that relate to each other hierarchically. In *The Symbolic Species* the anthropologist Terrence Deacon introduces this Peircean system (Deacon, 1997)[82]. In it there are icons, indices, and symbols. *Icons*

[82] Some of the following examples are mine.

have some similarity to what they stand for. For example, a small drawing of a file on a computer screen represents a file, and a photograph of a person stands for the person. *Indices* have temporal or spatial causal connections to the object that they indicate. For example, the footprint of a man's foot indicates that a man has walked here, and a reading on a thermometer indicates the temperature of the water. Most animal communication is through indices. Pheromonal odors, for instance, indicate an animal's proximity. Finally, *symbols* are linked to what they symbolize by agreements among their users, and they can be arbitrary. The agreement might be explicit, as in a contract, or implicit, as in social conventions. Most words are symbols. And grammatical rules are implicit contracts. Languages are symbolic in Peirce's sense.

COPROPHENOMENA – FORBIDDEN LANGUAGE

The word coprolalia was invented by Gilles de la Tourette and his mentor Jean-Martin Charcot. For them the behavior was a diagnostic criterion. Gilles de la Tourette's emblematic case for *maladie des tics* was the Marquise de Dampierre, even though he had never examined her himself and learned about her only from the reports of her physician, Marc Gaspard Itard. I introduced her in the chapter "Tourette's – a syndrome not a disease." Her shouts were sudden. They interrupted conversations, and they made "a deplorable contrast with her intellect and her distinguished manners" (H. I. Kushner, 1999:10).

To remove the foul connotation from the behavior, I replace *copro* with *forbidden*. I replace "coprolalia" with *forbidden speech*, and "copropraxia" with *forbidden gesture*. And as the current consensus in the cognitive linguistics community is

that gesture is part of language (Goldin-Meadow, 2008; Kendon, 2000; McNeill, 1996), I replace the across-modality "coprophenomena" with *forbidden language*.

"Cursing" or "swearing," as they are often used interchangeably, would do too. The Australian linguist Ruth Wajnryb has observed that even though the most common expressions in voluntary cursing are of sexual or religious nature, the complete cursing vocabulary goes beyond these two domains to include all that is *socially inappropriate* (Wajnryb, 2005). In that sense, coprophenomena could have been called "cursing" or "swearing." But I use these words seldom, since in everyday use people tend to limit the meaning of cursing to the domains of sex and religion. I use them when I want to highlight the emotional contents of the forbidden utterance, and always in the sense of Wajnryb – this which is socially inappropriate.

Descriptive statistics

In my own study, the majority of the 16 adults with Tourette syndrome (ATS) are afflicted with symbolic tics. Eleven (69%) use forbidden speech, and 8 (50%) use, in addition, forbidden gesture. Not a single person in the study is afflicted with forbidden gesture unless they are also afflicted with forbidden speech (Table 8.1). This correlation has similarities and differences with regular linguistic behavior. In regular language, gestures are used to accompany and enhance speech rather than being independent (Kendon, 2000; McNeill, 1996)[83]. Therefore people either only speak, or speak and gesture, but rarely do they only gesture. This is much like the population of my study, in

[83] It is important to distinguish between sign language and gesture. Sign language used by the deaf is equivalent to speech. In addition signers gesture, and their gestures are distinguishable from their signs (Lidell, 2003).

which those who use forbidden gesture are sure to use forbidden speech, but not vice versa.

ID #	Participant	Forbidden gesture	Forbidden speech
1	SRA	1	1
2	LRA	2	3
3	TQA	3	3
4	KGA	3	3
5	DQA	1	1
6	HBA		1
7	SZA	1	1
8	EEA		
9	CBA		
10	NFA		
11	DLA		
12	NYA		1
13	CFA	2	2
14	DGA	2	3
15	LLA		3
16	DGB		
	COUNT	8	11
N=16	**%**	50	69

Table 8.1[84] Forbidden language distribution in my pilot study. The numbers indicate intensity 1-mild, 2-frequent and forceful, 3-very frequent and very forceful.
Forbidden speech has been reported and demonstrated by 11 participants (69%), and forbidden gesture was demonstrated by 8 (50%) of the participants. Not a single one has forbidden gesture without forbidden speech.

On the other hand there is a difference. In the normal use of language, gestures co-occur with speech. In the temporal analysis of the tics that I discussed in the chapter "Tic classification," vocal and motor tics rarely co-occur, including forbidden speech and gestures. This suggests different relations between regular speech and gestures and forbidden speech and gestures.

A possible explanation to the greater prevalence of forbidden speech over gesture is that gesture is physical and might be perceived as more threatening than speech.

[84] The original TSSR distinguishes between modalities and uses "coprolalia" and "copropraxia." Since this is a self-report and the participants interpret coprophenomena as forbidden, I have taken the liberty to change the headings of the columns accordingly.

Therefore, if any volition contributes to the behavior, the forbidden speech is preferred over gesture, since its impact is less threatening.

The high percentage of probands afflicted with forbidden language in my study is by design and does not represent a randomly selected sample of ATS. One of the goals of the study was to explore the nature of forbidden language. Thus more than 50% of the participants demonstrate forbidden speech (See Table 8.1 for the participant distribution).

The DSM-IV-TR estimates that less than 10% of the Tourette syndrome (TS) population is afflicted with what they call coprolalia. In a large international database of Roger Freeman and colleagues, 13% of the 6805 TS participants have what he calls coprophenomena (R. D. Freeman et al., 2000). In a more recent study conducted in 15 sites in 7 countries, Freeman and colleagues have found higher numbers. Forbidden language occurred in 18.6% of males and 14.1% of females but only 7.0% of males and 3.8% of females had forbidden gesture. The mean onset of forbidden speech was age 11.3 and 10.1 for gesture; and the mean onset for forbidden language, regardless to its modality, was at about 5 years after the onset of the disorder. For 11.3% forbidden speech began before age 6 and in 6.4% after age 18 (R. Freeman et al., In process).

Forbidden language violates culture

The above onset ages of forbidden speech are puzzling. What is the meaning of a 6-year-old using forbidden language? Many of the words that make up the forbidden vocabulary are not yet understood at this age. They surely do not have the same meaning that they have for a 20-year-old. But while the semantics of a forbidden word might not be understood by a 6-year-old, the social knowledge that it is forbidden, regardless to its meaning, is already in place.

Evidence from the developmental literature

A growing body of research provides evidence that at the age of 6, children have internalized the social rules of their culture. In a 2004 review article, Cosmides and Tooby cite a series of studies by Núñez and Harris in which a child had to identify the picture in which a character is violating a social rule. For social contracts, British 3-year olds chose correctly 72%-83% of the time, and 4-year olds chose correctly 77%-100% of the time. By age 5, children could solve the full array of four-card Wason task[85] when the conditional rule expressed reasoning based on a social contract (Cosmides & Tooby, 2004).

In *The cultural origins of human cognition*, Michael Tomasello draws a developmental chart of understanding the behaviors of others, which is assumed to be concomitant with the level of social cognition. The beliefs, plans, and desires of others is already understood by the age of 4 (1999:180). In discussing moral reasoning, Tomasello makes a distinction similar to the one I suggest: That even though language is applied, "[n]ote again that it is not the *content* of the language that is crucial… but the process of engaging another mind in discourse dialogically" (181, my italics). And finally, Tomasello cites the observations of Palinscar & Brown and Gauvain & Rogoff that around the world children 5-7 are seen as entering a new phase of development, and that this is the age where schooling starts. Children at this age are able to use social and moral rules in a self-regulating manner (191).

So it seems that forbidden speech that starts at the age of 6 does not depend on the semantics of its vocabulary. It rather demonstrates the cultural and social sensitivity of

[85] The four-card Wason task is logic puzzle using 4 cards. It has been accepted as a reliable and valid test for the understanding of social relations.

the child, the knowledge that a certain word is culturally forbidden and socially inappropriate. And along with this knowledge comes the violation of the cultural and social prohibitions.

Evidence from people with TS

Forbidden language, whether in the form of speech or gesture, is not necessarily foul. The afflicted demonstrates great sensitivity to cultural and social prohibitions and violates them. The trigger is provided by the specific social situation. The prohibition is provided by the culture. An interesting example is of Japanese who are afflicted with TS. Their forbidden language is often expressed in their prosody, not necessarily in their vocabulary. This is in accord with their culture, which interprets certain prosodies as socially inappropriate, insulting, and unacceptable (Baron-Cohen & Robertson, 1998).

Examples from my own interviews further support the understanding that forbidden speech violates cultural conventions and social appropriateness, and is not necessarily foul or blasphemous: Since third grade, Daniel has been home-schooled in a Christian fundamentalist family. With only very little exposure to other kids, he had minimal exposure to a cursing vocabulary. The worst he knew was the family-forbidden "shut up" and "doodoo." These made up his forbidden speech until he grew up and started to meet other kids (Finkelstein et al., 2008-06-04). Dylan's father described his fear of flying with Dylan, knowing that screaming about explosives would be inevitable and would not be tolerated after 9/11 (Finkelstein et al., 2007-08-01b).

Ted is Jewish. During the High Holidays he and his family – mother, father, wife, and baby – attended the synagogue, which was packed with over 3000 people. "In the middle of the synagogue the worst you can say is 'Jesus, I love Jesus.' But I said that."

(Finkelstein et al., 2007-08-06). Obviously "I love Jesus" is not foul. Moreover, in many churches "I love Jesus" is welcomed and "amen"-ed. But in Ted's situation this was an inappropriate utterance. "People are stimulating to me," says Ted: Black people evoke "nigger," fat women evoke "you are so fucking fat," a gay friend evokes, "want to suck your dick," and a woman with large breasts evokes "you got big tits" (Finkelstein et al., 2007-08-06).

Dylan's forbidden speech is also relevant to the situation. "Spick" is reserved for his Hispanic physician, "nigger" for his Black friends or Black passers-by, and "I want to have sex with you" to the woman who exercised on the treadmill next to him (Finkelstein et al., 2007-08-01a). "Nigger" is in the vocabulary of many afflicted with the forbidden speech of Tourette's. In North America, racial slurs are considered swearing, they can be deeply insulting, and sometimes, like other insults, they can evoke a violent response (Flynn, 1977). Dylan is aware of possible violent responses from the target of his forbidden speech, and is often "scared," when it is directed at a "big Black guy" (Finkelstein et al., 2007-08-01a).

Lessons from bilinguals

Studies of bilinguals and multilinguals reinforce the notion that taboo words convey emotional information and the prohibition on their utterance is cultural. Studies of the emotional response of bilinguals to swearing words show that even though semantically the words in both the native and the later-acquired languages are equivalent, they generate different emotional responses.

To test the claims that taboo words generate less anxiety in a foreign language, Harris and colleagues studied 32 Turkish–English bilinguals, with Turkish as their

mother tongue, and English acquired later in life. The participants rated a variety of stimuli for pleasantness in Turkish and English while skin conductance was monitored via fingertip electrodes. The participants demonstrated greater autonomic arousal to taboo words in Turkish compared to English. This agrees with the subjective experience reported by the participants (Harris, Ayçiçegi, & Gleason, 2003). My own experience as a Hebrew-English bilingual, with Hebrew being my native language and English acquired later in life, is that I am able to utter certain taboo words in English with relative ease; I would never utter their semantic equivalence in Hebrew, not even in private.

An autobiographical account of the language-dependent emotional difference experienced by a bilingual has been published by the English-French bilingual Canadian Nancy Huston. Her writings have been studied closely by Celeste Kinginger (2004). And some of her words, written in French and translated into English, are cited by Jean-Marc Dewaele: "[I]f I am involved in an intellectual conversation… or any linguistic situation that draws on concepts and categories learned as adult, I feel most at ease in French. On the other hand, if I want to go mad… swear, sing, yell… I do all that in English" (Dewaele, 2004:207).

Dewaele conducted probably the largest study of the emotional force of taboo words on bi- and multilinguals. Using a web questionnaire, he recruited 1039 participants who speak a total of 75 languages. Despite some methodological issues of such a study, to which the author is sensitive, there are interesting findings in this study, and especially the subjective testimonies of the participants: Kevin who speaks Finnish, English, Swedish, and German in the order listed above, says, "I very rarely swear in Finnish" (Dewaele, 2004:213). Estela chooses to insult in her mother tongue, Romanian, since "it

carries more weight and I can distinguish more nuances" (213). There are more testimonies in his study -- all manifesting the emotional power of the dominant language, which is in most cases, the mother tongue. These emotional words can either favor or hinder their use, depending on the communicative intention of the speaker.

In my study, only one participant was bilingual, Claire. Her mother tongue is German and her English was acquired later. She has been married to an American for over two decades and has lived in the US for most of her marriage. Since I had failed to ask her about her bilingualism in the interview I called her up and got her permission to ask her some questions over the phone. To my question about which language she uses when she curses, she answered "English." I probed further and distinguished between voluntary and involuntary swearing; asked about what happens when the conversation is in German. The answer was always "English" (Finkelstein, 2009-04-16).

Based on the studies cited earlier and my own personal experience, I interpret Claire's use of English for taboo words as a manifestation of the strong emotional arousal that German evokes in her. I further interpret all the evidence about bilingual swearing as supporting the notion that taboo words are recognized and coded emotionally as forbidden. The prohibition on their utterance represents cultural norms, to which children are sensitive much before they can understand the semantics of the forbidden words.

Negativity bias

No matter how poor our understanding of forbidden language is, one question that comes almost immediately after being introduced to the phenomenon is, why cursing? Why doesn't a person scream involuntarily and inappropriately, "I love you? You are

beautiful?" The *negativity bias*, the asymmetry between positive and negative valences might hold a piece of the puzzle.

In a review of the literature, Rozin and Royzman bring evidence that negative entities are stronger than the equivalent positive entities (2001). In making decisions people give greater weight to negative aspects of a stimulus than to its positive ones (Kahneman & Tversky, 1984; Peeters & Czapinski, 1990, review). For example, Rozin and Royzman cite a 1975 study by Bloom and Price showing that short-term downturn in the economy cost the party of the incumbent American president, while upturn in the economy has no effect on the elections.

In addition, the negativity of negative events grows more rapidly when approaching them in space or time than does the positivity of positive events. For example, in 1948, Brown measured the rat's tendency to pull toward food at the end of an alley, at different points in the alley. He did the same for the tendency to pull away from shock at different points in the alley. He reported steeper negative than positive gradients in terms of distance from the site of feeding or shock (cited in Rozin & Royzman, 2001). Negative reinforcement results in faster learning (Öhman & Mineka, 2001, review). Physiological arousal is greater for negative than for positive stimuli (Taylor, 1991). Brain imaging data lead to a similar understanding. Late positive potential (LPP), a component of an event-related brain potential (ERP), has been found to be significantly greater in response to negative stimuli than to positive ones (Ito & Cacioppo, 2000).

The impact of the negative is greater than of the positive. Whatever motivates and drives the eruption of forbidden language would have hardly any effect had the eruption been positive. In such a case only the eruption itself would be inappropriate but not its

contents. The negative content of the involuntary forbidden language is with great force and leaves a strong impact.

Forbidden gesture

I have classified symbolic tics across modalities. An example of a linguistic, yet non-vocal, violation is described by Cory Friedman: In an English lesson in elementary school, "[b]efore I start, I sound out the word in my mind, the way she [the teacher, Mrs. Wilkens] told us to do. Then I turn to the blackboard and get the chalk ready. But instead of writing the word of the day, my hand writes the word SHIT in great big letters" (Patterson & Friedman, 2008:39).

But most of the non-vocal symbolic tics are through the use of gesture. During our interview, Ted raises his middle finger at me (Finkelstein et al., 2007-08-06). Dylan, whose tics have been greatly ameliorated after his DBS, still raises his middle finger during the interview (Finkelstein et al., 2007-08-01a). When I ask Stuart about his forbidden gesture, the very conversation evokes the raising of his middle finger (Finkelstein et al., 2007-10-26). Donna speaks of performing forbidden gestures (Finkelstein et al., 2008-02-08), and Claire occasionally squeezes her breasts with great force (Finkelstein et al., 2007-12-21).

In deaf population there is a distinction between forbidden "speech" and forbidden gesture, but sometimes they are more difficult to tease apart. Morris and colleagues describe a 29-year-old man that was found to be profoundly deaf at the age of 10 months. The man's IQ was 120 but he demonstrated pragmatic deficits in language and possible autism. He had some tics but whether he suffered from TS or not, is not clear. "At the age of 20 his tic behavior markedly worsened and his parents felt that he

had developed Gilles de la Tourette syndrome" (Morris, Thacker, Newman, & Andrew J. Lees, 2000:319). But "when he mixed with children with normal hearing, he learned the obscene palm-back V sign and this was incorporated into his repertoire of tics." (319). It is hard to be sure whether his palm-back V sign was indeed an involuntary or a voluntary use of a new curse sign. A more detailed description of his behavior would have been helpful. In another study, Lang, Consky, and Sandor report a woman who studied sign language after being diagnosed with TS. Her forbidden language, "speech" and gesture, were expressed through signing (Lang, Consky, & Sandor, 1993).

These last two cases serve also as support against the hypothesis presented by Marc Nuwer and based on random computer-generated strings of letters, that "certain obscenities are a concatenation of high-probability sequences of letters or phonemes" (Nuwer, 1982:366)[86]. There are too many examples to support the notion that forbidden language is indeed symbolic and not simply random and accidental. Van Lancker and Cummings assert the same. Their statistical analysis, based on distribution of phonemes in English, concludes that "the Markov model is unable to account for coprolalic utterances, even monosyllabic examples" (Van Lancker & Cummings, 1999:92).

Gestures in both the speaking and deaf communities, are part of language (Goldin-Meadow, 2008; Kendon, 2000; McNeill, 1996). Like words, they can be used for communication and they are culturally constructed and shared. Different cultures use different gestures to convey the same meaning. For example, the American equivalent of the British obscene palm-back V is a raised middle finger. In Israel the middle finger is lowered and pointing. But unlike words and signs in sign-languages, gestures are

[86] I thank Prof. Roger Kurlan for the reference.

typically iconic rather than symbolic. As such they are more primitive, carrying out only the simple mapping of resemblance, and do not require the intricate and supportive network of symbols. In view of such understanding it seems paradoxical that forbidden speech is more common than gesture. But maybe the paradox can be explained by the threatening quality of the gesture, which was mentioned earlier.

In normal use of language, gestures are involuntary; words are not. Familiar is the scene in which a person in a telephone booth enthusiastically or angrily gestures to someone on the other end of the line, who cannot see the gestures (de Rutter, 1995). The correlation between the meaning of the words and the gestures strongly suggests that, while involuntary, the gestures in normal communication express the intentions of the gesturer. Therefore "automatic" better applies to describe them than "involuntary." The gestures express the intentions of the speaker, but they are automatic in the sense that they do not require conscious attention, and they are hard to suppress. Maybe forbidden gesture expresses intentions more than forbidden speech.

TS and other cursing disorders

Involuntary cursing is not unique to TS. Degenerative diseases like Alzheimer's and Parkinson's disease are often accompanied with cursing. So are some brain lesions. Socially forbidden speech often happens in aphasia, the loss of speech. A famous aphasiac was Mr. M. Leborgne. He had lost his ability to speak except for cursing "*Sacre nom de Dieu*" and repeatedly saying "*Tan,"* after which he was nicknamed. A postmortem operation on his brain was performed in 1861 by the French neurologist Paul Broca, who found a large lesion in Leborgne's left hemisphere (Harrington, 1987). Up until today, Broca's area, the left posterior inferior front cortex comprising of

Brodmann's areas [87] 44 and 45, is considered important for speech production (Nolte, 2002).

There are important differences between the cursing of TS and aphasia. The differences are in the neurological substrata, in the behavior, and in the course of the disorders. To begin with, and as I discussed in the chapter "Tourette's – a syndrome not a disease," there are no robust findings of brain lesions in Tourette's. By contrast, aphasia results from an insult to the brain.

The very content of TS and aphasia cursing are different. Hughlings Jackson, with many aphasia patients said, as I have quoted in the beginning of this chapter, "We scarcely say anything when we swear, although we utter words." But in TS the swearing often does have meaning. In that it might differ from, say, that of Tan. Van Lancker and Cumming compare the cursing of TS to that of aphasia (Van Lancker & Cummings, 1999:93), and find a few differences. Most derive from the main difference between the two cursing: In severe aphasia the swearing is the only speech preserved, as with Tan. By contrast, people with TS are most often articulate. None of the 16 probands in my study have demonstrated linguistic deficiency[88]. The obvious differences are: (i) Aphasiacs, unlike some TS patients, cannot disguise their inappropriate utterances by substitution. Donna can substitute "fuck" with "fire" since her access to language is unimpaired (Finkelstein et al., 2008-02-08). Aphasiacs cannot do this. (ii) TS inappropriate expletives often occur within a conversation, disrupting it. Aphasiacs do not have conversations. (iii) The forbidden language of TS evolves. The vocabulary changes with the years. The manner can change too; expletives can become more elaborate. But chronic aphasiacs

[87] Brodmann's areas are number-labeled regions in the cortex defined by their cytoarchitecture.
[88] For Claire, English is a second language, so she often speaks with mistakes. However, they are not due to any linguistic deficiency.

have only their fixed repertory. Typically it remains the same. (iv) There is no evidence that aphasiacs have any premonitory urge to curse, as do TS patients. (v) And finally, the natural histories of aphasia and TS are different.

The natural histories of aphasia and TS, dissolution, and evolution

Hughlings Jackson, with his many aphasia patients who had lost their ability to speak but preserved their ability to curse, reflected on the phenomenon. He attributed the behavior to a situation in which "the speechless man is seen to have lost the most voluntary or special part of language (speech), and not to have lost the more automatic language of emotional manifestation. He is in this way reduced to a more automatic condition of language" (Jackson, 1958b:134). This interpretation, according to which the automatic is preserved while the more complex and advanced gets lost, is consistent with Jackson's model of evolution and dissolution. According to this model the higher, evolutionary-younger functions, are less perfectly organized and therefore more vulnerable to damage. Thus the voluntary is more fragile than the involuntary, and language and culture are the most vulnerable to damage. The dissolution reverses the evolution (Jackson, 1958a).

However, this is not how TS progresses. The natural history of TS parallels normal development rather than reverses it. First come the eye and facial tics; then more parts of the body get involved; then the phonic tics emerge; and only then does the forbidden language emerge. While this trajectory does not describe all patients, it is the course of most TS patients (Leckman, Bloch, King, & Scahill, 2006) [89].

[89] I thank David Ritz Finkelstein for productive discussions of this topic.

In addition, there is the waning and waxing pattern of TS. This pattern reminds us of the U-shape trajectory of development, in which performance often goes from better to worse rather than from worse to better. Such a developmental phase has been observed in many areas such as stepping, language development, phoneme discrimination, written presentation, artistic creativity, face perception, social cognition, and scientific reasoning (Siegler, 2004). In an issue of the *Journal of Cognition & Development* dedicated to the U-shape development, three articles claim that the U-shape pattern results from underlying monotonic improvements whose products generate the final behavior. The product changes since even though all its elements progress, they do not progress in the same pace. Stepping, for example, relies on increase in leg strength and increase in leg weight. When the weight outperforms the strength the behavior regresses (Gershkoff-Stowe & Thelen, 2004). In learning gesture-referent relations, 18-month olds perform better than 26-month olds. This seeming regression is attributed to the fact that at 26 months the child has already learned about iconic relations. When the gesture is not iconic, the child fails to generate referential link between a gesture and its referent (Namy, Campbell, & Tomasello, 2004). Between 3 and 4 months infants integrate facial features rather than focusing on the features themselves. By 6 months they fall back to features only, and by 7 months they integrate again. This sequence is attributed to improved visual acuity that increases the informational load; and to a new orientation of seeing more upright faces due to crawling (Cashon & Cohen, 2004). In all these examples it is the new relations among the underlying contributions to the behavior that results in a U-shape rather than in monotone progression, even though the individual contributions continue to progress monotonously.

It seems that the course of TS as well as its waning and waxing might follow a dynamics similar to that of normal development. Rather than manifesting dissolution, as aphasia might, it is possible that TS manifests an alternative developmental trajectory, parallel to and competing with the normal one. In many, the TS signs and symptoms disappear in adulthood (Bruun & Budman, 1992), as though the normal development has won over. In a minority, the TS developmental processes win over, and patients continue to be afflicted as adults, often very severely.

What do voluntary and involuntary cursing violate?

The involuntary forbidden speech of TS violates the same taboos that voluntary cursing violates. The English *taboo* is a descendant of the Polynesian *tapu* (Freud, 1913/1952). It is that which is marked or set apart, not common, not part of the mundane. Whether sacred or profane, it is forbidden from interacting with (Shore, 1989:144). The notion of *tapu* is not unique to the Polynesian cultures. It is found in many other cultures and languages. In ancient Hebrew, the root *k.d.sh*. is equivalent to taboo (Freud, 1913/1952)[90]. And like with *tapu*, the prohibition on certain behaviors and objects extend to their symbols, to speaking about them[91]. This is the prohibition that cursing, or swearing, violate. The taboo on swearing is as strong as the taboo on performing what the swearing symbolizes. And the legal consequences of violating the swearing taboo are

[90] The Hebrew root **k.d.sh** is in "**k**a**d**o**sh**" – holy, "**kd**e**sh**a" – prostitute, "he**kd**e**sh**" – a shelter home for the poor, "**k**i**d**u**sh**in" – marriage, and "ha**kd**a**sh**a" – dedication. All set apart, for better or for worse.

[91] Since the behavior of swearing is taboo, so is its symbol, the very word, "swearing." One such example is in the trial of Naboth, which the Bible describes this way: "[A]nd the men of Belial witnessed against him… saying, Naboth *blessed* God and the king." (1 Kings, 21:13). Swearing in the sense of naming a deity is also forbidden. This strong inhibition is instructed in the third commandment, "Thou shalt not take the name of the LORD thy God in vain; for the LORD will not hold him guiltless that taketh his name in vain" (Exodus 20:6-7; Deuteronomy 5:10-11). Naming God is an *act* of evoking Him and should not be done in vain. This ancient prohibition on swearing demonstrates cultural intuition that utterances are speech acts.

similar to those of violating physical taboos (Graves, 1936; Jay, 1999; Montagu, 1967; Wajnryb, 2005).

Profanity makes the bigger part of any cursing vocabulary. This is universally true. (Montagu, 1967/2001; Wajnryb, 2005). Often the behavior alluded to by a profanity is taboo and this taboo extends to its symbolic representation. Prominent among utterances of profanity are those pointing to incestuous behavior. Their linguistic description is highly insulting (e.g., motherfucker). Other modes of forbidden sexual relations and behaviors, attributed either to the targeted person or to his or her family members, are also the themes of swearing (e.g., son-of-a-whore). The intensity of the insult, when referring to the mother, the sister, or the wife, varies culturally. But cross culturally, profanities describe tabooed behavior, and uttering them is taboo too (Flynn, 1977; Jay, 1999). In some societies calling a person homosexual, whether he is or not, is an insult; an extension of a taboo from the realm of behavior into the realm of symbolic language. The intensity is culturally dependent (Flynn, 1977; Jay, 1992). Religious contents are taboo too, universally (Flynn, 1977; Jay, 1999; Wajnryb, 2005). In the US, racial slurs are socially forbidden, and thus are part of American cursing (Jay, 1992).

All these themes of voluntary cursing are also the themes of involuntary cursing, the forbidden speech of TS. They were uttered or described by the participants in my study. Some were already mentioned before to support other arguments. But despite their bad taste I bring them here again to demonstrate how involuntary cursing shares its vocabulary and symbolism with voluntary swearing: Before Steven transformed all his forbidden speech into grunts, he uttered “fuck, shit, nigger, [and] fucking slut.” Lionel’s involuntary cursing includes “damn, hell, fuck” (Finkelstein et al., 2007-11-07). Ted’s

eruptive utterances include “nigger” (Finkelstein et al., 2007-08-06). Kyle utters OCD-driven “fuck, fuck, fuck,” and in addition “the 4-letter word, and em-ef” (Finkelstein et al., 2007-09-21a). Dylan’s involuntary cursing includes “fuck, whore, spick, pussy, tits, nigger, [and] goddamn” (Finkelstein et al., 2007-08-01a). Stuart often utters “Good God damn,” “Jesus fucker,” and “Jesus fucking Christ” (Finkelstein et al., 2007-10-26). Donna screams “fuck you all,” “fuck you all the urine,” or “fuck you motherfucker” (Finkelstein et al., 2008-02-08). The racial nature of Louis’s forbidden speech is expressed with “Kwanzaa” and “Kwanzaa juice”(Finkelstein et al., 2008-05-30).

Social consequences of involuntary cursing

In the West swearing has lost much of its ancient power and for more than a century, the legal system does not punish it any more (Graves, 1936). This is well illustrated by the recently published book by the Princeton philosopher Harry Frankfurt, titled *On Bullshit* (Frankfurt, 2005). In cultures other than the West, much of the taboo on swearing still exists. For example, in 1989, Ayatollah Ruhollah Khomeini, then Supreme Leader of Iran, issued a *fatwā* – a Moslem edict – requiring Salman Rushdie’s execution for his 1988 novel *The Satanic Verses* being “blasphemous against Islam” (Holcombe, 2004). But swearing, even when on its way into the legitimate mainstream, has still maintained some of it taboo power. After all, much of the behavior that it symbolizes is still forbidden.

Violating the rules of the culture with forbidden utterances can be intensely embarrassing. Itard mentioned it when he described the behavior of the Marquise de Dampierre (H. Kushner, I., 1999). The participants in my study spoke about it too. To avoid this embarrassment, many of them give up relations and activities that they would

otherwise enjoy. Claire, who is often expelled from her favorite café, avoids public places, including shopping malls and cafés that she loves (Finkelstein et al., 2007-12-21). Donna, who cannot afford a computer, avoids using the public library computer since she is disruptive to the other patrons and is greatly embarrassed by it. And it goes beyond embarrassment: She cannot get a job and was threatened with eviction from her apartment (Finkelstein et al., 2008-02-08). Ted lost his job of fifteen years after calling a customer "nigger" (Finkelstein et al., 2007-08-06).

Can the social consequences change? There seem to be two ways that can relieve the painful social price of coprophenomena. One depends on the speaker, the other on the hearer.

The speaker

Speakers adopt several strategies. My study's participants conceal, suppress, avoid possible triggers, and disguise. Any form of control over the use of forbidden speech is affected by the interplay between the voluntary and the involuntary, the intended and the un-intended. Do TS people intend to swear but cannot inhibit the impulse? There is great variability as is told in the subjective reflections of my study's probands:

When Stuart was 19, his brother was murdered by a Black man. "[This murder] engrained some racial negative stereotypes towards Black people" (Finkelstein et al., 2007-10-26). By contrast, Dylan insists that he could not have any racial stereotypes since many of his friends are Black. His father though is not so sure: "Some of his friends are Black," he says. But he adds, "it may have something to do with some of the attitudes… in the last 30 years living in the South; may have developed some attitude

along the way to start thinking in this direction." (Finkelstein et al., 2007-08-01b). Some of Dylan's cursing seems to have intention and sometimes plays a role in defining his social territory. "Is it a little bit of a test?" I ask him. "Yes! Somebody that I don't know… I'll tic more towards them…" In the presence of a few new people, the one that Dylan likes the least gets the most tics (Finkelstein et al., 2007-08-01a). Claire who often screams "nigger!" says, "I have a lot of Black friend[s]." And when I ask her whether it matters if she likes the person or not, she first screams and then answers, "O gosh! No it don't matter. It don't matter" (Finkelstein et al., 2007-12-21).

Whether intended or not, some try to hide it. Henry covers his forbidden words under stuttering, that otherwise he does not have. When he speaks about the "4-letter word" he starts to stutter. This deforms the word, almost to non-recognition (Finkelstein et al., 2007-07-23). When Lionel's started to use forbidden speech at the age of 8, he tried "muttering the things under my breath" (Finkelstein et al., 2007-11-07). Kyle's "fuck, fuck" is so soft and melodic that it takes a while to recognize it for what it is.

Kyle has additional control. He can suppress some of his forbidden speech. "I will say [the] 4-letter word, and G-D, and bitch, and M-F," but *no racial slurs.* "I live in a Black neighborhood. I don't think that will be good" (Finkelstein et al., 2007-09-21a). Others, that can suppress some of their curses for a while, try to minimize the public impact and leave the worst for home. Elaine Shimberg, a mother of a child with Tourette syndrome, tells her readers that "coprolalia often is worse at home, when the individual feels safer" (Shimberg, 1995:31). This reminds us of Steven who "no one, no friends, no people have ever seen me to the highest degree [of screaming] as my Mom or my Dad" (Finkelstein et al., 2007-09-28). Lionel, after disguising or suppressing his forbidden

speech in public, screams it out when he comes home or goes out to the woods (Finkelstein et al., 2007-11-07). *S*till, even after the worst is preserved for home, what remains is bad -- embarrassing, and socially isolating.

Lionel has achieved some control over his utterances by creating trigger-free mental space. His involuntary use of forbidden speech started as a ritualistic sequence of "shit, damn, darn, hell, fuck," which "were not meant to offend anybody… I just had to spit it out." Its immediate trigger did not seem social. His utterances were set off by hearing or reading the letters R or T, or the number 4. He did his best to avoid them. For example, when the teacher asked him how much was 5+4 he escaped the "4" and mentally replaced it with "5." His answer therefore was "10." This set off a whole obsessive-compulsive cycle of repeating "5" to stay in the safe zone. But it also spared him the embarrassment of using in public forbidden language (Finkelstein et al., 2007-11-07). Claire has no escape. Her forbidden utterances happen mostly in public. "I don't feel comfortable being with other people, and my tics coming on. I scream, I say 'Nigger!'" (Finkelstein et al., 2007-12-21).

Ted cannot hold off a curse until he comes home. So he tries to disguise it with substitution. He often replaces "nigger" with "Nick," and "bitch" with "witch." Sometimes he succeeds, but sometimes he fails, as when he lost his job (Finkelstein et al., 2007-08-06). Dylan disguises his forbidden gesture: After raising his middle finger, he continues the gesture as though to brush hair from his forehead with the middle finger (Finkelstein et al., 2007-08-01a). Steven has achieved complete substitution. He refers to his forbidden utterances as "coprolalia contradiction… because it contradicts everything I want to say. Because I, if I look at someone and insult him, which had happened before,

it's the opposite of what I think of that person." The intention, Steven says, is not there. Steven replaced his "fuck, shit, nigger, [and] fucking slut" with a grunt: "I got rid of it [the coprolalia] by turning it all into grunts, all of it." However he does not know how long it would last (Finkelstein et al., 2007-09-28).

A common disguise for forbidden language is humor. In the height of his pain and embarrassment after uncontrollably writing "shit" on the blackboard and causing his classmates to laugh at him, Cory Friedman got the idea that "it will be better if I make it look like I'm doing everything on purpose, as a joke. The kids will think that I am funny and not just weird. So I make a few more silly faces, including a dopey grin. This makes the kids laugh again. I laugh along with them… I am feeling so bad… But I also realize for the first time that I can get kids to laugh *with* me instead of *at* me when I do something inappropriate. I can become the class clown" (Patterson & Friedman, 2008:40). Chuck applied a similar strategy for his vocal tics. He started with "raspy-froggy" voices at a summer camp before his fourth grade. This earned him the title of the "froggy man," which he enjoyed. But not for long. After a while it got on everybody's nerves (Finkelstein et al., 2007-12-21).

Neil the psychiatrist analyzed the nature of being funny, and expressed his enjoyment of being witty and quick. Then he reflected on how humor is simultaneously a bridge to others and a separator from them. "I… think that humor can be a way of building walls… not getting connected. And I probably do that too much… If you think of someone who makes you laugh all the time… you are not getting to know *them*, you're getting to know their humor… It's not necessarily somebody you would trust with your inner thoughts" (Finkelstein et al., 2007-08-22).

Some comedians and humorists have TS. Louis Centanni appeared on the Opie & Anthony show to the laughter and amusement of the crowd who observed his inability to suppress some OCD behavior while spicing his talk with dirty words (Opie & Anthony, 2005). Dan Ackroyd, who rose to stardom on *Saturday Night Live*, spoke with NPR's Terry Gross about his Tourette syndrome. He told how he approached many of his problems by covering them up "with humor" (Gross, 2004). In *Naked*, the American humorist and comedian David Sedaris described his childhood with TS and OCD (Sedaris, 1997).

Neil pointed to comedians as examples of people who overcome their shyness through comedy yet staying distant from other people: "Comedians, for example, are notoriously shy off camera, off stage, I think in general. And they don't necessarily make a lot of close friends … they get along really well with people but they don't necessarily make the strong bond connections, and I'm a little bit like that" (Finkelstein et al., 2007-08-22).

It seems that humor does not really solve the challenges of the person with TS. It might reduce some of the social embarrassment but still leaves a deep sense of alienation and isolation. "If I can change myself," says Neil, "that's a thing [being funny] I would change" (Finkelstein et al., 2007-08-22). Attempts to suppress or disguise apply to all tics, not only to the symbolic ones. And I will return to them in the chapter "Interventions."

The hearer

The burden of interpretation is on the hearer. Culture supports the process and provides the hearer with tools to decipher the intentions of the speaker. Two examples

from anthropology illustrate it. Bradd Shore tells about two Samoan social scripts for a dinner invitation. The correct way to respond to one of them is to decline the invitation. The social rules of this schema are internalized by the members of the culture and become automatic: "[T]o the people who used them, the scripts felt more like spontaneous speech, part of a taken-for-granted world of normal human intentions and feelings" (Shore, 1996:43).

An anecdote told in Harriet Joseph Ottenheimer's *The Anthropology of Language* provides another illustration of how indispensible cultural knowledge is for a correct deciphering of the intention of a speaker: Dr. Stirland, a biologist from England, visited Kansas State University, and had planned to continue to Toronto. She was overheard mentioning her plans by a Native American young man, who approached her in the parking lot and commented on how nice Toronto was, and that his family lived not too far, in Upstate New York, and that he missed them. It took a while before it dawned on Ottenheimer that the Native American young man was not narrating a family story but rather indirectly, as is customary and polite in his culture, was asking for a ride. "I miss my family" was finally understood correctly by Ottenheimer, who resolved the situation and told the young man that Dr. Stirland would fly to Toronto. The young man wished Dr. Stirland a safe flight and left (Ottenheimer, 2008).

How does one interpret a curse? The power of cursing and its ability to harm rely on how the hearer deciphers the utterance of the speaker. And like other interpretation of utterances, so in interpreting a curse, the process is mediated by the culture. The core of the tragedy of those afflicted with involuntary forbidden language is the mis-assignment of intentions to the speaker.

Raising awareness in the hearer about the non-intentional and involuntary nature of the forbidden language helps; as Chuck said about another Touretter, "[H]e couldn't help it" (Finkelstein et al., 2007-08-24). The Tourette syndrome Association (TSA) has invested much effort in making the disorder better known to the general public. Often children get permission from their teacher to introduce the disorder to their classmates in the beginning of the year, which often helps (Conners). When this approach is successful, the social sense of violation and hostility is diminished and makes room for acceptance and empathy. Dr. Carl Bennett is a respected and trusted surgeon in his community (Sacks, 2004). The (now deceased) owner of a bookstore in Atlanta's Virginia Highland neighborhood had many loyal customers who enjoyed browsing the long lines of interesting and rare books in his store despite the constant background of "fuck" (personal visits).

The acceptance though is rarely complete. When the audience is of family members who are close to the curser the responses vary. The family members often participate in the suffering of the patient and sometimes feel guilty for their inability to help. This is how Brad Cohen describes his mother in his book *Front of the class* (Cohen & Wyscocky, 2005). Sometimes, however, a family member estranges himself completely from the curser, as in the case of the TS patient John, whose father stopped participating in the family's dinners (Kaye, 1990). Dylan's brother is very close with him, but his sister estranges herself from him as much as possible (Finkelstein et al., 2007-08-01b).

It is tempting to criticize those who do not respond to the possibly benign nature of TS. However, they act automatically as well. From early age we develop an automatic

mechanism that supports the way by which we interpret intentions of others. Like with every undoing of automatic behavior, assigning a benign intention to someone who curses us is cognitively effortful. In a way the hearer is required to perform a Stroop[92] task: to understand a benign intention even when it has an expression that we automatically associate with a non-benign intention. This is difficult. Sometimes we simply forget; other times we are too slow to go through the cognitive process; and sometimes, especially if we spend much time with the TS patient, we simply get tired. But as in the Stroop training, we can automate the process that suppresses the original automatic interpretation of the involuntary cursing. This is accomplished by some even though it is initially effortful; especially by close family members and good friends.

When Dylan, Caucasian, calls his best friend, who is Black, "nigger," his friend responds with, "I am a nigger and you are a nigger." They both laugh and it brings an end to the tension and the "nigger" (Finkelstein et al., 2007-08-01a). But this is difficult. Dylan's friend needs to cognitively undo an automatic process. Their friendship is long enough that Dylan's friend had the opportunity to train himself to create a new automatic process in which he suppresses the usual association <nigger-insult> in favor of the associations <<'nigger' utterer- Dylan>,<Dylan-friend>>. This is effortful and Dylan is

[92] The American psychologist John Ridley Stroop (1897-1973) studied the ability of a person to handle a mismatch between two associations of a stimulus. In the first version of his study, 70 college students had to read the words **Purple Brown Red Blue Green** in black fonts**.** Then they had to read the series of the same words in color. However the names of the colors did not match with the colors of their fonts. An example of such a series is: **Purple Brown Red Blue Green.** The time that it took to read the mismatched series was much longer than the time it took to read the word series in black. But this difference shortened with training. Stroop interpreted the phenomenon as manifesting first, before the training, a conflict between two automatic associations -- one of reading, and one of evaluating color. Resolving the conflict was not originally an automatic process, and therefore it took effort and time. The training started forming an automatic process of suppressing the association of evaluating a color in favor of the association of reading a word. Thus, the time of reading the mismatched stimuli became with the training less effortful and shorter (Stroop, 1935).

lucky to have a friend who values their friendship enough to be willing to invest such effort.

Kyle came to the interview with his friend Monica, who used to be his roommate. She has stopped hearing his cursing. It seems she has trained herself to suppress the automatic association between a string of syllables and their semantics in favor of associating certain syllables, when they are uttered by Kyle, as non-words (Finkelstein et al., 2007-09-21b). Kyle is not always that fortunate. His stepfather not only has not attempted to go through the cognitive self-training that would allow him to interpret Kyle's behavior as benign, but he "thought that [Kyle] was just doing it to get on his nerves." (Finkelstein et al., 2007-09-21a).

But even in loving relations, the demand on the hearer is great and often hard to meet. Neil and his wife, who came with him to the interview, seem happily married. Neil tells the following: "One day I asked my wife, 'honey… how can you stand living with me?'… And she says, 'honey it's ok as long as you don't start saying fuck you bitch fuck you bitch.'" (Finkelstein et al., 2007-08-22). Neil's wife is willing to make the effort required to tolerate Neil's tics. But the cognitive effort required for the forbidden speech would be too much for her.

In a normal speaker-hearer dyad, the speaker's utterances are intentional and voluntary. The hearer's interpretation of the speaker's intention is automatic. It is fast and effortless and carried out with almost no intention. To alleviate the suffering and embarrassment of those who involuntarily utter forbidden speech, the hearer is required to take on the cognitive effort in this dyadic relationship. It is as though the cognitive effort of normal communication needs to shift from speaker to hearer, as in a seesaw.

Since now the speaker acts automatically, it is the hearer who needs to interpret the utterance intentionally and non-automatically.

The successful examples that we saw above are limited to one dyad. In many situations, though, social interactions include one-to-many relations. The one TS speaker has many listeners, which adds much complexity, and brings out a group dynamic – an important field that I will not study here. However, I speculate that as in other group situations leadership matters. If the hearer who suppresses the automatic response in favor of empathy is an alpha person, the Touretter would be accepted by the rest. Those who follow the leader would be willing to invest the cognitive effort required for acceptance. By contrast, there would be little or no effect on the group acceptance of the TS person if the empathizing hearer is of a low social rank.

Interestingly, the TS patient seems to not be impaired as a hearer. However, it is possible that further studies of the comorbidity of TS and attention deficit disorders (ADD and ADHD) would reveal some impairment of the TS patients as a hearer.

THE NEUROBIOLOGY OF INVOLUNTARY FORBIDDEN LANGUAGE

Most of the efforts to understand the neurology of language focus on the neocortex. Evolutionarily this is the newest part of the mammal brain, which is different in its structure from older parts of the brain. In humans it accounts for 90- 95% of the total cortical area. Such high ratio is considered by many to correlate with the complexity of social life (Nolte, 2002). Language, the uniquely human skill that is closely linked to culture and social life, is therefore assumed to relate to neurological activity in the neocortex. And indeed the data of lesions and imaging, and the recent mapping of neural activation in response to linguistic stimuli support such reasoning (for example see the

review by Martin, 2007). But some scientists also look into the contributions of the subcortex to language.

The subcortical system

Following the tribune brain evolutionary model of MacLean (1990), Philip Lieberman proposed his Functional Language System (FLS) model. His model attempts to explain the evolution and the neural substrata of language. His model is derived from motor activity and as such relies not only on the cortex but also on the basal ganglia and the cerebellum.

Traditionally the cerebellum was considered to be involved only with the motor system. But whole-brain scans of cognitive tasks discovered activation of the cerebellum during high-level cognitive tasks as well (Gernsbacher & Kaschak, 2003). Leiner, Leiner, and Dow have suggested that cerebellar output is directed to prefrontal as well as motor areas of the cortex (1989). To further test these hypotheses, Middleton and Strick studied the projection of the cerebellum and the basal ganglia (BG) to the dorsolateral prefrontal cortex (dlPFC). They traced a circuit in the brain of the Cebus monkey, which projects from the dentate nucleus of the cerebellum (called by some the neocerebellum), and the internal segment of the globus pallidus (GPi) to the thalamus; and from there to the fronto-cortical area 46.

Evidence from deep brain stimulation

The procedure of deep brain stimulation (DBS) can affect involuntary cursing and as such shed some light on it neurophysiology. An important and fundamental question related to brain architecture and physiology is the question about regions vs. circuits. Is it

the region or is it the circuit? If the circuit is essential for a certain function, would stimulating various regions that participate in the same circuit achieve similar results? How crucial is a specific region for a certain function as long as it is part of the relevant circuit? Currently there is no easy way to answer this question. Two points support the centrality of the circuits. (i) In different clinics, different regions have been targeted for DBS, with similar, even if not identical, results. (ii) A TS DBS typically targets one of the following: a) the internal segment of the globus pallidus (GPi); b) the centromedian parafascicular complex (CM-PF) of the thalamus, with afferents from the GP and efferents to the striatum and diffuse efferents to widespread cortical areas; and the ventralis oralis anterior complex (VOA) of the thalamus, with efferents to the motor area of the cortex and afferents from the BG and the cerebellum; c) the anterior part of the internal capsule (AIC) with connections between the dorsomedial and anterior nuclei of the thalamus and prefrontal and anterior cingulate cortex and many frontopontine fibers. All these regions participate in circuits that connect the subcortical and the cortex. The subcortical alone would not do for language. Any effect of the DBS on forbidden language reinforces the hypothesis about the importance of circuits, maybe more so than of regions.

Servello and colleagues report 18 TS patients, 10 of them with what the authors call coprolalia, who underwent bilateral thalamic DBS and were followed up between 3 and 18 months. While all have shown significant improvements in their YGTSS scores, the coprolaliac behavior has not been reported separately from the phonic tics (Servello, Porta, Sassi, Brambilla, & Robertson, 2008).

More specific information, one case study, is reported by Huetto and colleagues. They performed a bilateral stimulation in the CM-PF and in the antero-medial (limbic) territory of the GPi of a 36-year old woman. Her behavior was studied under thalamic, pallidal, sham, and thalamic & pallidal stimulation. While many improvements have been reported, with the thalamic and the pallidal electrodes on, the involuntary forbidden language has remained moderate. It improved but reappeared intermittently whenever there was a loss of mental control (Houeto et al., 2005).

Dylan and DBS

Dylan's first DBS was performed on April 5, 2006, when he was 28 years old. The targets were bilateral anteriomedial regions of the GPi. Infection necessitated the removal of the system. His second DBS was performed on December 11, 2006, again bilateral, but this time in the posterior lateral region of the GPi (Juncos, DeLong, & Finkelstein, June 2008).

His physician observed that generally there is amelioration of about 50% in Dylan's tics. He still performs forbidden gestures; but stopped touching himself and others in public, and often succeeds in disguising the nature of his raised middle finger.. He still utters forbidden speech but it's muffled down. However he has acquired a very mild stuttering, not frequent and not intense. This stuttering impairs the temporal order of his words, "I am getting ahead of myself" (Finkelstein et al., 2007-08-01a).

Daniel and DBS

Daniel's first DBS was performed on Novemebr 6, 2007, when he was 17, much younger than the recommended age of 25 (Mink et al., 2006). This allowance was due to

the severity of his TS and the lack of response to any combination or doses of medications. The target was bilateral anteriomedial GPi. His forbidden utterances disappeared but there was no amelioration in his motor behavior. On August 7, 2008 he had a second DBS, this time it was bilateral targeting the CM-PF/VOA with remarkable amelioration of his motor tics. Now Daniel has 4 electrodes in his brain. There are considerations of gradually turning some of them off, based on observations with other DBS patients that sometimes the circuits get "trained" by the electrodes. The hope is that this learning would be sustained even without the electrodes.

Lessons from Dylan and Daniel

There is still much that is not understood about DBS; neither why they work nor why sometimes they fail. In addition, there is still only little data about various targets and their impact on various behaviors. For the two participants in my study, Dylan's involuntary eruptions of forbidden language have greatly improved after bilateral DBS in the GPi. Alas, a slight temporal stuttering has been acquired by him. Daniel's forbidden speech, that had never been very intense or frequent, disappeared completely after the first DBS in the anteriomedial GPi.

A BRIEF REVISIT OF TIC CLASSIFICATION

When classifying tics by their complexity, I have identified four categories: simple, elaborate, symbolic, and violent. But I also brought the example of spitting as a challenge to the clear boundaries of this classification. Depending on the situation, spitting can be simple, symbolic, or violent. Added to this fuzziness are examples of

touching one's own or another's genitals. Is this a symbolic forbidden gesture, or is it a violent tic?

And finally, is Steven's grunt a simple phonic tic, as is viewed by others, or is it symbolic as might be experienced by him since it replaces his coprolalia? Moreover, could many phonic tics be in fact symbolic, like in the animal kingdom, in which a grunt or a bark often has meaning and is symbolic (Darwin, 1872/1965; de Waal, 2005)?

CONCLUSION

The so called coprophenomena are involuntary cursing in the sense of Ruth Wajnryb. They are not limited to sexual and religious themes but can violate any social domain, and their themes and vocabularies are culturally dependent. Their commonality is that they are *socially and culturally forbidden*. Forbidden speech is more prevalent than forbidden gesture. Together they represent a nexus where the emotional, linguistic, and the social malfunction. While voluntary cursing is often associated with anger, this is not necessarily the case for the involuntary cursing of TS. A wide emotional spectrum that depends on the individual and the situation might feed the behavior. But it almost always generates the embarrassment that comes with violation of social prohibitions.

Not all involuntary cursing are alike, and the forbidden utterances of aphasia differ from those of TS. Neurologically, there is still uncertainty about which parts of the cortico-striato-thalamo-cortiacl (CSTC) circuits participate in the TS behaviors, even though DBS points to some possible regions within the GPi and the CM complex of the thalamus.

Much is still unknown about voluntary cursing. And much more is enigmatic about involuntary cursing. The behavior is not dichotomic; it is rather spectral with

varying levels of volition. When there is more control, there are also more ways to disguise or suppress the forbidden language. But it seems that the greatest amelioration is achieved when the community participates in the cognitive processes of the phenomena and recognizing its involuntary nature.

INTERVENTIONS

Sensori-motor processes are the physical sides of, or, as I prefer to say, form the anatomical substrata of, mental states. It is with these substrata only that we, in our character as physicians and physiologists, are directly concerned.

J. Hughlings Jackson in "On the anatomical and physiological localization of movements in the brain: Preface" The substance originally appeared in papers on epilepsy published in the *Medical Press and Circular* 1873-5.

It is vain to begin careful analysis of this complex problem [of epilepsy] without aid from psychology.

J. Hughlings Jackson in "Opthalmology and nervous diseases" *Transactions Opthamological Society,* 1886.

CHAPTER CONTENTS

IN THIS CHAPTER I address the most important aspect of Tourette syndrome (TS) – interventions. Currently there is no cure for TS. At best, interventions ameliorate the condition with minimal adverse side effects and provide coping tools. Hughlings Jackson teaches us that the physician, who treats psychiatric disorders with the goal of curing the patient, should be concerned with the sensorimotor system, and at the same time seek aid from psychology. I discuss therapies that collectively address both. Earlier I discussed the current interventions and their efficacy based on the literature. Here I focus on the interventions administered to the participants in my study, and the contribution of the patient to his or her own therapy. I start with the diagnosis.

DIAGNOSIS

Before interventions there is the milestone of the diagnosis. For many, naming the disorder has brought relief from the fear of being crazy and hope for help. Lionel, who was diagnosed when he was 48, believed until then that he "was the only person in the world like that" (S. R. Finkelstein et al., 2007-11-07). Ted, who was diagnosed at the age of 12, three years after the onset of his disorder, was relieved to be diagnosed because, "I knew I was not crazy" (S. R. Finkelstein et al., 2007-08-06). Kyle, prior to being diagnosed in grammar school, felt that "it was kind of scary 'cause nobody knew what it was," and the diagnosis "made it kind of easier [in school] because I didn't get into trouble… anymore" (Finkelstein et al., 2007-09-21). At the age of 17, Stuart was glad to be diagnosed. He "like[s] having names for things. … because you can come with a treatment plan" (Finkelstein et al., 2007-10-26). For Chuck, who was diagnosed at 6th grade, two years after the onset of the disorder, the diagnosis was "the best thing ever. . It

finally had a name… I was doing all those weird things and I'd get in trouble for it, yelled at… When I found that [it] had a name I went 'oh! That's what it is.' So that helped me out a lot" (S. R. Finkelstein et al., 2007-08-24). Danielle was diagnosed when she was 15, about five years after her onset. "It was nice to finally know what was wrong with me" (Finkelstein et al., 2007-08-17).

The responses of parents have been more varied. For Steven's mother the diagnosis of her 7-year-old son was "like a death sentence" (Finkelstein et al., 2007-09-28). Stuart's mother, even today after diagnosis and years of treatments, "doesn't believe Tourette syndrome exists" (Finkelstein et al., 2007-10-26). It took Chuck's mother, who felt ashamed and guilty, about a year before she could openly speak about her son having Tourette syndrome (Finkelstein et al., 2007-08-24). Neil's mother was happier with her original explanation of her son's behavior due to allergy (Finkelstein et al., 2007-08-22). But for Dylan's parents, the diagnosis open a pathway to dealing with the disorder. (Finkelstein et al., 2007-08-01b).

To many diagnosis brings hope for help. But it needs to "make sense," (Seligman, 1992:41) and contribute to the formation of a coherent autobiographical narrative. In *Rethinking Psychiatry*, the psychiatrist Arthur Kleinman claims that "a psychiatric diagnosis is an *interpretation* of a person's experience" (1988:7, italics in the original), and that this interpretation takes place within a culture, constructed with the cultural concepts of health and illness. The diagnosis needs to simultaneously fit into cultural explanations and resonate with the subjective experience of the afflicted. While today many TS patients would reject an explanation of being invaded by an evil spirit that needs to be exorcised, even if they sometimes use such a metaphor to describe their

condition, they accept biological explanations as coherent and sensible. Some are even open to socio-psychological explanations. This is how today in the West psychiatric disorders are understood -- as biopsychosocial phenomena (Ghaemi, 2007).

In *Twitch and Shout*, Lowell Handler dedicates a whole chapter to the journey of his diagnosis. His brother Evan insisted "You've got to find out what's wrong with you" (Handler, 2004:30). His mother asked the doctor whether Lowell's behavior resulted from allergies "I don't think she actually believed this but was searching… for an explanation of my bizarre behavior" (31). The family has been searching for a diagnosis and cure for Lowell's condition for 16 years. They were sent repeatedly to family therapy with no results. The search for a diagnosis reached its end when they visited the neurologist Oliver Sacks, who named Lowell's condition -- Tourette syndrome. "I was both stunned and relieved to find out that my condition had a name" (33). For Lowell the biological and familial interpretations of the disorder *made sense*. He even started to wonder whether his father had some signs of TS. Even though the biological and hereditary interpretations informed Lowell and his family that he would have TS for the rest of his life, "[o]nce the diagnosis of Tourette was made, everything began to fall into place" (38). It relieved Lowell's feeling of being crazy and alone. "I learned that like me hundreds of thousands of people in this country have Tourette" (33).

As fit with the zeitgeist of his time, the patient "O". of Meige and Feindel explained his tics in psychological terms: "We who tic are consumed with a desire for the forbidden fruit" (cited in Meige & Feindel, 1907/1990:12). And, accordingly, O. attributed the amelioration of his tics to his psychological treatment: "I am conscious of very material gain. I do not tic so often or with such force. I know how to keep still.

Above all, I have learned the secret of inhibition"(22). The patient's understanding of his or her disorder is informed by the culture and the observations and understanding of the medical practitioner. But at the end it is the patient who is privileged to accept or reject a diagnosis. While this recognition is in agreement with the American respect to the individual and with its implications to the doctor-patient relationships, this is not a universal attitude. Serge Lebovici, the influential child psychoanalyst of the second half of 20th century France, sarcastically noted that the Americans summarize scientific research and share it with their patients, the laypeople (Kushner, 1999). Indeed, American culture assumes that laypeople count, especially when they are the afflicted.

Who Diagnoses

The public and medical awareness of TS is relatively new, but in the last two decades it has been widely publicized in the media. This enabled some of the adults in my study, who had lived with undiagnosed TS many years, to take an active role in their own diagnosis. When he was 48, the now 65-year-old Lionel read a magazine article describing TS. "This story was me," he said. The phone number in the article took him to the Tourette Syndrome Association (TSA) that referred him to a local physician who confirmed his self-diagnosis (Finkelstein et al., 2007-11-07). Neil, himself a psychiatrist, diagnosed himself when he was a medical student: "I think I made the diagnosis myself" (S. R. Finkelstein et al., 2007-08-22a). Nick was diagnosed in his late 20's, after his mother, with whom he still lived at the time, had read an article about Tourette syndrome in the newspaper. He and his mother suspected that he had TS, and it was confirmed in the local hospital (Finkelstein et al., 2007-12-07).

For the pediatric population, the diagnosis of TS typically takes place when the parents take their child to a physician. But the forcefulness of the parent involvement varies. Dylan's mother "noticed things that were going on that she couldn't explain, [and] she got involved. And once she gets involved you better get out of her way," says Dylan's father. "It was because of her that we found out what the problem was early on." (S. R. Finkelstein et al., 2007-08-01b). Stuart's diagnosis at the age of 17, a few years after the onset, followed the initiative of his school's counselor. She sent him to a psychiatrist: "I told her [the counselor] that sometimes… when I am sitting in a group and there is a person that I do not like, I start thinking 'bitch'… That's what made her suspect. She sent me to [a] psychiatrist" (Finkelstein et al., 2007-10-26).

WHY TREAT?

The question "Why treat?" is not as foolish as it sounds first. In fact it is another way of asking what the penalties of TS are and is it worthwhile treating them. In the chapter "Symbolic tics" I discussed the social penalties that follow the TS breaking of taboos. But social penalties are also the share of those who do not have symbolic tics. Phonic tics are disruptive and unwelcomed in public places. Not only are they unpleasant for the people at the neighboring table in the restaurant, they also disrupt a concert, a movie, or a public talk, as was shared by many participants, including Chuck and Claire (Finkelstein et al., 2007-08-24; Finkelstein et al., 2007-12-21). Self mutilation might cost the person an eye as for Henry (Finkelstein et al., 2007-07-23). And motor tics, when frequent and intense, might also cause physical injuries. For example, frequent bending can injure the lower back, as for Daniel (Juncos, DeLong, & Finkelstein, June 2008); clapping the knees against each others can injure them as for Elliot (Finkelstein et al.,

2007-08-13); and violence against objects has its own consequences as was shared by Claire, Dylan's father, and Ted's mother (Finkelstein et al., 2007-08-01b; Finkelstein et al., 2007-08-06; Finkelstein et al., 2007-12-21).

Adults with severe TS can be fired from work for insulting a customer (e.g., Ted) or the boss (Nick, Finkelstein et al., 2007-12-07). Motor tics reduce the motor control, and Louis, who is a nurse, had to switch to an administrative job because he used to fling the syringe out of his hand due to his wrist tic (Finkelstein et al., 2008-05-30). Danielle is not allowed to drive since she has eyeball tics that might be dangerous (Finkelstein et al., 2007-08-17). Henry is unemployed and is on disability. Donna lives on food stamps and cannot find an employment (Finkelstein et al., 2008-02-08).

Surely, there is a good reason to treat. But treatment, as was discussed earlier, is an individual road of iterative trial-and-error, in which the afflicted is simultaneously treated and while serving as an experimental subject for the efficacy of the treatment. The question then is about the balance between the amelioration, the cost of achieving it, and the adverse side effects that often come with it. Unfortunately the answer is complex. Therefore the question should be asked. It needs to be asked for and by each individual with TS, since each has an idiosyncratic phenotype and responds to treatment in a unique and unpredictable way. Borrowing James Leckman's metaphor, those who suffer from TS are like snowflakes, not two are alike. TS has no cure, but people can be helped, pharmacologically, surgically, and behaviorally; but often only temporarily.

PHARMACOLOGICAL AND SURGICAL THERAPY

The most widespread therapies are the pharmacological. They can help, but they often cause adverse side effects, and their long-term effect is unknown. When there is

amelioration it is often not clear whether the medications are to be credited or whether this is a waning phase in the trajectory of the disorder.

Often the road to the right mix of medications can be torturous. Ted's first medication was haloperidol. Many were helped with it (Shapiro, Shapiro, Bruun, & Sweet, 1988). But earlier I quoted Ted's experience with haloperidol. He was in a straight jacket in the hospital. He then scarred his face, damaged his own eyes, pulled his teeth out with his own hands, and had to be kept away from suicide. When the haloperidol was stopped, his condition improved almost immediately. But it took half a year before this happened. The pimozide that he takes now helps. But, like in many other cases, it is not enough. His depression is treated with Parnate, and this requires a rigid diet. And while this soup of medications helps, "the only thing that takes the tics away" is a daily dose "of 3 or 4 inch[es]" of marijuana (Finkelstein et al., 2007-08-06).

Danielle had to take twenty three (23) different medications daily. She stopped them all abruptly and "felt so much better after that…tremendously better." For four months she had no tics, and then they returned (Finkelstein et al., 2007-08-17). Even though her epilepsy-like tics are diagnosed as non-epileptic, the anti-epileptic anti-convulsant Topamax helps her. It does have cognitive side effects and her poor memory and constant fatigue might be such, but no one knows.

Chuck takes Tenex. He thinks that it helps but is not sure whether he owes the improvement in his situation to the medication or to having less stress in his life. "Is it better now while I'm on it [on Tenex]? Maybe. Just now, I'm fine. So if it's the medication, I give the medication credit. I don't know. But you see with the life issues and jobs, you see that all plays a role." Chuck still has his frequent vocal tics… [but it

used] to be louder, I had it more frequently, the pitch was higher" (Finkelstein et al., 2007-08-24). Lionel has given up. The antiepileptic Keppra was prescribed to him, but it made him sleepy and brought no relief with his tics. Would he do better with other medications? Being treated by another physician? He distrusts neurologists and psychiatrists so he has decided to cope without any medical help (Finkelstein et al., 2007-11-07).

A common side effect of Anafranil, which is prescribed for obsessive-compulsive disorder (OCD), is erectal dysfunction. Steven and Dylan had to be taken off the medication. Nick is pleased with its effects. Elliot takes Orap, with possible side effects of akathisia (motor restlessness). It is hard to be sure. His physician is reluctant to switch to another atypical antipsychotic since other antipsychotics are associated with aggravation of diabetes and hyperlipidemia, with which Elliot is diagnosed (Finkelstein et al., 2007-08-13).

Obviously, the pharmacological therapies are a mixed blessing. And whether to take them or not depends on the severity of the phenotype, the individual response to specific medications, and the severity of the adverse side effects. The question whether to medicate is not a yes-no question. And even for the same individual it is not asked and answered once and for all. It is an ongoing question with possibly different answers.

Neurosurgery

Claire, whose screaming, touching herself, and falling are debilitating, gets no relief from her medications. She feels that neurosurgery is her only hope. She would hopefully be included soon in a study of deep brain stimulation (DBS) for TS. The procedure is still highly debated and not yet approved as a therapeutic procedure by the

FDA. It is recommended only for people with severe phenotype who resist all medications and are older than 25 (Mink et al., 2006). The only access for those who, like Claire, cannot pay for this expensive procedure is within a study.

It may help her. DBS sometimes looks like a miracle. Dylan had elaborate tics, violent tics, self mutilation, and forbidden language. The intensity was high and his tics were very frequent. He resisted all medications. Nothing helped. "You say it took courage," Dylan's father says to me about the decision to operate. "I don't know what courage, because once we knew what it was and what it could do we were eager to do it and there was some apprehension of course because it's a brain surgery and, you know, anything can go wrong but it really [got] to the point where we couldn't imagine it getting any worse. Really there is no way that I could imagine it being any worse." Dylan is still on medications, still has tics, and still has eruptive cursing But the amelioration is great and the quality of his life has been greatly improved. He can now sit in the passenger seat next to the driver, he has a girlfriend, he can go to the mall, and he holds a job (Finkelstein et al., 2007-08-01a; Finkelstein et al., 2007-08-01b).

Daniel's life has changed dramatically after his DBS. From bending and touching the floor almost once every minute, he has acquired an upright gait with no bending. His medications have been reduced, and his quality of life has been greatly improved. The amelioration is remarkable. But he is not cured (Finkelstein et al., 2008-06-04).

Self medication

The adults in my study often rely on themselves for treatment. This can take many forms. Activism to legalize a medication that might help but has not been approved by

the FDA, taking illegal drugs, refusing any medications, or administrating drugs impulsively.

An important case of activism is that for pimozide, as told by Adam Seligman (Seligman, 1992) and Lowell Handler (Handler, 2004). In 1979 Seligman started to take the blocker of the dopaminergic receptor D2, pimozide. He found it to be highly effective and to have fewer side effects than the less-selective haloperidol. Alas, it was not available in the US, even though it had been available in Europe and Canada for years. Seligman was only 17 at the time. His mother decided to smuggle it for him from Canada. Handler requested to take pimozide in a clinical trial conducted by Shapiro. With the encouragement of Abbey Meyer of the Tourette Syndrome Association Seligman and Handler agreed to testify before Congress, and they succeeded. Pimozide was one of the first drugs to be approved under the Orphan Drug Act[93] that later became a law. Thus Seligman and Handler initiated, with some help, the use of a drug and worked to make it legally available.

Ted, as we heard above, supplements his pharmacological medication with the only drug that helps him, marijuana (Finkelstein et al., 2007-08-06). He is the only one on my study who admitted to using marijuana, but not the only TS patient who reports amelioration of tics after taking marijuana (Handler, 2004).

Another form of "self-medication" is refusal to take any medication. Neil (Finkelstein et al., 2007-08-22) and Lionel (Finkelstein et al., 2007-11-07) do not take any medications and do not see any physician. Neil considers himself a mild phenotype and is aware of possible side effects, which as a neurologist he often sees in others.

[93] The Orphan Drug Act is for a disease that affects less than 200,000 Americans, and therefore is not economically attractive enough to be "adopted" by any drug company. The Act created some tax incentives to encourage drug companies to develop drugs for rare diseases that affect small populations.

Lionel has found Keppra to be ineffective, and distrusts neurologists and psychiatrists in general.

While the decisions of Ted, Neil, and Lionel to medicate or not to medicate are a way to assert their agency, the sporadic undisciplined self-medications of Donna is not. Her dominant experience is of helplessness and giving up. While she is dreaming of a better future, she resists any attempt to improve her situation. With no job, living off food stamps, being isolated, and bitter, Donna starts and stops her medications at will, and sporadically and arbitrarily admits herself to the hospital (Finkelstein et al., 2008-02-08).

Such minimal agency is at its extreme with drug dependency. Dylan, who is grateful to his DBS, whose improvement after his DBS seems miraculous, has friends who love him and accept him. Alas, some of them consume drugs, including cocaine. And Dylan cannot resist. In addition to his medication, he takes cocaine. He underwent rehabilitation since his neurologist conditioned the continuation of his treatment on getting off the cocaine. But it did not last and he is currently on the drug again. His father wishes he had different friends (Finkelstein et al., 2007-08-01b)…

BEHAVIORAL THERAPY

No one in my study is "officially" involved in what is called "behavioral therapy." In this section, I will consider non-pharmacological and non-surgical as behavioral. Most of these behavioral strategies were discovered and developed by the participants themselves. For Lionel, his "obsessive and intrusive thoughts" are the greatest challenge of his TS; greater than the tics and his forbidden language, which are just "the tip of the iceberg." He noticed that direct fighting does not work "the stronger I fight the stronger it gets." He changed his strategy. He pretends he is in a horror movie that he has to watch.

His tension decreases and the experience is more bearable, less tormenting. Lionel extends his personal success to support his friend. He guides her away from fighting her thoughts and feelings into observing them as though she is in a horror movie. This helps her through the experience and makes it more tolerable (Finkelstein et al., 2007-11-07).

Lionel's strategy is of special interest since it includes some elements known to meditators. The modification of meditation for the specific purpose of healing and stress reduction has been labeled *mindfulness* to distinguish it from religious connotations and adapt it to a Western context. In mindfulness there is, in addition to stress reduction, also a shift in the relations between the individual and his or her own mind. The thoughts and feelings are observed as in a movie, to use Lionel's words. The valid observation of Lionel that the more he fights the worse the situation gets, describes a battle between the individual and his thoughts and feelings. Such a battle engages and fuels the thoughts and feelings, and thus the darkness thickens. By contrast, a non-combative observation reduces the power of the uninvited thoughts and feelings over the individual. It is a form of disengagement in which no one seeks control. The individual resigns from controlling his or her thoughts and feelings, and the thoughts and feelings have also lost their control over the individual; thus the vicious circle breaks and a relief is experienced (Personal experience. Kabat-Zinn, 2006).

Lionel keeps asserting his agency in every aspect of his TS. He is the one who diagnosed his disorder, he is the one who decided not to take medications, and he is the one who has found a strategy to deal with what for him is the greatest challenge of the disorder. Indeed, in behavioral therapy, the role of agency is crucial. In drug therapy the choice of medications, their doses, and how to administer them are decided by the

physician, and the patient has to follow instructions; in DBS the patient has to agree to the intervention but then it's all in the hands of the medical team. But behavioral modification is completely up to the individual and is not accomplished by a one-time decision. It requires an ongoing application of strategies, whether taught or suggested by others, or discovered or invented by the individual; a constant assertion of agency.

Agency and OCD

The question of agency is greatly complicated when OCD is part of the disorder. How does a person respond to a disorder, whose essence is a weakness of voluntary behavior? How to assert agency despite the disorder? There is a whole spectrum between success and failure, and it is not easy to predict.

ID #	Patient	obsession		Compulsions	
		Resistance	Severity	Resistance	Severity
4	KGA	3	17	2	15
5	DQA	3	8	3	7
7	SZA	3	14	2	9
8	EEA	2	3	4	8
10	NFA		0	2	11
11	DLA	3	9	4	15
12	NYA		1	2	9
14	DGA	4	19	4	15
15	LLA	3	7	2	7
16	DGB	3	4		1

Table 9.1: For 10 out of the 16 participants, Dr. Juncos filled in the Y-BOCS observational questionnaire: (1) stands for "Tries to resist all the time"; (2) stands for "Makes some efforts to resist"; (3) stands for "Yields to all"; and (4) stands for "Completely and willingly yields to all."

All the participants in my study suffer from TS *and* OCD. Ten were observed by their treating physician and he rated their resistance to obsessions and compulsions. Table 9.1 summarizes his observations. They reveal that the severity of the obsessions and

compulsions do not necessarily predict the ability to resist them. For example, Elliot (#8) whose compulsion severity rated 8, which is much less that that of others with 15, has a ranking of 4, "Completely and willingly yielding to all," in his resistance to the compulsions. In other words, Elliot's greatest challenge with his OC is neither the severity of his obsessions, nor of his compulsions; they are quite mild. But it is his low *resistance* to his compulsions. On the other hand, Kyle (#4), with the high severity 17 for obsessions and the high severity 15 for compulsions can resist them to some extent. While he is rated 3, "Yields to all" for obsessions, he is rated 2, "Makes some efforts to resist" for compulsions. The balance between resisting compulsions and obsessions varies among individuals.

Strategies: Suppression, disguise, and substitution

In my earlier chapter "Symbolic tics," I discussed how some participants have developed strategies to suppress, disguise, or substitute their forbidden language. I briefly repeat some here: Kyle succeeds in suppressing some of his tics until he comes home. So does Louis. Ted listened, for much of his interview, with a pencil between his lips to keep him from undesirable utterances. Daniel sat on his hands throughout his interview to avoid forbidden gestures. To hide his ticcing feet, he sat in lotus position. When standing, he put his hands in his pockets.

In a study of 135 (ages 8-71) TS patients, the YGTSS questionnaire was used to assess tic severity of the participants. In addition the participants were asked to self report and describe their premonitory urges. 93% reported such urges, and 92% reported that the tics following the urge are fully or partially voluntary (Leckman et al., 1999). Often the tics can be suppressed (e.g., in a job interview, in dating a new person). But typically the

suppression is limited to short periods of time and after a while, the tics are irresistible any longer, and come in intense bouts, with a great force. Lionel says, "You can suppress them [the tics] so long, and they come back with a vengeance" (S. R. Finkelstein et al., 2007-11-07). Sometimes Neil succeeds in suppressing his tics despite the urge. But then the tics come intensely later. "For example, I am not really suppressing now but I am a little bit and… when I leave here I probably will have more." He tries to explain the phenomenon: "It's not that it's [the suppression is] really conscious, I am concentrating more here. Probably after the concentrating, talking, I'll be ticcing more often" (S. R. Finkelstein et al., 2007-08-22a). Henry describes the fight to suppress his urge in real time, "I'm putting most of my energies into not yielding to the urge to tic. And I'm using the rest of my brain to try and get to work [what I am] saying to you. Three things are going on at once. I can't release. The urge is there right now. I'm thinking about the urge more than about what I'm trying to tell you" (S. R. Finkelstein et al., 2007-07-23). And Elliot does not "have the explosion of tics that some people have" after suppressing a tic. But his ability to suppress is limited and "there is a moment that you *have* to go" (S. R. Finkelstein et al., 2007-08-13a).

When the consequences of succumbing to the urge are extreme, people make special efforts. Henry suppresses his urge to poke his only eye with a knife (Finkelstein et al., 2007-07-23). Danielle suppresses her desire to get satisfaction from using razor blades (Finkelstein et al., 2007-08-17). But there are drawbacks, similar to Dylan's pattern with the cocaine. The fear of such a drawback is expressed by Steven about his substituting grunt: "But that can all [the inappropriate speech], can all come back" (Finkelstein et al., 2007-09-28).

As with symbolic tics, some of the participants spoke about ticcing most in the evening, at home, away from the public eyes. Louis says, "when I get home in the evening, when I walk through the door, it's like (shapes his hands as though holding a ball and exhales loudly)--- Because I'm trying to suppress it the whole day, it's like it's going to come out. And I feel like I *have* to do it." But from our conversation it is not obvious whether the pattern of more ticcing at the evening is due to cumulative energy, or whether being busy during the day keeps him busy with work and therefore ticcing less (S. R. Finkelstein et al., 2008-05-30). It's also possible that for some the tiredness at the end of the day diminishes their ability to voluntarily resist the urge: "The worst time of the day for me," said Neil, "is late afternoon after about 5" (S. R. Finkelstein et al., 2007-08-22a).

Kyle and Lionel disguise their forbidden speech by muttering it under their breath. Dylan disguises the gesture with his middle finger by extending the motion to remove hair from his forehead. Ted tries to substitute inappropriate utterance with an accepted one; "Nigger" becomes "Nick." Steven has substituted all his forbidden speech with a grunt. And Neil substitutes humor. These three strategies are not restricted to symbolic tics. They are applied to other tics as well.

The anthropologist Andrew Buckser, who interviewed adults with TS, describes tic substitution consciously applied by some of his participants: "In a situation of close social contact, where satisfying the urge for a facial or neck tic would be very noticeable, almost all of my informants said that they would occasionally induce a tic in the leg or foot instead" (Buckser, 2008:176). Henry spoke about replacing highly-penalizing tics with less-penalizing ones. He tries to replace his self mutilation tics: "I'm trying to switch

over from the physical tics to the verbal tics because my body just about had it" (S. R. Finkelstein et al., 2007-07-23).

Replacing old tics with new ones

Like suppression, disguise and substitution have limitations. Donna's "Fire!" often replaces her "fuck you all." However, it has acquired a life of its own. And while it is not insulting, "Fire!" is alarming. It has caused Donna many troubles, like losing a job and being threatened by her landlord to lose her apartment (S. R. Finkelstein et al., 2008-02-08).

While by exerting their will, many TS patients succeed in disguising some tics with other behaviors, or replacing injurious tics with less severe ones, often the very new behaviors become new tics. One of the participants in Buckser's study told him "of a time in his twenties when he had tried inducing abdominal tics, involving the clenching of his diaphragm and chest muscles, to relieve facial tics during conversations. The induced tics soon became chronic, leading to back spasms and breathing difficulties that lasted for months. Another man clenched his shoulders to divert energy from a noticeable facial tic; the practice produced muscle tension in his shoulders that progressed to more severe tics, then to intense headaches, and eventually to back pain that placed him on disability" (Buckser, 2008:176).

Silvia Sottofattori describes two patients whom she observed as part of her PhD research (Sottofattori, 2007): One of them, the adolescent OP, was greatly embarrassed by a tic that involved his forearm. To disguise it as a voluntary and meaningful gesture he added to his tic a pointing index finger and created a "fake deictic gesture." Now it appeared as a meaningful movement, not as an uncontrollable tic. Alas, it has assumed a

life of its own, and the elaborate combination of raising the forearm and pointing with the index finger has become a tic. Sottofattori suggests that the motion has undergone a process of automatization.

It is possible and even likely that this automatization relies on the same mechanism that is involved in the learning of many motor behaviors. And it might be that this mechanism contributes to the trajectory of the disorder and to the change in tic repertory often seen in the course of the disorder of many individuals. The typical progression of TS is from motor tics alone to additional vocals tics. But besides adding a modality, the tics themselves keep changing. Tics can move from one part of the body to another. And some tics can disappear and make room for new ones. This distinguishes them from most other hyperkinetic movement disorders (Fahn, 2005). Steven's first tics included neck-cracking, which is "the only tic that remains. All the other tics changed over time" (S. R. Finkelstein et al., 2007-09-28). Lionel's first tics at the age of 5 were all motor, including seizures. The seizures almost completely disappeared at the age of 15. But then forbidden speech kicked in (S. R. Finkelstein et al., 2007-11-07). At some point in their relations, Claire was violent against her husband. This has stopped. New tics -- falling and screaming – have taken over (S. R. Finkelstein et al., 2007-12-21). Louis's tics have changed over the year. His eye-blinking greatly diminished when he went to college. His trichotillomania has disappeared too. But he does have one persistent tic– a wrist jerk (S. R. Finkelstein et al., 2008-05-30).

It is hard to separate the variations in the nature of tics from the variation of their intensity and frequency and from their waning and waxing pattern. There are three forms of variability: (i) a volitional replacement of a tic with another behavior, which then turns

into a tic; (ii) a spontaneous tic disappearance and its replacement by other tics with no patient's interference; and (iii) a changing pattern of tic frequency and intensity. Similar variability is also characteristic to the urges. Some urgeless tics acquire an urge. For others the nature of the urge changes "The thing that was interesting about the swearing is that at first it felt like a physical tic; involuntary, uncontrolled, rapid. But soon it built up into a stronger feeling of internal tension" (Seligman, 1992:36).

Involuntary vs. automatic

The French psychologist Pierre Janet (1859-1947) considered all tics to evolve from voluntary actions, which a subconscious obsessive-compulsive conflict turns into a tic (H. I. Kushner, 1999). It is no longer sufficient to explain tics in psychological terms alone. But some points in Janet's discussion of tics deserve attention; especially his linking of the process of automaticity to OC.

Automaticity is an important aspect of learning. And even though in everyday speech we often interchange "automatic" with "involuntary," they are not the same, yet not mutually exclusive. *Tics are involuntary*, at least to some extent, and volition has only limited control over them. *Biking is automatic*. The activity is volitional and it can start or stop at our will. However, its elements do not require conscious attention. As I choose to bike I do not need to consider how to pedal. Automatic skills are quick, effortless, without conscious control, and once acquired are hard to change (Logan, 1988). Walking, swimming, playing tennis, and even writing and playing the piano are examples of automatic activities. They are learned behaviors that in the beginning are under conscious control, quite slow and often awkward. As they become automated they get quick and effortless. This is thanks to the fact that they do not require conscious

attention anymore. If after their automatization, an individual attempts to perform them consciously, the performance is inferior to the automatic one. And changing them requires great conscious attention and is very difficult.

The automaticity process relies on repeated rehearsals. If, as proposed by Janet, tics start as voluntary behaviors and later transform, OC might play an important role since it provides the intensive rehearsal that is important in transforming a learned behavior into an automatic one. The OC responds to common triggers. And it is highly probable that OC is not merely a comorbidity of TS, but an integral and necessary aspect of it. Whether OC stands for obsessive-compulsive disorder (OCD), obsessive-compulsive symptoms (OCS), obsessive-compulsive behavior (OCB), or obsessive-compulsive spectrum disorders (OCDS) is an important and unresolved question and requires greater clarification within the scientific and medical communities.

The human environment

Earlier on Meige and Feindel emphasized the role of the family in the therapy of the patient; thus their reluctance to send a patient to an asylum that separates the patient from her or his family (1907/1990). Ted speaks of his wife who “provides structure and a lot of strength.” He also speaks of his parents who have combined support with expectations (Finkelstein et al., 2007-11-07). It is difficult to determine whether his ability to have a family and hold a job results from family support, but it is a likely conjecture. Chuck, who is now a successful teacher and married, is grateful to his parents. “My Mom, my Dad, my brother -- we experienced it, it’s life. I mean, we weren’t sheltered. It wasn’t like ‘stay at home.’ It was more like, ‘What d’you want to get involved with? You can do sports, you can do soccer, do baseball, basketball, you’re

going to be in camp, you're going to get out there'" (Finkelstein et al., 2007-08-24). As for Ted, Chuck too experienced a combination of expectations as he were like his brother, along with opportunities, love and support.

Maybe not only what the individual *gets* from a close community matters but also what the individual *gives* matters. Chuck has developed a sensitive and rich philosophy of compassionate and individualized teaching. Ted believes that the way he copes with his difficulties makes him a role model for the children with disabilities with whom he works. Steven, who also works with children with disabilities, loves what he is doing and has befriended some of the older students in his program (Finkelstein et al., 2007-09-28). After a long personal journey, Lowell Handler has found home in his job at Cobblestone Road, a non-profit organization that provides housing, food, job training, and social work services to homeless men and women. He reflects, "All we have in this world is each other. And we are all different yet bound by our humanity"(Handler, 2004:203).

This longing to be bound with others in common humanity makes the social environment important, a source of support, and a source of pain. Buckser discusses three strategies that intend to remove tics from the social arena: displacement, misattribution, and contextualization (2008). His displacement and misattribution are like my suppression, disguise, and substitution. But his *contextualization* is a strategy through which the person with TS "invites" the others for tolerance. Earlier I discussed the opportunity given in some schools for a child with TS to introduce the disorder to his or her classmates; thus remove a stigma of the other. The common knowledge today that TS has biological origins removes the stigma often attached to psychological disorders. And finally, remembering that the TS behavior does not result from inappropriate intentions

would hopefully change the perception of the individual who suffers from TS and of the disorder in general.

In the chapter “Symbolic tics” I discussed the challenges faced by the afflicted to change the TS and by the people in the human environment of the afflicted to tolerate it. Buckser is more optimistic than I. His focus is on the relations between the TS sufferer and the social construction of the disorders. And his conclusion is that “TS sufferers are not passive victims of social stigma, ground between the gears of a misperceiving society and an oppressive physical symptomology; they are central agents in determining how their illness will be perceived and understood” (184). We studied very different populations. He believes that the current misconception of TS is because “the larger culture, associating the disease with florid symptoms like cursing—symptoms that, although not at all typical of TS, are the ones most resistant to these kinds of management [of displacement, misattribution, and contextualization].” (167).

I disagree. I think that the research and medical communities have accepted mild cases and diagnosed them with TS, while those with the florid phenotypes are often marginalized and get little help. Buckser’s study and mine are examples, maybe important ones, of how the definition of the disorder matters; how the clinical definition includes many mild cases; while, as folk intuition knows, there are some, or many, out there with florid phenotypes. They are marginalized and often cannot receive adequate help, and they desperately need it, in the form of research, clinical treatments, and social support.

PERSONAL REFLECTIONS

Many times during this study I asked myself, what would I do had my child been afflicted with TS? I realize that I can never be sure how I would behave in such a situation until I am in it. But from listening to the people in my study, patients as well as their parents, spouses, and friends, and drawing lessons from my own life experience I have developed some intuitions.

Even though the population of my study is of adults, the pediatric population is important. That's where it all starts. The first trap to avoid is to define a child with TS by the disorder. This rule has been captured by the title of the DVD "I have Tourette's but Tourette's does not have me" (Tourette-Syndrome-Association). Like every other child, a child afflicted with Tourette's is to be raised to the fullest of his or her potential towards having meaningful and fulfilling life. But, of course, the job here is harder; often much harder. The purpose of life and its meaning is the same for the individual afflicted with TS as for any of other child. Therefore the purpose of intervention should be linked to the purpose of raising a whole human being. Cure is not there. Not yet. Coping might be a more realistic and more productive goal. All people have to learn how to cope. All face challenges. However, the challenge of TS is great and calls for a greater response.

Medications

Medications are widely used, even for mild cases of TS. Many have adverse side effects; should they be administered or not? This is related to the question of how we distinguish between transitory tics, which afflict about 25% of the pediatric population, and those that continue into adulthood. This question is rarely asked today. The most common assumption today is that a child diagnosed with TS has to be medicated. But

maybe we should pay less respect to the name of the disorder. What we call Tourette's today is not what was identified by Gilles de la Tourette. Moreover, what differentiates TS from other tic disorders is the presence of vocal tics in addition to motor. However, it is possible that most vocal tics are phonic, and just another form of motor tics. This would change the diagnosis of many from TS to another tic disorder. And we know that most children diagnosed with TS recover before adulthood.

The question we need to ask and that I have not heard throughout my study is: **what are the predictors for a tic disorder that will become a lifelong-disorder rather than a transitory one**. These are the individuals who most require care. The rest may recover spontaneously, without any intervention. If their disorder is, as we discussed earlier, a result of disharmony among developing subsystems, then hopefully when all their systems mature they synchronize and the person is cured. No intervention, no side effects. The phenotypes of those that stay with the disorder throughout their adulthood are typically severe and need special attention. Maybe the place to start looking for a predictor is in the TS-OCD comorbidity. Throughout this dissertation I returned to various situations in which the OCD seemed to be the driver of the tics and the force behind their persistence.

A child with a transitory syndrome, whatever its name is, should acquire coping skills. Administering drugs to him or her should be considered carefully, taking side effects and the possibility of still unknown long-term effects into account. At times medications do a lot of good. But I think that we tend to overuse them. It is difficult for parents and physicians to admit how little we know and how helpless we are. Prescribing drugs is a way acting, fighting, and keeping hope. Therefore research and development of

drugs should continue. Marijuana might prove to be potent and with not so many adverse side-effects. It's time to invest more in studying this plant, even if the ease of growing it and its wide availability would make it hard to patent.

Behavioral therapy

As a parent, I would try other avenues before resorting to our current pharmacology. What is to replace it? "It takes a village to raise a child." And indeed the support of school, family, and friends is indispensible in allowing a child to simply grow, tics or no tics, and concentrate on the experience of life. Much can be learned from those in my study who cope. Ted's parents treated him like they treated his healthy brother. Same expectations, same love. Demanding yet always available, they escorted him through many difficult and scary moments. But he has grown to have his own family, have a job where he is liked and appreciated, and live a full life.

For most with TS, the greatest challenge is the social. Last summer I visited a TS weekend camp in Georgia. Children and their families stayed in cabins, played, participated in sports activities, spent time together, and enjoyed gentle southern landscape, with a lake, open meadows, woods, and many mosquitoes. Chuck, who never let the TS be in his way and took twenty four job rejections before being hired as a teacher, presided over the family Olympiad. It was normal and happy. There were tics, many. But they did not matter much. This need for people like one's own is important. Louis expressed his need for people with whom he could share problems and solutions. The support group is a familiar idea to Americans. Support groups get organized for weaning from alcohol, reading books, and hiking. Such groups can be powerful. And even though they are not there with the person all the time, they provide moments, hours,

sometimes days of relief and joy. There are local chapters of Tourette syndrome that provide a sense of community. Active mothers deserve much credit for this. And the Tourette Syndrome Association is supportive of such initiatives as well. Adults, however, are left to fend for themselves. They need help too. While many adults with TS would take advantage of group meetings and activities, only few, if any, would initiate the creation of such a group. Much of behavioral therapy can be provided in groups. And maybe this is the practical way to combine the teaching and practice of coping techniques with the needed community support.

I see hope in behavioral approaches. However, I suspect that the current Cognitive behavioral therapy (CBT) is too direct. Tics have to be tricked. Many CBT methods give prominence to the tics and keep them at the center of attention, drawing from limited cognitive and especially emotional resources. Emory is uniquely positioned now to study the effect of meditation, or mindfulness training, on TS behavior. To the best of my knowledge, this form of behavioral therapy has not been tried with TS anywhere. Meditation is indirect in the sense that it does not draw attention to the tics. Instead it creates a state of mind, a new cognitive space that may be less liable to react to tic triggers. In addition, by learning how to observe oneself, the person becomes less under the control of the TS urges. Instead of fighting them or succumbing to them, one observes them. Lionel's response to his dark thoughts, watching them as though sitting in a horror movie, is such a success story. He cannot help having his dark thoughts but he can help how he experiences them. Moreover, he extends his insight and technique to help his friend. When she gets tormented by her dark thoughts he keeps her company watching her horror movie with her. Together they cope (Finkelstein et al., 2007-11-07).

Lionel is not the only one whose coping strategy can be translated into successful behavioral therapy. Many in my study shared how when they are motivated and concentrating they tic less or do not tic at all. Maybe less enforced routine and imposed activities and more licenses for activities that the person loves and is motivated to perform can bring the person with the Tourette's more hours of satisfaction and self-fulfillment.

Oliver Sacks reported his patients as responding positively to rhythm. While the people in my study did not exclude this possibility, they subjectively emphasized instead their concentration and self- motivation. A possibility that has not been studied to the best of my knowledge is that the effect of music is not necessarily the rhythm but the fact that it is acoustic rather than visual. Designing programs that are richer in acoustic input and have as little visual input as possible might provide relief.

Where the efforts should go

Behavioral therapies can help some. They can provide islands of peace and groups can relieve some of the loneliness. Some will still need pharmacological help. Some will need neurosurgical help. Severe TS is a debilitating disorder, and it deserves much more attention from physicians, researchers, and granting agencies. Children who may outgrow their tics should not be rushed to be medicated. The efforts should go toward identifying and helping those who are doomed for life. In the next chapter I will propose future directions…

FUTURE DIRECTIONS

CHAPTER CONTENTS

IN THIS CHAPTER I conclude my study and suggest future directions.

Since the time of Gilles de la Tourette, the disorder named after him has undergone different constructions. Its very definition has changed to include mild phenotypes and to marginalize the severe ones. Many adults with TS have severe phenotypes and are jobless, socially isolated, on disability, and with no one to turn to. There are no reliable data on their prevalence in the population or statistics about the distribution of their phenotypes. In parallel to epidemiological studies to gather these data there are additional research directions that may lead to better understanding of Tourette syndrome (TS) and to better treatments.

With the exception of two out of the sixteen, the people in my study were recruited from a clinical population. They exhibit severe phenotypes, including the eruptive utterances of forbidden language, and self injurious behavior. They also suffer from obsessive-compulsive disorder (OCD) even though this was not a selection criterion for the study. Their self-reflections, the questionnaires that they filled, the medical observations, and study of their tics strongly suggest that OCD contributes to the severity of the disorder. Therefore, future studies should pay attention to the contribution of OCD and consider the *spectrum nature* of TS beyond its current DSM-IV definition.

As I pointed in the chapter "Interventions." finding predictors that identify tic disorders that are likely to become life-long rather than transitory is important: It will free children with transitory tic disorder from medications that might have long-term adverse effects, and it will encourage research focusing on those who are in real need of help. For this life-long population, I have identified a few directions of research that I believe and hope will be productive.

Temporal patterns of the tics

Rich data from the video-recorded interviews was gathered and analyzed. It was found that motor and vocal tics tend not to co-occur, and their co-occurrence is significantly less than the co-occurrence of speech and gesture. In addition, elaborate tics that are ritualistic series of simple tics seem to have the same intensity for all the simple tics in a specific instance of the ritual. These data are difficult to interpret and may need additional support from other studies that include data about temporal behavior, like the frequency of electrodes in deep brain stimulation (DBS); or fMRI studies that follow temporal activation patterns. They might lead to better understanding of the gating of CSTS circuits, which some consider as central in the underlying mechanism of TS (Jankovic & Mejia, 2006). More detailed studies of temporal behavior of tics are in order.

TS and infection

While there is a wide consensus about the role of excessive dopamine transmission in TS and the implication of the basal ganglia (BG) in the disorder, the origins of the abnormalities of the BG and the dopaminergic pathways in TS are still debated. In the historical background chapter, I discussed the causal relation that has been established between streptococcus bacterial infection and Sydenham's chorea. Inspired by this link, the PANDAS project investigates possible links between streptococcal infection and TS for pediatric population. But not much progress has been achieved yet. It is a path worth further pursuit.

The somatosensory system

One qualitative finding of my study is that the following experiences lead to ticcing: (i) the urge for tactile symmetry, (ii) the irritation of being touched, and (iii) the urge to touch. In addition, intentional self-inflicted pain can help in suppressing tics and bringing a sense of relief. The threshold of the pain that provides the relief may increase, much like in addictive processes (See the chapter "Tic classification: Triggers of behavior"). The interactions with the somatosensory system, which comprises all of touch (non-noxious), pain, and proprioception (movement sense) inputs, may generate the experience of reward in a subject-specific manner[94].

Relying on existing paradigms of pain studies, the pain threshold of TS patients can be compared to that of healthy controls. And imaging studies can further inform the question of possible links between, pain, ticcing, and reward circuits.

The visual world

Based on participant testimonies and tic observations, I suggest that visual stimuli are important triggers of tics: (i) eye tics are typically the first to emerge, (ii) the eyes are common target for self mutilation (iii) antisaccade[95] of TS is abnormal, and (iv) reading is challenging to many with TS. In addition, many do not tic in their sleep or have ameliorated tics during their sleep or in situations in which their eyes are closed (See the chapter "Tic classification: Triggers of behavior").

The superior colliculus (SC), which is implicated in saccadic eye movement, has a direct path to the substantia nigra par reticulate (SNr) and direct projection from the

[94] I thank Krish Sathian for discussing this possibility with me.

[95] In the antisaccade paradigm the subject is instructed to saccade away from a novel visual stimulus.

retina. These pathways make the SC an interesting candidate for participating in TS. One speculation is that an SC abnormality may be linked to having two incompatible representations of visual stimuli; one from the direct retina pathway and one from the cortex. If the two disagree, attempts to align them, by eye blinking or head movement, might be the origin of tics.

Another, more robust, hypothesis is that saccadic eye movements can serve as possible surrogates for failure of inhibition in TS. Abnormal antisaccadic eye movements in TS patients have been documented in a number of studies (Munoz & Everling, 2004; Munoz, LeVasseur, & Flanagan, 2002). Antisaccadic eye movements are a simple and readily measurable index of inhibition similar to the time tested "go/no-go" paradigm. This paradigm is used to test inhibitions in frontal lobe dysfunction, which is a consistent finding in neuropsychological studies in TS (Butler, Stern, & Silbersweig, 2006). But their significance to the pathophysiology of TS is unknown.

Antisaccadic studies might interrelate a number of well described but poorly understood cognitive and physiological defects in TS, many of which could be tested through the study of saccadic eye movements. In addition to tics, failure of inhibition may contribute to attentional, obsessional and even sleep disorders in TS. These co-morbid conditions significantly contribute to the disability of the disorder, yet are not part of the primary of TS research (King , Leckman, Scahill, & Cogen, 1999).

Behavioral therapies

Behavioral therapies are important since they can support successful coping and improve the individual's quality of life. In the chapter "Interventions" I proposed that the meditation-derived techniques of mindfulness can ameliorate tics and provide the

afflicted with coping strategies. To the best of my knowledge, this has not been tried for TS. Emory, with its meditation studies and research of the effect of meditation on cognition and health, is in especially good position to try this therapy. And I intend to carry out such a study in the future.

In closing

Each new observation and treatment dynamically changes our understanding of TS. The disorder's definition is in constant dynamical relationship with the *zeitgeist* and with its presentation by the afflicted to his or her society. Despite public awareness and growing scientific activity, TS remains elusive and its hard questions resist solutions. But we should not give up. Some afflicted with TS provide models of persistence. On the 28th of April, 2009, Nick van Bloss, 41, played in a comeback concert at the Cadogan Hall in London. This award-winning student of the Royal College of Music had given up his performing career 15 years ago, since his TS got worse with the years. At his youth, whenever he played his tics stopped, until one day he had an attack on stage. He stopped performing.

He calls the Tourette's "a curse," but after many years, rather than fighting it, he has embraced it. "It is a fundamental part of me," he says. "I am a person with Tourette's who happens to be a pianist and a musician. That's really my identity - not the freak pianist" (Davies, 2009). It seems van Bloss has learned how to cope, and with excellence: "[I]n the Beethoven *Emperor* concerto… a capacity for expression shyly revealed itself. The grand opening really was grand, and the gentler almost nostalgic melody that comes later was sensitively molded," writes Ivan Hewett in the *Telegraph* on the day after the concert in Cadogan Hall (Hewett, 2009). But it took van Bloss 15 years of playing only

"to the simplicity of the four non judgmental walls at home." His coping can become a model for the afflicted, his persistence a guide for the scientist.

ACKNOWLEDGEMENTS

I am grateful for the friendship and encouragement of many people and institutions. Applying for a second doctoral program late in life required emotional support and words of wisdom. Those I received from my sister Ziva Joseff, and my friends Prof. Mel Konner and Leslee and Dean Bobby Paul. They encouraged me even before I applied and continued throughout my journey.

Two departments at Emory University have been my home in the last six years. The Institute of Liberal Arts (ILA), of which I am a graduate student, has been supportive, both intellectually and financially. Faculty members have always made themselves available and provided helpful guidance; especially Prof. Angelika Bammer, Prof. Kevin Corrigan, Prof. Howard Kushner, Prof. Michael Moon, Prof. Laura Otis, and Prof. Walt Reed. My cohorts have allowed me to feel as one of them despite our differences; especially Aukje Kluge and Liz Milewicz.

The psychology department has been my foster home. This is where I took most of my courses and found the intellectual space within which to study perceptions, memory, language, emotions, and social cognition within a monistic framework. The faculty members have been generous to me, as though their department were my native home; especially Prof. Larry Barsalou, Prof. Robin Fivush, Prof. Stephan Hamann, Prof. Debra Mills (now at Bangor University in the UK), Prof. Laura Namy, Prof. Lynne Nygaard, Prof. Philippe Rochat, Prof. Kim Wallen, and Prof. Phillip Wolff. My student fellows made me feel welcome and young; especially the students in Barsalou's lab: Christie Wilson with whom I exchanged ideas about cognitive psychology and life; Aron Barbey, now at the NIH, with whom I discussed research ideas and exchanged

experimental tips; and Kyle Simmons, now at the NIH, who has become a dear friend and a wonderful colleague.

Prof. Stuart Zola provided me with financial support through my first summer as a student, and introduced me to the hippocampus. Prof. Bob McCauley introduced me to different schools of philosophy of mind. Prof. Paul Lennard helped me refine my presentation of Wernicke's ideas about brain regions and circuits. Prof. Mahlon Delong, Prof. Bob Gross, and Prof. Klaus Mewes introduced me to the world of deep brain stimulation. Prof. Jim Rilling helped me getting acquainted with the cerebellum. Prof. Krish Sathian introduced me in a very generous way to Dr. Juncos, and as my work progressed helped me thinking about the somatosensory system in TS patients. Prof. Michael Haber helped me decide which statistics to apply for my data analysis. And Prof. Claire Sterk initiated me into qualitative research and the world of grant applications.

Rob Poh of Georgia State University was a sensitive and generous cameraman who video-recorded all my interviews and provided me with helpful feedback. Jocelyn Taylor spent long hours coding many of the video-recorded interviews and ran various statistics on them.

The Graduate School of Emory provided me with a stipend for my first four academic years in school. It also contributed towards conference expenses during my six years in school. I received much administrative support from the office of the ILA, especially from Tracy Allen and Michele Kelley. I also received much technical support at the ECIT services of the Woodruff library, especially from Wayne Morse and Rob O'Reilly and the graduate students at the service desk. The Emory Clinic for Movement

Disorders allowed me to use the same one room on 16 different Friday afternoons, even though the clinic is crowded and its space is limited.

My advisor is Prof. Howard Kushner, Nat C Robertson Distinguished Professor of the History of Medicine; and my committee members are Prof. Larry Barsalou, Samuel Candler Dobbs Professor of Cognitive Psychology; and Prof. Jorge Juncos, neurologist, psychiatrist, and a member of the Medical Board of the Tourette Syndrome Association. I owe Kushner everything that I know and understand about the complexity and subtlety of illness and health and how they are constructed from the interactions of biology, culture, family, and everything else in the individual's environment. Barsalou opened the world of cognitive neuroscience for me. In his laboratory and seminars I learned much about quantitative methods, and how to apply them to studies of behavior and the brain. His scholarship has been inspiring; his commitment to embodiment assured me that I have been in the right place for my monistic search. Juncos, who treats so many Tourette syndrome patients and knows so much about them and their disorders, helped me recruit his patients for my study, answered many of my questions, taught me about medications, and further contributed to my study by filling medical observational questionnaires about the participants. Prof. Alan Cienki, linguist, who has left us for the *Vrije Universiteit* in Amsterdam and is not on my committee any longer, opened up the world of cognitive linguistics to me. Metaphors, syntax, semantics, and gestures have taken new meaning under his tutelage. I have been most fortunate with all these mentors, and am deeply grateful to them.

My study would be impossible if it were not for the generosity of sixteen adults who suffer from Tourette syndrome and agreed to share their suffering, hopes, and

insights with me. Most of them came with a relative or a close friend who were also interviewed. Meeting all of them changed the course of my study and has been a deeply moving experience. I thank them all and hope that enough progress would be made in understanding their Tourette's so that they would be better helped.

My husband David and our daughter Aria are my deepest joy. No words can express how deeply grateful I am for having them and their total support in my life.

TIC SYMPTOM SELF REPORT

Name:	ID#:
Date:	
Form completed by: (check 4 all that apply) o self o mother o father o other:	

DIRECTIONS: Below is a list of "tic" symptoms. Tics are usually quick, jerking movements or repetitive sounds that are difficult to control. Considering both their **frequency** and **forcefulness**, please rate the severity of each one of your tics **during the past week** by placing one score (0-3; see below) in the appropriate box.

SCORING:

0 = No symptoms at all this past week.	**2** = Tics were frequent and forceful.
1 = Tics were infrequent and not forceful.	**3** = Tics were very frequent and very forceful.

MOTOR TIC SYMPTOM	SCORE	VOCAL TIC SYMPTOM	SCORE
Eyeblinking		Grunting	
Eye movements		Throat clearing	
Jaw or mouth movements		Coughing	
Facial tics		Snorting	
Head jerks		Whistling	
Shoulder jerks		Bird noises (hooting)	
Arm movements		Animal noises (barking)	
Finger or hand movements		Squeaking	
Leg kicking		Other noises:	
Abdominal tensing		Gulping	
Tensing arms or legs		Humming	
Repetitive touching		Breathing tics	
Repetitive finger tapping		Repeating single words or syllables	
Hitting self		Blocking in speech	
Picking at things (clothing, etc.)		Voice changes (volume or pitch)	
Unusual body postures		Obscene words or cursing	
Skipping or twirling		Repeating own words/sentences	
Obscene gestures		Repeating other's speech	
Combined movements (specify):		Combined tics (specify):	
Other:		Other:	

Total Motor ☐ **Total Phonic** ☐ **Overall Total** ☐

.

APPENDIX II: YALE-BROWN OBSESSIVE COMPULSIVE CHECKLIST

Name _____ **Date** _____

CY-BOCS OBSESSIONS CHECKLIST

Check all items that apply (Item marked "*" may or not be OCD phenomena.)

Current Past Contamination Obsessions

____ ____ Concern with dirt, germs, certain illnesses (e.g., AIDS)
____ ____ Concerns or disgust with bodily waste or secretions (e.g., urine, feces, saliva)
____ ____ Excessive concern with environmental contaminants (e.g., asbestos, radiation, toxic waste)
____ ____ Excessive concern with household items (e.g., cleaners, solvents)
____ ____ Excessive concern about animals/insects
____ ____ Excessively bothered by sticky substances or residues
____ ____ Concerned will get ill because of contaminant
____ ____ Concerned will get others ill by spreading contaminant (aggressive)
____ ____ No concern with consequences of contamination other than how it might feel *
____ ____ Other (Describe) _____

Aggressive Obsessions

Fear might harm self
____ ____ Fear might harm others
____ ____ Fear harm will come to self
____ ____ Fear harm will come to others (may be because something child did or did not do)
____ ____ Violent or horrific images
____ ____ Fear of blurting out obscenities or insults
____ ____ Fear of doing something else embarrassing *
____ ____ Fear will act on unwanted impulses (e.g. to stab a family member)
____ ____ Fear will steal things
____ ____ Fear will be responsible for something else terrible happening (e.g. fire, burglary, flood)
____ ____ Other (Describe) _____

Sexual Obsessions

[Are you having any sexual thoughts? If yes, are they routine or are they repetitive thoughts that you would rather not have or find disturbing? If yes, are they:]
____ ____ Forbidden or perverse sexual thoughts, images, impulses
____ ____ Content involves homosexuality *
____ ____ Sexual behavior towards others (Aggressive)
____ ____ Other (Describe) _____

Hoarding/Saving Obsessions

____ ____ Fear of losing things
____ ____ Other (Describe) _____

Magical Thoughts/Superstitous Obsessions

____ ____ Lucky/unlucky numbers, colors, words
____ ____ Other (Describe) _____

Current	Past	Somatic Obsessions
____	____	Excessive concern with illness or disease *
____	____	Excessive concern with body part or aspect of appearance (e.g., dysmorphophobia) *
____	____	Other (Describe) _____

Religious Obsessions (Scrupulosity)

____	____	Excessive concern or fear of offending religious objects (God)
____	____	Excessive concern with right/wrong, morality
____	____	Other (Describe) _____

Miscellaneous Obsessions

____	____	The need to know or remember
____	____	Fear of saying certin things
____	____	Fear of not saying just the right thing
____	____	Intrusive (non-violent) images
____	____	Intrusive sounds, words, music, or numbers
____	____	Other (Describe) _____

TARGET SYMPTOM LIST FOR OBSESSIONS

Obsessions (Describe, listing by order of severity, with #1 being the most severe, #2 the second most severe, etc.):

1. _____
2. _____
3. _____
4. _____

APPENDIX III: YALE GLOBAL TIC SEVERITY SCALE

NAME: TODAY'S DATE :

/ /

RATER:

MOTOR TIC SYMPTOM CHECKLIST (Check motor tics present during past week.)

•**Simple Motor Tics** (Rapid, Darting, "Meaningless"):

- o Eye blinking
- o Eye movements
- o Nose movements
- o Mouth movements
- o Facial grimace
- o Head jerks/movements
- o Shoulder shrugs
- o Arm movements
- o Hand movements
- o Abdominal tensing
- o Leg, foot, or toe movements
- o Other (describe):

o Other (describe):

•**Complex Motor Tics** (Slower, "Purposeful"):

o Eye movements

o Mouth movements

o Facial movements or expressions

o Head gestures or movements

o Shoulder movements

o Arm movements

o Hand movements

o Writing tics

o Dystonic postures

o Bending or gyrating

o Rotating

o Leg or foot or toe movements

o Blocking

o Tic related compulsive behaviors (touching, tapping, grooming, evening-up)

o Copropraxia

o Self-abusive behavior

o Paroxysms of tics (displays), duration ___ seconds

o Disinhibited behavior (describe):*

o Other (describe):

PHONIC TIC SYMPTOM CHECKLIST (Check phonic tics present over the **past week**.)

•**Simple Phonic Symptoms** (Fast, "Meaningless" Sounds):

o Sounds, noises (circle: coughing, throat clearing, sniffing, or animal or bird noises)

o Other (list):

•**Complex Phonic Symptoms** (Language: Words, Phrases, Statements):

o Syllables (list)

o Words (list)

o Coprolalia (list)

o Echolalia

o Palalalia

o Blocking

o Speech atypicalities (describe)

o Disinhibited speech (describe)*

* Do not include disinhibitions in ratings of tic behaviors

NUMBER	Motor	Phonic	
None	o	o	0
Single tic	o	o	1
Multiple discrete tics (2-5)	o	o	2
Multiple discrete tics (>5)	o	o	3
Multiple discrete tics plus as least one orchestrated pattern of multiple simultaneous or sequential tics where it is difficult to distinguish discrete tics	o	o	4
Multiple discrete tics plus several (>2) orchestrated paroxysms of multiple simultaneous or sequential tics that where it is difficult to distinguish discrete tics	o	o	5

FREQUENCY	Motor	Phonic	
NONE No evidence of specific tic behaviors	o	o	0

RARELY Specific tic behaviors have been present during previous week. These behaviors occur infrequently, often not on a daily basis. If bouts of tics occur, they are brief and uncommon.	o	o	1
OCCASIONALLY Specific tic behaviors are usually present on a daily basis, but there are long tic-free intervals during the day. Bouts of tics may occur on occasion and are not sustained for more than a few minutes at a time.	o	o	2
FREQUENTLY Specific tic behaviors are present on a daily basis. tic free intervals as long as 3 hours are not uncommon. Bouts of tics occur regularly but may be limited to a single setting.	o	o	3
ALMOST ALWAYS Specific tic behaviors are present virtually every waking hour of every day, and periods of sustained tic behaviors occur regularly. Bouts of tics are common and are not limited to a single setting.	o	o	4
ALWAYS Specific tic behaviors are present virtually all the time. Tic free intervals are difficult to identify and do not last more than 5 to 10 minutes at most.	o	o	5

INTENSITY	Motor	Phonic	
Absent	o	o	0

Minimal intensity Tics not visible or audible (based solely on patient's private experience) or tics are less forceful than comparable voluntary actions and are typically not noticed because of their intensity.	o	o	1
Mild intensity Tics are not more forceful than comparable voluntary actions or utterances and are typically not noticed because of their intensity.	o	o	2
Moderate intensity Tics are more forceful than comparable voluntary actions but are not outside the range of normal expression for comparable voluntary actions or utterances. They may call attention to the individual because of their forceful character.	o	o	3
Marked intensity Tics are more forceful than comparable voluntary actions or utterances and typically have an "exaggerated" character. Such tics frequently call attention to the individual because of their forceful and exaggerated character.	o	o	4
Severe intensity Tics are extremely forceful and exaggerated in expression. These tics call attention to the individual and may result in risk of physical injury (accidental, provoked, or self-inflicted) because of their forceful expression.	o	o	5

COMPLEXITY	Motor	Phonic

None If present, all tics are clearly "simple" (sudden, brief, purposeless) in character.	o	o	0
Borderline Some tics are not clearly "simple" in character.	o	o	1
Mild Some tics are clearly "complex" (purposive in appearance) and mimic brief "automatic" behaviors, such as grooming, syllables, or brief meaningful utterances such as "ah huh," "hi" that could be readily camouflaged.	o	o	2
Moderate Some tics are more "complex" (more purposive and sustained in appearance) and may occur in orchestrated bouts that would be difficult to camouflage but could be rationalized or "explained" as normal behavior or speech (picking, tapping, saying "you bet" or "honey", brief echolalia).	o	o	3
Marked Some tics are very "complex" in character and tend to occur in sustained orchestrated bouts that would be difficult to camouflage and could not be easily rationalized as normal behavior or speech because of their duration and/or their unusual, inappropriate, bizarre or obscene character (a lengthy facial contortion, touching genitals, echolalia, speech atypicalities, longer bouts of saying "what do you mean" repeatedly, or saying "fu" or "sh").	o	o	4

Severe Some tics involve lengthy bouts of orchestrated behavior or speech that would be impossible to camouflage or successfully rationalize as normal because of their duration and/or extremely unusual, inappropriate, bizarre or obscene character (lengthy displays or utterances often involving copropraxia, self-abusive behavior, or coprolalia).	o	o	5

INTERFERENCE	Motor	Phonic	
None	o	o	0
Minimal When tics are present, they do not interrupt the flow of behavior or speech.	o	o	1
Mild When tics are present, they occasionally interrupt the flow of behavior or speech.	o	o	2
Moderate When tics are present, they frequently interrupt the flow of behavior or speech.	o	o	3
Marked When tics are present, they frequently interrupt the flow of behavior or speech, and they occasionally disrupt intended action or communication.	o	o	4
Severe When tics are present, they frequently disrupt intended action or communication.	o	o	5

IMPAIRMENT _____

None	o	0
Minimal Tics associated with subtle difficulties in self-esteem, family life, social acceptance, or school or job functioning (infrequent upset or concern about tics vis a vis the future, periodic, slight increase in family tensions because of tics, friends or acquaintances may occasionally notice or comment about tics in an upsetting way).	o	10
Mild Tics associated with minor difficulties in self-esteem, family life, social acceptance, or school or job functioning.	o	20
Moderate Tics associated with some clear problems in self-esteem family life, social acceptance, or school or job functioning (episodes of dysphoria, periodic distress and upheaval in the family, frequent teasing by peers or episodic social avoidance, periodic interference in school or job performance because of tics).	o	30
Marked Tics associated with major difficulties in self-esteem, family life, social acceptance, or school or job functioning.	o	40
Severe Tics associated with extreme difficulties in self-esteem, family life, social acceptance, or school or job functioning (severe depression with suicidal ideation, disruption of the family (separation/divorce, residential placement), disruption of social tics - severely restricted life because of social stigma and social avoidance, removal from school or loss of job).	o	50

APPENDIX IV: YALE-BROWN OBSESSIVE-COMPULSIVE SCALE

For each item circle the number identifying the response which best characterizes the patient.

1. Time Occupied by Obsessive Thoughts

How much of your time is occupied by obsessive thoughts? How frequently do the obsessive thoughts occur?

0 = None

1 = Mild (less than 1 hr day) or occasional (intrusion occurring no more than 8 times a day)

2 = Moderate (1 to 3 hrs a day) or frequent (intrusion occurring more than 8 times a day, but most of the hours of the day are free of obsessions)

3 = Severe (greater than 3 and up to 8 hrs day) or very frequent (intrusion occurring more than 8 times a day and occurring during most of the hours of the day)

4 = Extreme (greater than 8 hrs day) or near consistent intrusion (too numerous to count and an hour rarely passes without several obsessions occurring)

2. Interference Due to Obsessive Thoughts

How much do your obsessive thoughts interfere with your social or work (or role) functioning? Is there anything that you don't do because of them?

0 = None

1 = Mild, slight interference with social or occupational activities, but overall performance not impaired

2 = Moderate, definite interference with social or occupational performance but still manageable

3 = Severe, causes substantial impairment in social or occupational performance

4 = Extreme, incapacitating

3. Distress associated with Obsessive Thoughts

How much distress do your obsessive thoughts cause you?

0 = None

1 = Mild, infrequent and not too disturbing

2 = Moderate, frequent and disturbing but still manageable

3 = Severe, very frequent and very disturbing

4 = Extreme, near constant and disabling distress

4. Resistance Against Obsessions

How much of an effort do you make to resist the obsessive thoughts? How often do you try to disregard or turn your attention away from these thought as they enter your mind?

0 = Makes an effort to always resist, or symptoms so minimal doesn't need to actively resist

1 = Tries to resist most of the time

2 = Makes some effort to resist

3 = Yields to all obsessions without attempting to control them, but does so with some reluctance

4 = Completely and willingly yields to all obsessions

5. Degree of Control Over Obsessive Thoughts

How much control do you have over obsessive thoughts? How successful ae you in stopping or diverting your obsessive thinking?

0 = Complete control

1 = Much control, usually able to stop or divert obsessions with some effort and concentration

2 = Moderate control, sometimes able to stop or divert obsessions

3 = Little control, rarely successful in stopping obsessions

4 = No control, experienced as completely involuntary, rarely able to even momentarily divert thinking

6. Time Spent Performing Compulsive Behaviors

How much time do you spend performing compulsive behaviors? How frequently do you perform compulsions?

0 = None

1 = Mild (less than 1 hr day performing compulsions) or occasional (performance of compulsions occurring no more than 8 hrs a day)

2 = Moderate (1 to 3 hrs day performing compulsion) or frequent (performance of compulsions occurring more than 8 times a day, but most of the hours of the day are free of compulsive behaviors)

3 = Severe (greater than 3 and up to 8 hrs day performing compulsions) or very frequent (performance of compulsions occurring during most of the hours of the day)

4 = Extreme (greater than 8 hrs day performing compulsions) or near consistent performance of compulsions (too numerous to count and an hour rarely passes without several compulsions being performed)

7. Interference Due to Compulsive Behaviors

How much do your compulsive behaviors interfere with your social or work (or role) functioning? Is there anything that you don't do because of the compulsions?

0 = None

1 = Mild, slight interference with social or occupational activities, but overall performance not impaired

2 = Moderate, definite interference with social or occupational performance but still manageable

3 = Severe, causes substantial impairment in social or occupational performance

4 = Extreme, incapacitating

8. Distress Associated with Compulsive Behavior

How would you feel if prevented from performing your compulsions?

How anxious would you become? How anxious do you get while performing compulsions until you are satisfied they are completed?

0 = None

1 = Mild, only slightly anxious if compulsions prevented or only slightly anxious during performance of compulsions

2 = Moderate, reports that anxiety would mount but remain manageable if compulsions prevented or that anxiety increases but remains manageable during performance of compulsions

3 = Severe, prominent and very disturbing increase in anxiety if compulsions interrupted or prominent and very disturbing increases in anxiety during performance of compulsions

4 = Extreme, incapacitating anxiety from any intervention aimed at modifying activity or incapacitating anxiety develops during performance or compulsions

9. Resistance Against Compulsions

How much of an effort do you make to resist the compulsions?

0 = Makes an effort to always resist, or symptoms so minimal doesn't need to actively resist

1 = Tries to resist most of the time

2 = Makes some effort to resist

3 = Yields to all compulsions without attempting to control them, but does so with some reluctance

4 = Completely and willingly yields to all compulsions

10. Degree of Control Over Compulsive Behavior

How much control do you have over obsessive thoughts? How successful ae you in stopping or diverting your obsessive thinking?

0 = Complete control

1 = Much control, experiences pressure to perform the behavior but usually able to exercise voluntary control over it

2 = Moderate control, strong pressure to perform behavior, can control it only with difficulty

3 = Little control, very strong drive to perform behavior, must be carried to completion, can only delay with difficulty

4 = No control, drive to perform behavior experienced as completely involuntary

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Source: Goodman WK, Price LH, Rasmussen SA, et al. The Yale-Brown Obsessive-Compulsive Scale, 1: development, use, and reliability. *Arch Gen Psychiatry* 46:1006-1011,1989

APPENDIX V: PATIENT-COPROLALIA INTERVIEW GUIDE

Comments for the interviewer are in square brackets [].The **bold headings** define topics and are not questions.

Greetings

(1) Thank you for your participation in the study and for giving me some of your time

The emergence of symptoms

(2) What is your earliest memory of any Tourette symptoms?
(3) What were they?

Naming the behavior and describing it

(4) What was the first time that you remember saying things that you did not mean to say?
(5) How do you name these expressions? [This is motivated by possible denial. The subjective view of the syndrome is of interest and has to be respected. For the rest of the interview I will use the person's own label for coprolalia. I mark it below as OOO].
(6) What are some examples?

The effects of diagnosis

(7) When were you diagnosed with TS?
(8) How were things different before and after your diagnosis?
- In the response of your family
- In the response of your friends
- In the response of your teachers
- In your own feelings about yourself

Patterns of tics and coprolalia

(9) When do you have the most tics?
(10) What about OOO and copropraxia?

(11) When do you have the least tics?
(12) What about OOO and copropraxia?
(13) Why do you think you have these tics?
(14) Why do you think you have OOO and copropraxia?
(15) What is the nature of the tics
- When you are relaxed?
- When you perform rhythmic activities (e.g., biking, running, washing dishes)?
- When you are excited (i.e. aroused)?
- When you experience a new situation?
- When you are alone?
- What happens during sleep?

(16) What are the OOO and the like
- When you are relaxed?
- When you perform rhythmic activities (e.g., biking, running, washing dishes)?
- When you are excited (i.e. aroused)?
- When you experience a new situation?
- When you are alone?
- What happens during sleep?

(17) What about copropraxia?
(18) [If the urge was not mentioned so far] The OOO can be anticipated or come as a surprise. How is it for you?
(19) How about copropraxia?

The urge

(20) Tell me about the urges
- Before a OOO, for instance in the examples that you brought earlier (question 6).
- What is the relation between the urge and what you say? Is the urge specific and "telling" you what to say or is there just an urge to say something?
- When you OOO a person, how does it relate to whether you like or dislike the person?
- Some know before they OOO what they are going to say. How is it for you?

(21) What do you feel towards the person immediately before you OOO him/her?
(22) What is the difference between times that you suppress and do not suppress the urge to OOO.
(23) What is the difference between the urge before a motor or a vocal tic and OOO?

After the coprolalia behavior

(24) Let's talk about what happens immediately after the OOO.

- Does the urge get fulfilled by the tic or the OOO, or you are left with a feeling that some of the urge is still there, unfulfilled.
- How do your feelings after the tic or the OOO depend on the respond of the other people who are present?

(25) I am trying to see if there is any connection between tics and OOO. Do you have any thoughts or feelings about such possible connection?

Money

(26) Please describe to me your money handling habit: Shopping, saving, credit cards, etc.

(27) [SRF: Probe here about arithmetic and math]

Reading

(28) Do you read?

1. If yes – what do you read?
2. If not – why not?

Closing questions

(29) Are there more things that you care to tell me?

(30) Are there questions that I should have asked and have not asked you?

(31) Thank you.

APPENDIX VI: PARTNER-COPROLALIA INTERVIEW GUIDE

Comments for the interviewer are in square brackets [].The **bold headings** define topics and are not questions. The patient's name is designated in the Guide with XXX.

Greetings

(1) Thank you for your participation in the study and for giving me of your time

Observing the development of symptoms

(2) What is your relationship to XXX?
(3) What is the first time that you remember noticing any Tourette signs for XXX?

Observing the response of the patient and of other people

(4) What did you think they were?
(5) How did you respond?
(6) How did other people respond?
(7) How did XXX respond?

Observing the signs and the response to them

(8) What was the first time that coprolalia occurred?
(9) How did you respond?
(10) How did others respond?
(11) How did XXX respond?
(12) When was XXX diagnosed with TS?
(13) How did the diagnosis change things?

Observing patterns of tics and coprolalia

(14) When would you say, XXX has the most tics and coprolalia?
(15) When does XXX have the least tics and coprolalia?
(16) Please describe XXX's tics and coprolalia
- When s/he is relaxed
- When s/he performs rhythmic activities (e.g., biking, running, washing dishes)

- When s/he is excited (i.e. aroused)
- When s/he is in a new situation
- What happens to him/her during sleep?

(17) Are there any copropraxia tics?
(18) Are there other situations that I have not mentioned that affect the patterns of the tics and coprolalia?
(19) Please describe to me XXX's money handling habits: Shopping, saving, credit cards, etc.

Patterns of coprolalia expression

(20) How would you say, the coprolalia relates to whether XXX likes or dislikes a person?
(21) How would you say, the coprolalia relates to whether the person is familiar or is a stranger?
(22) How would you say, the copropraxia relates to whether the person is familiar or is a stranger?
(23) How do the people who are the target of the coprolalia and/or copropraxia respond?

Response of other people who are present

(24) How do other people who are present respond?
(25) How does the anticipation of people's response affect XXX's behavior?

Before and after the coprolalia expression

(26) You are close with XXX. What are any clues that predict the upcoming of a coprolalia expression?
(27) How would you describe the change in behavior after the expression of coprolalia?
(28) How would you describe the change in mood after the expression of coprolalia?
(29) [If XXX named the coprolalia as OOO]. XXX refers to his/her coprolalia as OOO. What can you tell me about this?

Coprolalia vs. other aspects of TS

(30) How would you compare the tics and the coprolalia?
- For XXX?
- For you?
- For other people?

Money

(32) Please describe to me XXX's money handling habit: Shopping, saving, credit cards, etc.
(33) [SRF: Probe here about arithmetic and math]

Reading

(34) Does XXX read?
1. If yes – do you know what XXX read?
2. If not – do you know why not?

Closing questions

(31) Are there more things that you care to tell me?
(32) Are there questions that I should have asked and have not asked you?
(33) Thank you

APPENDIX VII: PATIENT-NO-COPROLALIA INTERVIEW GUIDE

Comments for the interviewer are in square brackets [].The **bold headings** define topics and are not questions.

Greetings

(1) Thank you for your participation in the study and for giving me some of your time

The emergence of symptoms

(2) What is your earliest memory of any Tourette symptoms?
(3) What were they?

The effects of diagnosis

(4) When were you diagnosed with TS?
(5) How were things different before and after your diagnosis?
- In the response of your family
- In the response of your friends
- In the response of your teachers
- In your own feelings about yourself

Patterns of tics

(6) Which tics do you have? [See if there are any possible copropraxia]
(7) When do you have the most tics?
(8) When do you have the least tics?
(9) Why do you think you have these tics?
(10) What is the nature of the tics
- When you are relaxed?
- When you perform rhythmic activities (e.g., biking, running, washing dishes)?
- When you are excited (i.e. aroused)?
- When you experience a new situation?

- When you are alone?
- What happens during sleep?

(11) [If the urge was not been mentioned so far] The tic can be anticipated or come as a surprise. How is it for you?

The urge

(12) What is the urge like
- Before a motor tic
- Before a vocal tic

(13) Sometimes it is possible to suppress the urge. What is the difference between times that you suppress and do not suppress the urge?

After-the-tic behavior

(14) Let's talk about what happens immediately after the tic.
- Does the urge get fulfilled by the tic or you are left with a feeling that some of the urge is still there, unfulfilled.
- How do your feelings after the tic depend on the respond of the other people who are present?

(15) I am trying to see if there is any connection between motor tics and vocal tics. Do you have any thoughts or feelings about such possible connection?

Money

(16) Please describe to me your money handling habit: Shopping, saving, credit cards, etc.

(17) [SRF: Probe also about math abilities]

Reading

(18) Do you read?
a. If yes – what do you read?
b. If not – why not?

Closing questions

(19) Are there more things that you care to tell me?

(20) Are there questions that I should have asked and have not asked you?
(21) Thank you.

APPENDIX VIII: PARTNER-NO-COPROLALIA INTERVIEW GUIDE

Comments for the interviewer are in square brackets []. The **bold headings** define topics and are not questions. The patient's name is designated in the Guide with XXX.

Greetings

(1) Thank you for your participation in the study and for giving me of your time

Observing the development of symptoms

(2) What is your relationship to XXX?
(3) What is the first time that you remember noticing any Tourette signs for XXX?
(4) What were the symptoms?
(5) Which tics does XXX have now? [SRF: see if there are any copropraxia]

The response of the patient and of people who play a role in the patient's life

(6) What did you think they were?
(7) How did you respond?
(8) How did other people respond?
(9) How did XXX respond?
(10) When was XXX diagnosed with TS?
(11) How did the diagnosis change things?

Patterns of tics and coprolalia

(12) When would you say, XXX has the most tics?
(13) When does XXX have the least tics?
(14) How are the tics of XXX
- When s/he is relaxed
- When s/he performs rhythmic activities (e.g., biking, running, washing dishes)
- When s/he is excited (i.e. aroused)
- When s/he is in a new situation
- What happens to him/her during sleep?

(15) Are there other situations that I have not mentioned that affect the patterns of the tics?

Money

(22) Please describe to me XXX's money handling habits: Shopping, saving, credit cards, etc.

Response of other people who are present

(16) How do other people who are present respond?
(17) How does the anticipation of people's response affect XXX's behavior?

Before and after the tic

(18) You are close with XXX. What are any clues that predict the upcoming of a tic?
(19) How would you describe the change in behavior after the tic?
(20) How would you describe the change in mood after the tic?

Money

(23) Please describe to me how XXX handles money: Shopping, saving, credit cards, etc.
(24) [SRF: Probe here about arithmetic and math]

Reading

(25) Does XXX read?
   a. If yes – what do you read?
   b. If not – why not?

Closing questions

(21) Are there more things that you care to tell me?
(22) Are there questions that I should have asked and have not asked you?
(23) Thank you

APPENDIX IX: PATIENT-AFTER-DBS INTERVIEW GUIDE

Only very severe cases of TS patients are selected for deep brain stimulation (DBS). All had coprolalia prior to the DBS.

Comments for the interviewer are in square brackets [].The **bold headings** define topics and are not questions. The patient's own labeling of coprolalia from the previous visit is designated in the Guide with OOO.

Greetings

(1) Thank you for coming again and giving me of your time
(2) You have recently undergone a surgery. How are you?

The change

(3) How are things now compared to how they were before the surgery
- What happened to your motor tics?
- What happened to your vocal tics?
- What happened to your OOO?
- What happened to your copropraxia?
- Have your habits with money changed?
- Have your reading habits changed?

(4) Please describe how people respond to the changes in you?
- Relatives
- **[**See if there is room to mention friends, teachers, neighbors, and colleagues].

(5) Which symptoms of Tourette are still with you?
(6) What would you say you have gained from the surgery?
(7) If you lost anything because of the surgery, what is it?
(8) What happened to the urges that preceded your tics and OOO?
(9) If an urge appears now, how do you respond to it?
- For tics
- For OOO

(10) What are other changes in your life that follow the DBS.

- Gait
- Concentration
- Being awake
- Having interest in things and curiosity
- Physical activities
- Other activities
- Friends
- General sense of well being
- Money handling habits: Shopping, saving, credit cards, etc.

Closing

(11) In our previous interview I told you that I am looking to see if there is any connection between tics and OOO. Do you have any new thoughts about such possible connection?
(12) Are there more things that you care to tell me?
(13) Are there questions that I should have asked and have not asked you?
(14) Thank you [Add, as appropriate, wishes for complete recovery from the surgery]

APPENDIX X: PARTNER-AFTER-DBS INTERVIEW GUIDE

Only very severe cases of TS patients are selected for deep brain stimulation (DBS). All had coprolalia prior to the DBS.

Comments for the interviewer are in square brackets [].The **bold headings** define topics and are not questions. The patient's name is designated in the Guide with XXX.

Greetings

(1) Thank you for coming again and giving me of your time

The Change

(2) XXX has recently undergone a surgery. How are things now compared to how they were before the surgery
- What happened to his/her motor tics?
- What happened to his/her vocal tics?
- What happened to his/her coprolalia?
- What happened to his/her copropraxia?

(3) How do people respond to these changes
- Relatives
- Friends
- Teachers
- Neighbors
- Colleagues.

(4) Which signs of Tourette are still there?
(5) What would you say has been gained from the surgery?
(6) If there was any loss resulting from the surgery, what is it?
(7) Please describe other changes in the life of XXX after the DBS.
- Gait
- Concentration
- Being awake
- Being interested in things
- Physical activities

- Other activities
- Friends
- General sense of well being
- Money habits: shopping, credit cards, etc.
- Reading habits

Closing

(8) I am trying to understand possible relations between coprolalia and other tics. Do you have any thoughts about such possible connection?
(9) Are there more things that you care to tell me?
(10) Are there questions that I should have asked and have not asked you?
(11) Thank you

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