



Critical aspects in the legal defence of patients with Tourette's Syndrome: An Italian case series

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ABSTRACT

Tourette's syndrome (TS) is a developmental neurobehavioral disorder with childhood onset and relevant burden in terms of disability and reduced quality of life. In Italy the biological basis of this syndrome is still frequently ignored and TS is often recognised as a psychiatric manifestation, or even it is not recognised as pathology, which may result in inadequate treatment, social isolation and improper hospitalization. Indeed, the organic medical nature of TS needs to be taken into great consideration in evaluating causality of committing crimes in affected patients. In addition, delaying the diagnosis and consequently proper treatment has a devastating impact on social as well as legal aspects in patients with TS. The present report is aimed to present an Italian case series of 4 TS patients who faced legal problems related to their mental condition, in the intent to add further evidence, raise the level of awareness and encourage further investigation in the field, as in most of the cases, patients' illness was not taken into adequate account by the Justice. The relevant law is discussed in detail.

1. Introduction

Tourette's syndrome (TS) is a neurobehavioral movement disorder (Jankovic & Kurlan, 2011) with childhood onset characterized by multiple motor and phonic tics, starting before the age of 18 and lasting for more than one year, as defined in the Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition (DSM 5) (American Psychiatric Association, 2013). TS is not an uncommon condition: according to a recent meta-analysis, in fact, TS prevalence estimate was 0.52% in the general population, and prevalence of TS in children was estimated to be 0.3% to 0.9% (Scharf et al., 2015). Indeed, TS prevalence is often underestimated due to misdiagnosis: affected individuals and caregivers, in fact, often do not recognize the symptoms of TS, which thus may delay proper identification and diagnosis (Mol Debes, Hjalgrim, & Skov, 2008) of a potentially treatable and manageable condition. TS pathophysiology is characterized by a dysregulation within the cortico-striato-thalamo-cortical pathway, and by an alteration of the neurotransmitter systems, principally dopamine and serotonin (Zapparoli et al., 2016; Zapparoli, Porta, & Paulesu, 2015). TS

seems to have a genetic basis, even though a single gene has not been identified (Dietrich et al., 2015). Streptococcal infections (Martino, Zis, & Buttiglione, 2015), also, could have an etiological role.

TS is considered a neurobehavioral disorder because -in 90% of cases- it includes also neurobehavioral manifestations (Kurlan et al., 2002; Robertson, 2000), such as attention-deficit/hyperactivity disorder (ADHD) (Schlander, Schwarz, Rothenberger, & Roessner, 2011), obsessive compulsive behaviors (OCB) (Eapen & Robertson, 2015) and, more rarely, obsessive compulsive disorder. In addition to these, TS patients can show co-occurring psychopathologies such as anxiety, depression, learning difficulties, personality and impulse control disorders, aggression, Non-Obscene Socially Inappropriate behavior, oppositional defiant disorder, conduct disorder and addiction. Considering the most prevalent components (ADHD and OC component), TS can be classified in different phenotypes (Eapen & Robertson, 2015; Robertson, 2015). In this TS classification, obsessive compulsive component results almost always present, therefore in 2017 a new subtype of TS was born in Italy with the name of OCTD (Obsessive-Compulsive Tic Disorder: a TS subtype characterized by both tics and OCB)

Abbreviations: TS, Tourette's syndrome; DSM 5, Diagnostic and Statistical Manual of Mental Disorders Fifth Edition; ADHD, Attention-Deficit Hyperactivity Disorder; OCB, Obsessive Compulsive Behaviors; OCTD, Obsessive-Compulsive Tic Disorder; DBS, Deep Brain Stimulation

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(Dell'Osso et al., 2017).

Even though tics are the most evident and known aspect of TS, the psychopathological manifestations and comorbidities are often the most disabling and worrying elements for TS patients: these symptoms often interfere with subjects' daily activities and can lead to a severe social impairment (Eddy et al., 2011), e.g. patients with TS may have difficulties in finding or keeping a job (Verdellen et al., 2011) and young patients might face marginalization at school. Exactly for this reason, social impairment, working and school issues may often become even more important than objective tic severity in therapeutic decisions (Roessner et al., 2011). As therapies, Habit Reversal Training (Verdellen et al., 2011) (i.e. the psychotherapy gold standard in TS), pharmacological treatments (Roessner et al., 2011; Shapiro & Shapiro, 1968) and DBS (Deep Brain Stimulation) surgery (Porta et al., 2016) are implemented nowadays; specifically, the use of off-label and not reimbursable drugs in TS patients is another current social/legal problem.

Ultimately, TS-related symptoms can lead to civil, penal, work-related and matrimonial law problems. The susceptibility of these patients regarding lawsuits and their inclination to adopt unlawful conducts has already been reported in literature (Jankovic, Kwak, & Frankoff, 2006). Actually, the impulse control deficit is a common element among TS symptoms. It manifests because of the above-mentioned cortico-striato-thalamo-cortical pathway dysregulation, and the neurotransmitters' alteration. A TS patient -because of impulsivity- maintains the capacity of understand, whereas the capacity of taking action is not maintained during the inconstant manifestation of symptoms. Consequently, it's hard for a judge to define the culpability of a TS patient in an unlawful act. And, potentially, patients can profit from their TS condition. Moreover, besides patients' involvement in law issues, caregivers and clinicians are frequently involved in lawsuits as well.

Criminal behaviour is a multifactorial and hard to frame issue: social and cultural aspects as well as personal reminders and actual context, together with psychological and neurological mechanisms are involved in its origin. Brain imaging has been used to study neurological features of people showing unlawful behaviors and some structures such as the amygdala, hippocampus and orbitofrontal cortex have shown some abnormalities in murderers (Anderson, Bechara, Damasio, Tranel, & Damasio, 1999; Laakso et al., 2001; Raine, Lencz, Bihle, Lacasse, & Colletti, 2000). Moreover, hypoperfusion and decreased glucose uptake in the prefrontal cortex have been associated with impulsiveness and aggressive, violent behaviors (Braun et al., 1995; Brower & Price, 2001; Soloff et al., 2003). It is well established that some medical conditions are linked to unlawful behaviors; just as other patients affected by neurological and psychiatric diseases, patients with TS (Gullucayir, Asirdizer, Yavuz, Zeyfeoglu, & Ulucay, 2009; Jankovic et al., 2006; Siponmaa, Kristiansson, Jonson, Nyden, & Gillberg, 2001) and patients with neurodevelopmental disorders who share some key-aspects with TS (Gittelman, Mannuzza, Shenker, & Bonagura, 1985; Siponmaa et al., 2001; Stewart, Cummings, Singer, & Deblouis, 1981), might face different legal problems linked with their condition due to the above-mentioned TS neurobehavioral symptoms and co-occurring psychopathologies. Taken as a whole, these elements increase the risk of indictment.

As reported by Jankovic et al. (Jankovic et al., 2006), there is a lack of data on the prevalence of unlawful behaviors or crimes in TS. As a consequence, prevention, treatment strategies and legal defence are currently insufficient.

The aim of this study is to prevent late diagnosis and late therapy in TS because they cause a high social impairment in patients and in their families, with frequent legal issues. To better explain TS-related law problems, we here report the cases of four Italian TS patients who faced legal issues linked to their disease: one for coprolalia and stalking, one for Non-Obscene Socially Inappropriate behavior and two for copropraxia. All the 4 cases were late diagnosed -only once they were adults- and therefore therapies couldn't prevent the social impairment during

their childhood and adolescence, leaving a sign in their psychological integrity. Actually, adolescence is the worst period for TS manifestations (Leckman & Cohen, 1999) and 2 of the 4 cases (i.e. case 2 and case 3) were also bullied for their TS. In the case description, though, we didn't report their bullying episodes. Viceversa we underline the fact that they were unjustly treated in front of the justice because of their pathological TS diseases.

2. Cases

The following 4 subjects consented for the publication of their syndrome history, therapies and legal reports. In the study, two different TS clinical scales have been administered by clinicians: the first one, the Yale Global Tic Severity Scale (YGTSS) (Leckman et al., 1989) is worldwide used to assess tics, and the second one, Yale-Brown Obsessive Compulsive Scale (Y-BOCS) (Goodman et al., 1989) is worldwide used to assess the obsessive-compulsive component. Global YGTSS scores are 0 to 100 with half points for the objective disease, and half point for the subjective disease i.e. social impairment. Y-BOCS results are included in a 0 to 40 range, 20 points are attributable for obsessions and 20 points for compulsions. In both scales, high results mean severe status of pathologies.

1) The first case is a OCTD 35 year old man (he is painter in a hospital), who at the beginning of the disease had several motor and phonic tics, later he manifested a severe addiction to cigarettes and an OCB towards his ex-wife as TS psychopathological diseases.

The patient's baseline global YGTSS scored 89/100, and his baseline Y-BOCS scored 31/40.

He was treated with: Habit Reversal Training -one year of weekly sessions- and pharmacological therapy,

both typical and atypical neuroleptics for tics, together with both selective serotonin reuptake inhibitors and tricyclic antidepressants for the obsessive-compulsive component (Roessner et al., 2011).

The patient had a poor compliance in following pharmacological and psychotherapeutic treatments, and he also had a poor response to them. Therefore, he underwent DBS with a thalamic bilateral target i.e. Voa-CM/Pf. After DBS, global YGTSS scored 69/100 and YBOCS scored 26/40, indicating a partial therapeutic effect of treatments. After the separation from his partner, he started persecuting her with threats and stalking because of his OCB; so the partner sued him. During the trial, the judge had not considered that he was suffering from TS and indicted him to six months of house detention. After the condemnation, he continued to display an erratic bizarre behavior associated with coprolalia and stalking. He asked for a second court hearing, during which the law's medical consultant proved that the specific conducts under accusation were depending on OCB and impulse control disorder. The patient did not have the control of his acts, consequently his behaviors could not be qualified as voluntary. This time the judge acquitted him considering his illness of TS.

2) The second case is a OCTD 38 year old woman with multiple tics, Non-Obscene Socially Inappropriate behaviors and obsessive-compulsive behaviors focused on personal hygiene as TS psychopathological diseases.

The patient's baseline global YGTSS was 88/100, her baseline Y-BOCS was 38/40. She had been treated with psychotherapy, pharmacological and surgical therapy (DBS). She took part in 8 months of Habit Reversal Training sessions with scarce results. She was prescribed typical and atypical neuroleptics, together with selective serotonin reuptake inhibitors and tricyclic antidepressants. Medications fatten the patient, who had to follow a diet but she failed because of her low compliance.

As she was still resistant to treatments, clinicians opted for DBS. Considering her DBS targets, she was first implanted in Voa-CM/Pf and Accumbens/ventral striatum. Then she had the removal of the device because of rejection; she was finally re-implanted in anterior GPI. DBS resulted in a reduction of both tics (global YGTSS decreased to 67/100)

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