

## Tourette Syndrome and Consciousness of Action

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### Abstract

**Background:** Tourette syndrome (TS) is a neuropsychiatric disorder characterized by the chronic presence of multiple motor tics and at least one vocal/phonic tic since childhood. Tics typically change and vary in both intensity and severity over time, with remission and exacerbation common. In the vast majority of patients, tic expression is characteristically accompanied by discomfiting bodily sensations, known as sensory phenomena or premonitory urges.

**Methods:** We reviewed the existing literature on premonitory urges associated with the sense of voluntariness of action in TS.

**Results:** Although the wish to move is perceived by the patient as involuntary, the decision to release the tic is often perceived by the patient as a voluntary capitulation to the subjective urge. Most patients with TS can exert a degree of control over the urge and constantly try to inhibit the movement. Based on these features, it has been suggested that tics performed in response to an urge to move should be classified as ‘unvoluntary’, as opposed to voluntary or involuntary acts. However, recent experimental data suggest that the brain areas involved in the generation of the wish to act show considerable overlap between healthy subjects and patients with TS.

**Discussion:** The simultaneous presence of both voluntary and involuntary aspects in the expression of tic symptoms by patients with TS is consistent with the hypothesis that tics can have the same neurophysiologic substrate as voluntary acts, even though they are misperceived as being involuntary. This reinforces the view of TS as a hyperkinetic movement disorder primarily affecting the conscious experience of action.

**Keywords:** Tourette syndrome, consciousness, tics, premonitory urges, voluntary movement, will, self-identity

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### Introduction

Tourette syndrome (TS) is a neurodevelopmental disorder affecting up to 1% of school-age children.<sup>1</sup> It is defined by the chronic presence of both multiple motor tics and one or more vocal/phonic tics.<sup>2</sup> According to *Diagnostic and Statistical Manual, 5th edition* (DSM-5) criteria, TS is a tic disorder characterized by an early onset before 18 years of age (typically 3–8 years for motor tics and 11 years for vocal tics) and that is not secondary to the administration of drugs known to cause motor side effects (e.g. stimulants) or to the presence of other disorders (e.g. Huntington’s chorea, postviral encephalitis).<sup>3</sup>

Tics are rapid movements or spasms of individual muscle groups (motor tics) or brief sounds resulting from the passage of air through the mouth, throat, and nose (vocal/phonic tics).<sup>4</sup> Common motor tics

include eye blinking, smirking, squinting, grimacing, nose twitching, and shoulder shrugging.<sup>5,6</sup> Vocal tics are more appropriately referred to as phonic tics as they do not necessarily involve the vocal chords, and include coughing, grunting, mumbling, throat clearing, and sniffing. Simple motor tics are quick and purposeless movements, which nonetheless can be distressing and painful.<sup>7</sup> Complex motor tics, by contrast, are slower and seemingly purposeful, so that they can be described in terms of deliberate actions (e.g. adjusting clothes, forced touching, echopraxia). Likewise, simple vocal/phonic tics are meaningless emissions of sounds or noises, while complex vocal/phonic tics involve the production of meaningful words and short sentences (e.g. palilalia, coprolalia).<sup>8</sup> Although complex tics are not included in the current diagnostic criteria for TS, they are not rare and were in fact

**Table 1.** Definitions and Descriptions of Different Types of Premonitory Urges

Term	Definition	Description
Sensory tic	Somatic sensation in the body, especially in bones, muscles, and joints, that leads the individual to perform voluntary movements to relieve the sensation.	Uncomfortable tactile, visceral, or musculoskeletal sensation that comes immediately before or accompanies the repetitive behavior. The individual is driven to repeat certain movements until he/she experiences a sense of relief.
Sensory phenomenon/ premonitory experience	Uncomfortable physical sensations in skin, muscles, joints, and other parts of the body that may be accompanied by perceptual stimuli (visual, auditory, tactile).	Itchy, tense, or tight sensation with a specific anatomic location, which leads to the feeling of wanting to release the repetitive behavior.
Just-right experience	A force, triggered by visual, auditory, or tactile perceptions, as well as a feeling of imperfection about actions and intention, that leads to the individual performing compulsive acts until the actions are felt by the individual to be complete.	A need to feel that objects look a certain 'just-right' way; that objects and people sound a certain 'just-right' way; or that objects and people have to be touched in a certain 'just-right' way.
Urge	A drive or impulse to perform the repetitive behavior in the absence of any obsession, worry, fear, or bodily sensation.	A need to perform repetitive actions that is not preceded by obsessions or sensory phenomena.

incorporated in Georges Gilles de la Tourette's original description (symptom triad of motor tics, echolalia, and coprolalia).<sup>9-11</sup>

The exact pathophysiology of tics is still unknown, although findings from neurophysiologic and neuroimaging studies suggest an underlying dysfunction within the cortico-striato-thalamo-cortical pathways.<sup>12</sup> Moreover, transcranial magnetic stimulation studies have indicated possible involvement of the primary motor cortex and Broca's area in the performance of repetitive behaviors, as well as abnormalities of intracortical inhibition that facilitate access of the sensory stimuli to motor responses.<sup>13</sup> This pathophysiological model is further supported by evidence that patients with TS can have deficits in sensorimotor gating as compared with healthy individuals.<sup>14</sup> In turn, these abnormal mechanisms, linked to the phenomenon of 'somatic hypersensitivity' or 'site sensitization,' seem to be supported by instances of sensory hypersensitivity that are frequently reported by patients with TS.<sup>15,16</sup> Moreover, tics can sometimes be induced by environmental stimuli such as a particular siren sound or specific word ('reflex tics').

In the next two sections, we review the classification of sensory phenomena and premonitory urges (PUs) and illustrate their relevance to the understanding of the full spectrum of TS symptoms. Subsequently, we discuss in detail the volitional and non-volitional components of tic production and the suggestion that TS might also affect the conscious experience of action.

### Sensory phenomena and premonitory urges

The symptomatology of tics in patients with TS is multiform: both motor and vocal/phonic tics frequently change and vary in intensity

and severity over time, with remissions and exacerbations common.<sup>17</sup> Moreover, various degrees of awareness and intentional control can accompany both tics and habits or behavioral routines, so that the two can be difficult to distinguish on clinical examination.<sup>18</sup> In fact, the vast majority of patients (>77% of those older than 13 years) experience discomforting bodily sensations immediately before the tics, known as sensory phenomena or PUs.<sup>19-23</sup> PUs, which are inner urges accompanied by unease or anxiety and relieved by the volitional capitulation to movement, are thought to be an involuntary and fundamental component of the subjective phenomenology of tic symptoms.<sup>22</sup> These experiences are also identified as sensory tics and have been reported to be focal or generalized intrusive sensations that drive the patient to gain relief through motor or vocal performances.<sup>24-26</sup> PUs are mostly localized to the eyes, palms, shoulders, and throat, with 40% of patients reporting these unpleasant experiences exclusively in the muscles and the remainder also localizing them to the joints and skin. Table 1 summarizes the definitions and descriptions of different types of PUs. Of note, premonitory sensations and PUs do not necessarily indicate the same phenomenon. The former are associated with uncomfortable physical sensations, while the latter can be linked to drives or impulses that are mental processes rather than sensory experiences.

As a whole, PUs are now recognized as a core symptom of TS and have been incorporated in clinical rating instruments such as the TS Diagnostic Confidence Index.<sup>27</sup> Moreover, the presence of PUs, along with tic severity and family history of TS, has recently been identified as an important childhood predictor of poorer health-related quality of life in adults with TS.<sup>28</sup> According to Leckman et al., patients become

aware of PUs at a mean of 10 years of age, with onset of the tic preceding awareness by an average of 3 years.<sup>21</sup> Data gathered with the Premonitory Urge for Tics Scale—a specific psychometric scale to measure the severity of PUs—confirm this latency of awareness, suggesting that patients become aware of PUs through a maturational process that is largely independent of tic onset.<sup>29,30</sup> This process could be the sign of transition from simple sensory perceptions to fully fledged consciousness.<sup>31,32</sup> Thus, a refined degree of self-consciousness could help patients recognize their subjective symptoms and thereby improve their ability to suppress them, as is shown in behavioral techniques such as exposure and response prevention and habit-reversal therapy, which can be effective strategies for regulating tics.<sup>33–36</sup> The significance and function of PUs is not known. It has been suggested that they could be a manifestation of neural dysfunction below the threshold of tic production or the result of abnormal attention to sensory information.<sup>19</sup> Converging evidence supports the hypothesis of sensory-gating dysfunction, which may result in a disproportionate inflow of somatosensory information and, consequently, generate PUs by intensifying the activation of the supplementary motor area and specific corticostriatal circuits.<sup>37</sup>

### More than tics

TS is increasingly recognized as a complex disorder with a wide spectrum of tic-associated behavioral problems.<sup>38</sup> About 90% of patients with TS develop comorbid psychiatric conditions, obsessive-compulsive disorder and attention-deficit hyperactivity disorder (ADHD) being the most common.<sup>39</sup> Of note, these comorbid conditions have been shown to affect quality of life even more profoundly than tic severity in TS populations.<sup>40</sup> With regard to the relationship between obsessive-compulsive disorder and TS, the scientific literature suggests that the two conditions share some neurobiological substrates and that a degree of compulsivity appears to be intrinsic to TS, although the reported percentage of patients with TS who also fulfill diagnostic criteria for obsessive-compulsive disorder varies from 11% to 80%.<sup>41–43</sup> Interestingly, tic-related obsessive-compulsive symptoms tend to be related to counting (arithmomania), concerns for symmetry (resulting in evening-up behaviors), and ‘just-right’ experiences, in addition to checking rituals.<sup>44</sup> Interestingly, a recent study on adults with TS using the Premonitory Urge for Tics Scale showed a significant association between the severity of PUs and measures of compulsivity, as well as perceived tic severity.<sup>45</sup> With regard to ADHD and TS, the rate of comorbidity between the two conditions is considerably high, especially in young people: as many as 60–80% of patients with TS fulfill diagnostic criteria for ADHD.<sup>42,43</sup> Moreover, the clinical features of these conditions partially overlap. In fact, the exact boundary between these two neuropsychiatric disorders is often unclear. On the one hand, problems related to attention, impulsivity, and hyperactivity commonly precede the manifestation of tics; on the other, the ability of patients with TS to focus and sustain concentration may be profoundly affected by the constant effort to monitor their PUs and suppress tics.<sup>46–49</sup>

Other comorbid behavioral problems often reported by patients with TS are related to impulse control disorders, a heterogeneous set of conditions (including intermittent explosive disorder, self-injurious behavior, trichotillomania, impulsive-compulsive sexual behavior) where patients have considerable difficulty controlling their urges to perform rewarding behaviors.<sup>50</sup> Personality disorders are also more commonly reported in clinic populations with TS than in the general population.<sup>51</sup> The susceptibility for schizotypal features in patients with TS can contribute to the development of non-obscene socially inappropriate behaviors (e.g. making inappropriate comments or actions).<sup>52–54</sup> These behaviors are often preceded by specific urges, which patients try to suppress. Finally, certain difficulties in interpersonal interactions have been associated with underlying alterations in social cognition, possibly resulting in unwanted behaviors.<sup>55–58</sup>

Conflicting frames of mind also characterize the voluntary attempts to inhibit unwanted behaviors and their urges, which, by contrast, are perceived by patients as involuntary. These attempts are tightly linked to the generation of tics and other repetitive behaviors.<sup>59,60</sup> The next sections discuss the exact nature and the putative roles of these different components in the consciousness of action in patients with TS.

### Volitional and non-volitional components of tic behaviors

Traditionally, voluntary and involuntary behaviors have been kept separate. The common view is that voluntary and conscious actions are flexible and controlled, while involuntary and automatic actions are fast but inflexible (i.e. predetermined). The former have often been associated with the activation of specific areas in the frontal cortex and use of perceptual information to guide goal-directed conscious behavior,<sup>37</sup> whereas the latter have been more closely related to subcortical pathways that are automatically triggered by perceptual stimuli.<sup>61,62</sup>

One well-studied example is the visual grasp reflex, which occurs whenever a visual stimulus suddenly triggers a quick eye movement (i.e. a saccade); this reflex is so automatic that it occurs even when the individual tries to look at something else.<sup>63</sup> Therefore, perceptual processing can easily cause motor activity and, at the same time, interfere with the execution of other responses. Moreover, this interference is not confined to the context in which reflexive behavior is elicited, but can also occur during the execution of voluntary actions whenever various potential stimuli in the environment grab the individual's attention, thereby subconsciously eliciting specific action schemata.<sup>64</sup> Indeed, it has been suggested that such automatic activation may be part of the process that leads to the voluntary behavior.<sup>34</sup> Counterintuitively, involuntary motor activity can be greater in intentional processes than in the activation of automatisms. In fact, an automatic response results from the activation of a specific sensorimotor connection that is dominant among others (e.g. the aforementioned visual grasp reflex), while in the presence of the volitional process several sensorimotor patterns compete to lead to a particular behavioral performance (e.g. the voluntary control of gaze

**Table 2.** Classification of Movements According to the Subjective Perception of Will

<b>Voluntary</b>	<b>Involuntary</b>	<b>Unvoluntary</b>
An action is voluntary when it is consciously performed, is flexible, and can be controlled. The perceptual information is used to guide goal-oriented behavior.	An action is involuntary when it is automatically performed and is inflexible. It is usually faster than a voluntary action. It cannot be controlled, because it is mechanically triggered by specific perceptual stimuli.	An action is unvoluntary when it is perceived as a voluntary response to an uncontrolled and involuntary urge to move.

during antisaccades). This model is also confirmed by the observation that the dorsal premotor cortex initially appears to represent multiple motor outputs in a choice situation context, in order to select the final action.<sup>65</sup>

In patients with TS, the distinction between voluntary and involuntary movements appears to be even more complex and clinically challenging. The sense of urge or the wish to move and feeling of uneasiness that leads to the repetitive behavior being performed is involuntary but, in contrast, the decision to release the tic is often perceived by the individual as voluntary.<sup>20</sup> In fact, it is possible for a patient with TS to exert a degree of control over the urge and try to inhibit the movement, even though the final release can be more intense in proportion to the length of the tic-suppression interval.<sup>66</sup> In light of this, it has been proposed that tics performed in response to a specific urge to move could be categorized as ‘unvoluntary’; that is, a third category of movement halfway between voluntary and involuntary behaviors.<sup>17</sup> Within this framework, a relevant feature of tics is that the urge appears to be an endogenous cue, anatomically linked to the movement. In a sense, the origin of the movement is completely involuntary (the urge), but the decision to move to relieve the unpleasant feeling is voluntarily taken (Table 2). It should be highlighted, however, that a subgroup of patients with TS (especially younger patients) do not report PUs. At present, it is unclear whether this indicates the actual absence of PUs for certain tics or the inability to recognize and verbally report them (in both cases as a possible consequence of brain developmental stages). Based on these cases, it is important not to conflate the dichotomies between voluntary versus involuntary and conscious (i.e. being aware, having ‘sense of agency’) versus unconscious.

A neuroanatomic model to clarify the brain mechanisms underlying the complex behavior of patients with TS experiencing the urge to tic has been proposed by Jackson et al.<sup>67</sup> The authors highlighted the overlap of sensory and motor neural circuits between urges for normal behaviors—such as swallowing, yawning, and micturition—and repetitive behaviors in TS. In particular, the shared functional brain-activation patterns seem to involve the limbic sensory and motor areas, as well as the insula and mid-cingulate cortex.<sup>68</sup> This functional network should be considered crucial for the motivation-for-action but not for the performance of an intentional action. In fact, the willed intention to perform a goal-directed movement would also require activation of both the premotor and parietal cortices. This model has the merit of identifying sound evidence for the involvement of

common neuroanatomic areas in a specific functional network for both normal and pathologic urges to move.

Despite these significant advances, unanswered questions remain. For example, it is not clear how the brain processes the transition between unconscious and conscious urges.<sup>69</sup> In particular, it is unclear how the model proposed by Jackson et al. could fit in the widespread network of regions that are known to be activated in conscious perception.<sup>70,71</sup> Moreover, it should be taken into account that an urge necessarily implies the possibility of voluntarily inhibiting the action (at least transiently).<sup>72</sup> In fact, if the urge did not make it possible to suppress or delay the action then the movement would be performed immediately. The commonly reported ability of patients with TS to delay the tic release implies that the urge to move precedes the conscious performance of the action, allowing conscious tic suppression. Therefore, a repetitive behavior that is urged by an involuntary feeling of discomfort, anxiety, and uneasiness, is perceived by patients with TS as not freely chosen, but it is nonetheless voluntarily performed insofar as the patient can decide to inhibit the action, thereby delaying the movement.

### The voluntary control of action

The picture of unvoluntary action in patients with TS outlined above remarkably resembles Benjamin Libet’s account of free will in his famous experiment in which healthy subjects were given the possibility of vetoing their actions.<sup>73</sup> Libet wanted to ascertain whether consciousness of action precedes or follows the *Bereitschaftspotential*, or readiness potential (RP), reported by Kornhuber and Deecke in 1965.<sup>74</sup> Libet’s experiment is well known and has been replicated several times with the same result: awareness of willingness to move follows activation of the premotor cortex and the supplementary motor area, which are the main contributors to the RP.<sup>75</sup> Libet also mentioned tic expression in patients with TS as an example of movement performed in the absence of conscious will.<sup>76</sup> He came to this conclusion because the pathologic tic behavior does not appear to be frequently preceded by the RP,<sup>77,78</sup> which Libet considered the hallmark of cerebral preparation to a willed action. It is worth noting, however, that the RP involves early and late components; patients with TS have been demonstrated to have the late component.<sup>79</sup> Furthermore, there is significant analogy between the ability of patients with TS to suppress their tics and the ability of healthy individuals to consciously veto their movements in the presence of a wish to act. In fact, both patients with TS and healthy subjects can inhibit the action that is driven by an urge to act, with the only

difference that the wish to act emerges involuntarily and automatically in the former, but voluntarily and consciously in the latter. Of note, however, the final motor output is described by both groups as voluntary, even though there is no RP in patients with TS.

The observation that the RP is not exclusively associated with voluntariness is supported by the finding that in healthy subjects at rest or engaged in mental tasks, unconscious movements are generally preceded by RP.<sup>80</sup> Thus, the RP probably signals the activation of a set of brain processes that are involved in causing the movement, independent of the interpretation of the movement as voluntary or involuntary. It has been suggested, therefore, that there could be two parallel pathways involved in the genesis of action: the first leading to movement execution, and the second to movement awareness.<sup>81</sup> The brain areas mostly involved in the experience of voluntary action appear to be the presupplementary motor area and the insula. The presupplementary motor area seems to be important for the cognitive features of action, such as the production of urge and a sense of agency.<sup>82–84</sup> The insula, in turn, may be particularly involved in the experience of agency while performing a movement, as there is evidence that activation of this region is more intense when subjects feel in control of their actions.<sup>85</sup> In addition, functional neuroimaging investigations have suggested that the insula is part of the brain networks responsible for tic generation.<sup>86</sup>

This picture seems consistent with the theory developed by Jackson et al., who have provided strong evidence that the brain areas involved in the generation of urges (i.e. limbic sensory and motor regions—insula and mid-cingulate cortex) are the same in healthy individuals and those with TS.<sup>67</sup> Of note, the chronic presence of PUs and the resulting tic expression and/or suppression have been associated with plastic remodeling at the level of frontostriatal circuitries and motor inhibitory pathways.<sup>87,88</sup> What remains unclear is the exact mechanism by which the urge to move in patients with TS can lead to impaired consciousness of action, whereby tics are perceived as a combination of voluntary and involuntary aspects. A possibility is to consider tics as equivalent to voluntary acts with regard to the physiologic underpinnings, even though they are mistakenly perceived as having an involuntary component.<sup>89</sup> In this case, TS might involve not only a movement disorder but also a pathologic conscious experience of action. Although this abnormal experience of conscious will does not account for the generation of tics, it could explain why patients with TS are sometimes unable to detect them in advance and thereby fail to suppress the stereotyped movements.<sup>31,90,91</sup> Interestingly, reduced awareness of self-initiated movements has been documented in other disorders characterized by hyperdopaminergic states, such as Huntington's disease and levodopa-induced dyskinesias.<sup>93,94</sup> However, it can be argued that the analogy between stereotyped movements in TS and pure choreic movements (such as those in Huntington's disease or levodopa-induced dyskinesias) might be inappropriate, given the total lack of volitional control in the latter movements.

Finally, intriguing similarities can be found between the hypothesis that tic generation involves brain processes associated with altered

consciousness of action and the hierarchical Bayesian formulation of brain function recently proposed by Edwards et al.<sup>94</sup> Although this model aims to account for the genesis of functional motor and sensory symptoms, rather than tics, abnormal activity of hierarchical Bayesian networks might be involved in the pathophysiology of different conditions associated with altered consciousness of the voluntary nature of movements.

## Conclusions

The process that brings about repetitive behavior in patients with TS is multifarious (e.g. tics are of different types, they can wax and wane) and complex rather than rigorously linear, as reflected by the multiple underlying neuronal networks possibly involved. Conscious and unconscious pathways appear to be strictly entangled. However, there is evidence of a close relationship between two modalities for movement generation: the first produces the actual movement, while the second allows conscious recognition of the authorship of the action. Patients with TS might have alterations in both pathways, thereby generating ambiguous judgments with regard to the actions performed as a consequence of their PUs. Beyond movement disorders, a similar model has been advocated to account for the altered consciousness of action in patients with schizophrenia, a psychiatric condition in which fragmentation between the execution of the action and a sense of agency (the recognition of ownership of one's own actions) is even more severe.<sup>95</sup> With the chronic presence of urges and tics and an enduring struggle to control their bodies, children with TS might also have to confront significant challenges to maintain a sense of a locus of control.

Future studies will clarify the complex interplay between motor execution and consciousness of action in patients with TS and healthy people. In turn, a better understanding of these mechanisms may help patients improve strategies for recognizing their subjective symptoms and maximizing tic control.<sup>34–36,96,97</sup> Finally, translational research might benefit from these advances as they open up new avenues for pharmacologic<sup>98–100</sup>—or even surgical<sup>101–104</sup>—interventions aimed at suppressing the urge to tic in patients with treatment-refractory conditions.

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