

# TICS STATUS: A MOVEMENT DISORDER EMERGENCY

*(Observations)*

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## Abstract

We describe a case of an 18-years-old boy with GTS, who developed severe, continuous, disabling tics after an arbitrary and abrupt withdrawal of haloperidol, tiapride and clonazepam therapy. This exacerbation considered as tics status lasted for weeks despite of intravenous re-administration of these drugs and interfered with sleeping; therefore, propofol and midazolam sedation, and later relaxation with artificial ventilation were applied for an immediate relief. Changing the medication to olanzapine and levetiracetam in parallel to sedation was effective to improve the motor symptoms to the baseline level. As far as the authors are aware, our case is the first published tics status case lasting for weeks and requiring sedation, intubation, and relaxation.

## Introduction

Guilles de la Tourette's syndrome (GTS) is a chronic complex neuropsychiatric disorder characterized by the combination of hyperkinetic movement disorders (especially tics) and psychiatric symptoms (e.g. obsessive-compulsive disorder –OCD and attention deficit hyperactive disorder –ADHD). Some case studies have described sudden exacerbation of GTS symptoms either due to drug withdrawal or acute infection resulting in a physically demanding state characterized by severe and permanent tics[10]. This condition shares some common symptoms with status dystonicus; therefore it might be considered as 'tics status'.

## Case report

The 18-years-old man was born in 1990. At the age of 7, stereotypic motor and vocal tics developed, and the diagnosis of GTS was made in 1998 in accordance with the Diagnostic and Statistical Manual 4th edition. Until 2003, he was on haloperidol monotherapy; however, later the combination of haloperidol, tiapride, clonazepam medication and hypnosis was needed for adequate symptomatic control.

Because the symptoms of GTS were well suppressed and only slight and non-disabling facial tics were observable, the patient arbitrarily and abruptly stopped his medication in 2008. Approximately two weeks later, he was admitted to the Department of Neurology because of subacute worsening in motor symptoms. Persistent and severe motor and vocal tics developed over a few days before admittance, which involved all of his extremities, trunk and neck. The motor tics were different from the pre-existing symptoms and dominated the clinical picture (**motor tics severity: 24/25, vocal tics severity: 16/25, impairment: 44/50 points** on Yale Global Tic Severity Rating Scale-YGTSRS) which was accompanied by relatively mild obsessive symptoms. Between tics,

we could not observe any sustained muscle contractions or abnormal postures; therefore the presence of tardive or drug-withdrawal dystonia was unlikely.

At the time of admission, the patient had slight fever (**37.5Celsius**), permanent sinus tachycardia (**120-130/min**), excessive sweating probably due to the physically demanding hyperkinetic symptoms. The level of creatine-kinase was elevated (**8000U/l**) most probably as a consequence of increased muscle activity and the numerous incidental injuries caused by the complex motor tics (e.g. the upper extremities involuntarily bumped into the furniture during sitting or laying on the bed).

Besides tics, there were no other neurological signs (e.g. dystonia, chorea, akathisia, or altered consciousness) present. Based on the otherwise normal blood chemistry and the clinical symptoms, the presence of neuroleptic malignant syndrome was also unlikely.

Bearing in mind that presumably drug-withdrawal might have provoked the subacute worsening, first intravenous tiapride, haloperidol and clonazepam was administered; however, without any symptomatic relief. Because the frequent and severe motor tics produced long-lasting extreme physical exhaustion, cardiovascular demand and pain interfering with sleeping, this condition was considered life-threatening by both intensive care specialists and neurologists. Because tetrabenazine is unavailable in Hungary, continuous intravenous propofol (up to 10 mg/kg/hour) and midazolam sedation (up to 0.3mg/kg/hour) was introduced on the second day after admittance for achieving immediate improvement[1]. During this light sedation tics were completely vanished; however, the temporary suspension of sedation resulted in reappearance of severe motor tics. After 3 days of light sedation, we decided to apply intubation, relaxation and deep sedation analogously to the treatment guideline of status dystonicus[7]. Because temporary suspension of sedation again resulted in reappearance of severe tics, we decided to change the haloperidol, tiapride and

clonazepam combination to olanzapine (10 mg bid) with the later augmentation by levetiracetam (1000 bid)[4]. After another five days of deep sedation, motor symptoms began to improve and subsequently sedation could be stopped.

The CK levels dramatically lowered during sedation and completely normalized within two weeks. With the combination of hypnosis, olanzapine and levetiracetam, the symptoms improved to the baseline level over the period of six months, which effect is still persistent over a one-and-a-half year follow-up period (**motor tics: 7/25, vocal tics: 3/25 points, impairment 8/50, YGTSRS**).

We obtained written consent from the patient to present his case in accordance to the Declaration of Helsinki.

## Discussion

Fahn commented on a rarely observed phenomenon in which motor and/or vocal tics become continuous over a period of several minutes, which he referred as 'tic status'[3]; however, the existence of this entity has been remained debatable in the literature. It is probably due to the lack of diagnostic criteria and the genuinely rare occurrence. Definition of tics status may vary by case-reports to case-reports. For example, Sachdev et al. suggested the following criteria[10]:

- Tics occur in one or more groups of muscles in a stereotyped and repetitive manner
- The episode is sustained for at least several minutes or hours
- During the episode, the patient is unable to suppress the tics, and they intrude into normal activities
- The episode is in clear consciousness.

Sachdev also noted that during tics status, the tics were different from the preexisting tics by being more severe, complex and long-lasting.

Probably the most important core feature of tics status is the continuously disabling tics and not the other behavioral problems of GTS. In our case, the hyperkinetic symptoms reached a point of severity such that incessant movements caused cardio-respiratory difficulties, autonomic signs, and indirect suggestions of incipient rhabdomyolysis. The whole picture resembled that of other severe hyperkinetic “storms”, e.g. status dystonicus. Therefore, to diagnose tics status, the presence of drug withdrawal or tardive movement disorders (e.g. dystonia, chorea), akathisia and hyperkinesias due to ADHD should also be excluded[5].

To clarify the definition of tics status, therefore, we propose diagnostic and exclusion criteria based on the previously published case studies and our experience (**Table 1**).

Several case studies describe tics status-like episodes; however, most of these exacerbations were not accompanied by systematic effects. In most cases, drug withdrawal (haloperidol[8, 10] or clonidine[6]), mycoplasma reinfection[2] and extreme stress by an assault[10] were identified as provoking factors. Because most case studies did not provide information about the autonomic symptoms (e.g. tachycardia) and laboratory findings (e.g. CK levels), we cannot ascertain whether these cases would have fulfilled our proposed criteria for tics status.

Tics status is considered an emergency situation by several centers[9]. There are no well-established guidelines available for the treatment of tics status.

One logical solution might be the re-administration of previously withdrawn drug (if drug-withdrawal is suspected as a provoking factor) or the application of antipsychotics in increased dosage. In our case the re-administration and dosage increase of base therapy (haloperidol, tiapride and clonazepam) was ineffective; however, this approach was efficacious in some cases[6, 8, 10]. Addition of

benzodiazepines (e.g. clonazepam) or switching to second generation antipsychotics might also be useful, if the increased dosage remains ineffective[10].

The treatment of long-lasting and disabling tics status requires a team approach including intensive care-, movement disorder specialists and psychiatrists. Because tics in our case were severe, permanent and physically demanding, our team considered this condition to be life-threatening. In similar situations light or deep sedation might be required, as well. Because in status dystonicus symptomatic improvement might occur after sedation[7], temporary suspension of sedation might also be tried to check for recovery from tics status. During sedation, one might change the antipsychotic medication (in our case to the combination of olanzapine and levetiracetam), which might also bring resolution.

Tics status represents an extremely rare disorder requiring an immediate and adequate treatment. As far as the authors are aware, our case is the first published case lasting for weeks and requiring sedation, intubation, and relaxation. We think that recognizing similarly severe tic exacerbations might be important to achieve immediate relief and avoid the potentially harmful consequences of physical exhaustion and increased cardiovascular demand.

**Table 1.** Clinical features of the proposed tics status.

<b>Suggestive features</b>	<b>Exclusion criteria</b>
<ul style="list-style-type: none"><li>• Pre-existence of GTS fulfilling the appropriate criteria of DSM-IV text revision.</li><li>• Acute/subacute worsening in the hyperkinetic features of the preexisting GTS usually preceded by acute infection or drug withdrawal/dosage reduction.</li><li>• The tics are different from the preexisting tics (e.g. more severe, complex and long-lasting)</li><li>• The hyperkinetic movements predominate the clinical picture resulting in severe systematic effects (e.g. cardio-respiratoric difficulties, autonomic signs or incipient rhabdomyolysis)</li><li>• The duration of continuous and disabling motor symptoms may vary from minutes to days (of note, shortest duration published was cc. 15 min).</li></ul>	<ul style="list-style-type: none"><li>• Presence of neuroleptic malignant syndrome</li><li>• Presence of altered consciousness</li><li>• Presence of acute akathisia</li><li>• Presence of other movement disorders (e.g. tardive or drug withdrawal dystonia or chorea)</li><li>• Presence of hyperkinetic movements due to OCD or ADHD</li><li>• Fulfillment of the criteria of malignant GTS[1]</li></ul>

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