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# A multifaceted approach to challenges in Huntington's disease

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## Abstract

Huntington's Disease patients often suffer from severe cognitive impairments and personality changes throughout the course of their disease. One of the areas patients display severe impairment is in their executive functioning, which is responsible for planning and multitasking, flexible thinking, and response control. Individuals with executive function impairment may become apathetic, irritable, disinhibited, and impulsive. Research has been conducted to assess the efficacy of pharmacological and nonpharmacological interventions in addressing these cognitive deficits. Our case report examines the application of both types of interventions in addressing behavioral dysregulation in a Huntington's disease patient, as well as the challenges these interventions may pose. This should serve as a reminder to always consider a multifaceted approach in order to provide individualized care to patients, especially those with multiple debilitating medical conditions.

## Keywords

Huntington's disease; nonpharmacologic intervention; executive function; hypersexuality; aripiprazole.

## Introduction

Huntington's disease is a neuropsychiatric degenerative disease characterized by motor dysfunction, cognitive decline, and behavioral/mood dysregulation [1]. The neuropsychiatric problems are often the most distressing to both patients and the care teams, requiring a multidisciplinary approach to address the complexity of symptoms. The behavioral symptoms in Huntington's disease are a result of changes in the frontal cortex of the brain and are attributed to neuronal degeneration in the caudate nucleus [2]. Nuclear imaging studies have found degeneration in the fronto-striatal circuits even prior to patients showing severe cognitive symptoms [3,4], and the degree of caudate atrophy correlates with degree of cognitive function [1,5]. It is crucial to understand that these symptoms can be further exacerbated by the onset of

loss of independence and depression as the disease progresses. Thus, early intervention is necessary to provide both symptomatic control and improvement in quality of life.

Current pharmacological interventions targeting behavioral dysregulation consist of the use of antipsychotics, mood stabilizers, and/or selective serotonin reuptake inhibitors. The antipsychotics aripiprazole and quetiapine have shown significant improvement in symptoms of both chorea and depression in Huntington's disease patients and are preferred as first line therapy in individuals with severe motor dysfunction [1,6]. Quetiapine specifically has been found to be effective in reducing aggressive behavior and impulsivity after just 12 weeks [7]. In addition to antipsychotics, mood stabilizers can be added as adjunctive therapy, due to their ability to promote cell proliferation and neurogenesis in the central nervous system [8]. A recent study found that the combination of lithium and valproate "alleviated movement deficits, suppressed anxiety- and depressive-like behaviors, improved motor-skill learning and coordination, and prolonged average survival" in mouse models with Huntington's disease [8]. Another study found that the combination of antipsychotic drugs plus divalproex was more effective in diminishing hostility than the treatment with an antipsychotic alone [9]. Due to their neuroprotective and stabilizing capabilities, mood stabilizers in addition to antipsychotics prove to be very therapeutic for treating Huntington's disease and also potentially slowing disease progression [10].

Additionally, the use of a nonpharmacologic approach has been encouraged when treating neuropsychiatric and movement dysfunction in Huntington's disease patients. Studies show that routine physical therapy consisting of graded aerobic exercises has reduced symptoms of chorea in Huntington's disease patients and improved mood in certain cases [11]. Practice guidelines should emphasize the importance of one-to-one interaction between patients and the care team to address physical and cognitive barriers present at that moment. Because Huntington's disease patients experience memory problems on a daily basis, providing a structured and calm environment can help ameliorate exacerbations of irritability, impulsivity, and apathy that are triggered by having to process new information [12]. It is also important for family members, nursing staff, and recreational therapists to use appropriate interactional styles to avoid direct confrontation with Huntington's disease patients. Rather than using restraints or attempting to touch a Huntington's disease patient during a behavioral outburst, using soft words and providing space gives patients an opportunity to practice impulse control, and minimizes risk of escalation of untoward behaviors. This allows health care members to form a more personalized relationship with patients, allowing the opportunity to provide more individualized care. This also allows patients the opportunity to maintain a certain amount of control and maintain dignity over their actions and life.

In this case report, we examine the effects of pharmacologic and nonpharmacologic interventions on improvement in behavioral regulation and impulsivity in a Huntington's patient. We used a psychiatric evaluation and Barkley Deficits in Executive Functioning Scale Short Form (BDEFS-SF) questionnaire before and after said interventions to assess this patient's executive functioning.

## Case Report

Mr. Y is a 30-year-old male with a 15-year history of Huntington's disease, methamphetamine use, mood and psychotic symptoms who presented to our facility with destructive, violent, and aggressive behavior after assaulting staff at his previous care facility, then stating that he wanted to kill himself.

At time of admission, Mr. Y displayed significant chorea and motor deficits, cognitive and memory impairments, as well as apathy and depressive symptoms. Mr. Y was started on duloxetine, valproic acid, and risperidone. Mr. Y completed the BDEFS-SF questionnaire and scored 72, which indicated severe impairment in executive functioning. Initially, Mr. Y interacted minimally with the care team, and did not attend any group classes. Recreational Therapy services were consulted, and recommendations of one-to-one therapy were provided to Mr. Y. The Speech Language Pathology (SLP) service was also consulted, and they taught breathing exercises and speaking in short paragraphs to improve Mr. Y's speech quality, which he found to be helpful over the course of this hospitalization. Recreational Therapy encouraged morning stretches and exercise to improve gait and balance. Due to ongoing symptoms of irritability, where Mr. Y would suddenly refuse to engage in therapeutic services, or spontaneously dump his meal tray onto the ground while cursing, Mr. Y was switched from risperidone to aripiprazole in order to improve symptoms of chorea and mood dysregulation simultaneously.

After two weeks on aripiprazole, which was up titrated to 15 milligrams per day, Mr. Y began showing increased energy, interest, and motivation in regard to activities of daily living (ADL). Mr. Y requested help with shaving, showering, and feeding; all activities he had previously neglected for the past several months. He began attending daily group activities and engaged in meaningful conversations with the care team, as well as unit staff. Mr. Y began to be seen ambulating around the unit with increasing frequency and would remind staff about his wish to engage in outdoor activities, to ensure he didn't miss outdoor activity. During these activities Mr. Y showed bright affect without behavioral issues.

During the third week with aripiprazole treatment, Mr. Y continued to display improved affect and mood. However, he also began to demonstrate increased impulsivity, as well as hypersexual behavior. Mr. Y began to spend hours in his bathroom masturbating, and on multiple occasions was verbally abusive towards staff and peers. His racial epithets and profanity caused a significant amount of emotional distress to staff, resulting in use of restraints on two separate occasions. In the days following these incidents, Mr. Y displayed shame and frustration in response to the emotional trauma he had endured. As a result, he was switched from aripiprazole to quetiapine. However, due to personal preference, Mr. Y requested that quetiapine be changed to olanzapine. Mr. Y experienced rapid resolution of his hypersexual and impulsive behavior after being switched to olanzapine. He continued his regimen of valproic acid, duloxetine, and olanzapine in addition to speech and recreational therapy throughout the remainder of the hospital course. A repeat BDEFS-SF questionnaire was administered at the end of four weeks, in which Mr. Y scored a 65.

## Discussion

It is important to acknowledge the challenges our care team faced as we managed this case of Hun-

tington's Disease. Cognitive impairment and motor dysfunction are some of the most distressing symptoms Mr. Y was experiencing. Pharmacologic treatment proved effective in managing these symptoms, but it was at the expense of increasing sexual impulsivity in Mr. Y. While increased hypersexuality with use of aripiprazole has been documented in prior psychiatric cases, it is not known which patients may develop this behavior. In fact, there are no known documented cases of aripiprazole induced hypersexuality in an individual with Huntington's disease. However, it is recommended that physicians be aware of this potential adverse effect [13]. In the case of Mr. Y, aripiprazole appeared to increase his libido, precipitating an uncontrollable urge to masturbate, which became an activity very distressing to other patients and staff, and proved to be very difficult to address. Care team members engaged in daily debriefing sessions with both nursing staff and Mr. Y, in order to minimize emotional distress to both. It is also important to note that by utilizing both speech therapy and recreation therapy services, Mr. Y significantly increased his engagement in various physical and mental activities in the unit. These activities improved the mood, motor, and cognitive capabilities of Mr. Y, evidenced by the relatively rapid improvement in BDFES-SF scores. During Mr. Y's hospital course, it became apparent that he had the greatest improvement in behavior regulation and cognitive functioning when nonpharmacologic interventions were used as an adjunctive therapy to pharmacologic treatment.

## Conclusion

It is important to remember that individuals with Huntington's disease experience a significant amount of loss in both independence and dignity throughout the course of their disease. These losses can exacerbate the neuropsychiatric symptoms they experience. This case report illustrates the importance of a multifactorial approach to treatment in order to address the debilitating symptoms Huntington's disease patients experience, while maintaining patient dignity. In addition to providing pharmacologic therapy, nonpharmacologic therapy should be utilized to address specific needs and formulate individualized care plans. Physicians should be encouraged to spend one-on-one time with patients so that individual needs can be recognized and provide resolution to obstacles that develop. Educating care team members on the challenges of managing Huntington's disease and teaching de-escalation and coping strategies to both staff and patients can help achieve a significant improvement in quality of life. Understanding the importance of a multifaceted approach to treatment of this difficult, degenerating illness allows medical professionals to provide individualized care, improve quality of life, and most importantly, maintain dignity of people that suffer from Huntington's disease.

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