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A Case Report of Response to Quetiapine Treatment in Neuropsychiatric Manifestations of Juvenile Huntington's Disease (JHD)

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ABSTRACT

Background and Objective: Juvenile Huntington's disease (JHD) represents a rare form of neurodegenerative genetic disorder characterized by potential neuropsychiatric symptoms. Within the broader spectrum of Huntington's disease (HD), JHD cases constitute a relatively small subset. The importance of understanding JHD lies in its distinct clinical presentation, particularly the emergence of primary psychiatric symptoms during different phases of the disease. The purpose of this case report is to highlight the clinical manifestation and treatment outcome of a JHD patient, focusing on mood and behavioral changes following the initiation of Quetiapine treatment.

Case Report: The patient is an 18-year-old woman presented with prominent neurological and psychological symptoms including mood symptoms, self-harm, and suicidal attempts. Neurocognitive assessment using the Montreal Cognitive Assessment Test revealed impairment with a score of 17 out of 30. The patient had mild neurocognitive impairment due to JHD. Sodium valproate was initially prescribed, and due to the presence of mood symptoms, irritability, and behavioral problems, quetiapine was added to the treatment regimen, which resulted in significant improvement in the patient's mood. The patient was discharged with Quetiapine 150 mg and Epilim 600 mg once a night. During the two-week follow-up, the patient showed significant improvement in mood regulation and self-harm behaviors after starting Quetiapine.

Conclusion: The results of this case report emphasize the effectiveness of Quetiapine in improving mood, suicide attempt, and behavioral symptoms in JHD.

Keywords: *Huntington's Disease, Suicide, Quetiapine.*

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Introduction

Juvenile Huntington's disease (JHD) is a rare neurodegenerative genetic condition with neuropsychiatric features. JHD accounts for 1% to 9.6% of all Huntington's disease (HD) cases (1). In 5% of cases, they manifest before the age of 21 (2). It is typified by a triad of neuropsychiatric symptoms, progressive movement disorder, and dementia (3). Approximately 30% of people with JHD exhibit psychiatric or behavioral issues before the onset of motor symptoms, and this proportion rises to 75% as the disease progresses (4). Behavioral problems and cognitive regression are common early signs in people with JHD, typically preceding motor symptomatology by many years, similar to the temporal sequence seen in adult-onset HD (5). In comparison to adult-onset HD, individuals with JHD, particularly those with large-size mutations with more than 80 CAG repeats, exhibit different features such as faster development of symptoms (6).

JHD takes a heavy toll on individuals and their families because of its rapid advancement and the significant care requirements it imposes (7). Family members and carers also highlight the disproportionately distressing condition due to the disruptive nature of these behavioral manifestations relative to the motor symptoms (5). In addition to behavioral abnormalities caused by HD pathology, patients frequently face increased socio-medical challenges as a result of having parents affected by the disease from an early age, as well as unstable family environments, which may include parental drug abuse or frequent partner changes (1).

Therefore, conducting well-designed research on JHD is critical to addressing existing gaps in knowledge and improve patient outcomes. By elucidating the underlying mechanisms of JHD and evaluating the efficacy of emerging therapeutic interventions, research endeavors aim to enhance the quality of life for individuals affected by this debilitating condition. This case report aims to emphasize the clinical presentation and treatment response of a patient with JHD, particularly focusing on mood, suicidality, and behavioral alterations after the commencement of Quetiapine.

Case Report

The Medical Research and Ethics Committee of the Ministry of Health Malaysia authorized this case report with the code RSCH ID-24-02113-4OT. The patient was an 18-year-old woman currently single and employed as a part-time salesgirl. She was initially presented due to self-harming behavior, which transpired following a disagreement with her mother. The incident unfolded after a verbal altercation, during which she requested her mother's phone, and her mother declined. This denial triggered anger in her, leading her to engage in destructive behavior in her home, culminating in the breaking of a window. In response to her actions, the patient's mother slapped her, further escalating the situation. She then resorted to using scissors to cut the sofa linen and scratched her left wrist, all transpiring in front of her mother. Despite attempts to intervene, the patient's mother could not prevent her, and in a more alarming turn, she threatened to jump out of the window. This prompted her mother to seek police assistance, resulting in the patient's admission to the hospital and subsequent placement in the psychiatric ward. Upon delving into the patient's medical history, it was revealed that she had experienced an accidental fall from a third-floor apartment three months before her current presentation. This incident resulted in cerebral concussion and polytrauma, although head CT scan showed normal findings with no intracranial bleeding. At that time, the patient received an

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evaluation from the neuromedical team, who tentatively diagnosed her with JHD. This diagnosis was based on the presence of chorea movements persisting for over a year and a robust family history of HD, which included her paternal grandmother, four paternal siblings, and her father. Following her initial treatment and recovery, she defaulted subsequent appointments until she reemerged at the center, now exhibiting self-harming behavior.

She has exhibited behavioral issues since her secondary school years, characterized by deceitfulness and stealing. She displayed heightened irritability with deficient impulse control, frequently resorting to self-harm by cutting her wrist with a knife during distressful moments. Additionally, she demonstrated forgetfulness, poor judgment and engaged in frequent quarrels with her mother over trivial matters. Despite these behavioral concerns, she denied experiencing persistent symptoms indicative of major depressive disorder, manic or hypomanic episodes, or psychotic symptoms in the past.

After a one-week stay, she was discharged from the ward. However, she was readmitted a week later due to disruptive and suicidal behavior. During this episode, she appropriated her maternal aunt's phone, asserting it was hers, leading to a confrontation with her maternal uncles and aunts that resulted in physical assault. She exhibited impaired judgment, attempting to jump from her apartment building, but she abandoned the plan after being comforted by a cousin. In light of her behavioral problems and irritable mood, the patient's mother, feeling overwhelmed, expressed a desire to send her to a rehabilitation center. This distress is further compounded by her responsibility for caring for her husband, who is currently in an advanced stage of HD.

The physical examination revealed chorea movements observed over the left shoulder. In the mental state examination, irritability was evident, while there were no apparent thought or perceptual disturbances. The neurocognitive assessment using the Montreal Cognitive Assessment (MOCA) indicated impairment, with a score of 17 out of 30. She was diagnosed with mild neurocognitive disorder due to JHD. Initially, during the first admission, a mood stabilizer, Tab. Sodium Valproate was initiated to address impulsivity. However, upon re-admission, considering the presence of mood symptoms, irritability, behavioral issues, and cognitive impairment, Tab. Quetiapine was added to the treatment regimen. Subsequently, there was a notable improvement in terms of irritable mood and behavioral issues, leading to her successful discharge under the care of her mother. The discharge medications included Tab. Quetiapine IR 150mg once nightly, and Tab. Epilim 600mg once nightly. During her follow-up outpatient appointment (two weeks after discharge from the ward), her behavior was reported to be more manageable, and there were no further instances of self-harming behavior. After six months post-discharge, she was reported to be well with no more challenging and suicidal behavior.

Discussion

It becomes evident that patients with JHD are often presented with a broad array of manifestations, including but not limited to, violence, aggression, oppositional conduct, fixation, despondency, anxiousness, impulsivity, attentional deficits, psychotic episodes, and substance misuse (5). When compared to previously published cases, this patient demonstrated improvement in mood symptoms and suicidality following the initiation of Quetiapine.

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The treatment of psychiatric comorbidities in JHD remains challenging, with a lack of well-established characterization and definitive best practice recommendations (8). The provision of multidisciplinary care stands as a cornerstone in the endeavor to optimize the quality of life for individuals afflicted with HD. This holistic approach encompasses an array of interventions, ranging from home remedies to non-pharmacological modalities, targeting the mitigation of behavioral and psychological symptoms. Effective management options focus on alleviating symptoms and promoting general well-being, emphasizing the critical role of holistic care paradigms in mitigating the negative impact of this challenging disease (9).

Presently, therapeutic interventions for HD primarily focus on symptom relief, predominantly addressing neurobehavioral manifestations and chorea, although robust trial evidence supporting their efficacy remains somewhat sparse (3). For cases characterized by refractory depression, the utilization of atypical antipsychotics such as olanzapine, risperidone, aripiprazole, or clozapine has demonstrated efficacy, as evidenced by case reports and small open-label case series. Pharmacological management of irritability typically involves selective serotonin reuptake inhibitors (SSRIs) such as fluoxetine and sertraline as first-line therapy, with clomipramine and buspirone representing second-line alternatives (10). In this case, the initiation of Quetiapine and Sertraline led to an improvement in behavior, which was a significant finding. However, it's essential to acknowledge the complexities involved in the management of JHD, including considerations such as maintaining ideal body weight and managing the risk of seizures associated with antipsychotic medication (11, 12). Therefore, it is paramount to weigh the benefits and risks associated with the medication to make informed decisions regarding its continuation or modification (3).

This case underscores the importance of individualized treatment approaches and highlights the ongoing need for further research to enhance our understanding and management strategies for JHD. Moreover, the patient and caregiver’s understanding of JHD symptomatology and treatment are crucial components in optimizing patient care and maximizing the quality of life for individuals with JHD. Despite the current limitations in treatment options, the support of family and the community remains instrumental in addressing the multifaceted challenges posed by JHD (13).

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