

# Personality Changes in a Patient with Wilson's Disease

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## ABSTRACT

Personality changes in a patient with Wilson's disease

Wilson's disease (WD) is a rare disorder that primarily affects liver and brain, with the onset frequently in adolescence. Psychiatric symptoms are remarkable and the first manifestation in most cases. WD, when first presented with psychiatric symptoms, delay in the diagnosis can often occurs. This study describes a case of WD starting with tremor and depression and developing permanent personality and behavioral changes later.

**Key words:** Personality change, psychiatric symptom, Wilson's disease

## ÖZET

Wilson hastalığına bağlı kişilik değişikliği olgusu

Wilson Hastalığı (WH), karaciğer ve beyni tutan, sıklıkla adolesan çağda başlayan, nadir görülen bir hastalıktır. WH'nin psikiyatrik belirti ve bulguları da oldukça dikkat çekicidir ve birçok olguda başlangıç belirtisi olarak ortaya çıkar. WH psikiyatrik belirtilerle ortaya çıktığında, tanı gecikmesi yaşanması sıkır. Bu yazıda, tremor ve depresyonla başlayan, sonrasında kalıcı kişilik ve davranış değişikliklerinin geliştiği bir WH olgusu sunulmuştur.

**Anahtar kelimeler:** Kişilik değişikliği, psikiyatrik semptom, Wilson hastalığı



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## INTRODUCTION

Wilson's disease (WD) is a rarely seen genetic disease which frequently starts in adolescence period and primarily affects liver and brain (1). WD is related with copper accumulation and occurs in 1-2 per 100000 people. Most of the patients present with liver symptoms during childhood ranging from asymptomatic liver disease to cirrhosis and hepatic failure (2). In WD, a decrease is seen in incorporation of copper with ceruloplasmin, which transport copper, due to mutation in ATP7B gene localized to chromosome 13 (3). Therefore, copper cannot be excreted via bile and accumulates in liver at first and then in brain, kidneys and cornea (4). Extent of neuronal damage in the brain is highly variable including structural abnormalities in striatum and atrophy in the brain (5).

About 40-50% of the people with WD present with neurological and neuropsychiatric symptoms (6). These neurological symptoms are akinetic-rigid syndrome like Parkinson disease, tremor of the pseudosclerosis, ataxia and dystonic syndrome (7). Other frequently seen

neurological symptoms are motor coordination loss, dysarthria and spasticity. Migraine, insomnia and seizures are also reported (8).

Besides neurological symptoms, psychiatric symptoms are also remarkable in WD (9). Wilson, in his original article in 1912, reported permanent psychiatric symptoms in 8 of the 12 cases of WD (5). Psychiatric manifestations of WD are personality changes, affective fluctuations, psychosis and cognitive impairments (10). Personality changes, particularly irritability and aggression are mostly seen symptoms among these psychiatric manifestations (45.9%) and secondly depression (27%) (5).

In this study we report a case of WD starting with neurological symptoms and depression early in the second decade, then developing permanent personality and behavioral changes later.

## CASE

Ç.Ç. was a 27 year-old male patient who interrupted his university education (physics). He was unemployed

and living with his parents in Istanbul. He was brought to our psychiatric emergency department by his family due to suicide attempt by jumping in a sewer as a result of having a heavy argument with his family.

His complaints had begun six years ago, when he was in his first year of university education, with tremor in hands, ataxia, demoralization. He had been treated with an antidepressant drug but his symptoms had not improved. He had returned to live with his family after he had left school. He had been diagnosed as WD as a result of investigations and he had no symptoms other than tremor in his hands at that moment.

Seven months ago he had a suicide attempt with drugs of WD including trientine and zinc, due to the thought that he would never recover, but he did not apply to hospital. After ten days, he thought his body could not excrete those drugs, drugs were circulating in his blood and he would never get rid of this situation, so he had another suicide attempt with the same drugs. However, he stated that nothing happened to him other than nausea and vomiting. After six months, he once again attempted suicide taking drugs for gastric complaint and breathing in stove gas in the kitchen. Then, he accepted inpatient treatment in a psychiatry clinic because of insistence of his family and that was his first admission to a psychiatrist. Amisulpirid 400 mg per day treatment was started but this drug was stopped by internal medicine specialist due to a mild increment of liver function tests. His irritability and aggression increased for seven months. According to the family, he decreased self care, he did not take bath and he damaged household goods when he was angry. He never got out of his room. He began to masturbate more often. His family said that before WD, he had been a very good child and they never had any problem with him.

After diagnosis of WD he had problems especially with his mother and could not stand to hear his mother's voice. He claimed that his mother was speaking all the time, created many problems to his father and in fact never wanted his father. The day before, while he was arguing with his mother, his father interfered. He wanted to show to the other people how much he suffered, then he jumped to a

water-channel and he thought that the media would learn this situation. He could not reach to his goal and returned home.

His physical examination was normal. On his neurological examination sensorial, motor, cerebellar system and deep tendon reflexes were normal. There were tremor and mild rigidity in his upper extremities. Coreoathetoid movements were not seen. Kayser-Fleischer ring was not detected on the examination of the eyes.

At psychiatric examination, he was in his chronological age with normal eye contact and his self-care was decreased slightly. He was speaking in detail and was explaining the events theatrically. He behaved in a puerile manner. His affect was normal and mood was dysphoric. His associations were normal and directed to the goal. His thoughts were often about the conflicts with his family. He had no active suicide ideation. There was no perception disorder. He had insight about his disorder. His mother had depression and conversion disorder. He had no history of substance use except smoking one cigarette package in a day and rarely drinking alcohol. His biochemical, hepatitis, syphilis, HIV, urine substance metabolites and thyroid function tests were normal. In his hemogram trombocyte level was 65.000/mm<sup>3</sup> and he was under control of hematology. Hamilton depression scale score was 11. In Rorschach test psychotic symptoms were not detected, but histrionic and somatic mechanisms were seen to be used. According to the neuropsychological test, cognitive functions and memory were sufficient other than mild deficiency in attention and ability to sustain attention. Histrionic, borderline personality characteristics were prominent in assessment with structured clinical interview for DSM IV disorders-II (SCID-II). Any axis-1 disorder was not detected, whereas he was diagnosed as B class personality changes due to his general medical disorder (WD). He was treated with 1 mg/d of risperidone. His compliance was good at the hospital. He was sent home three times with permission. He lived conflicts with his family when he was at home. His symptoms were partially improved and he was discharged from the hospital to be followed up as outpatients.

## DISCUSSION

There is not a specific test for diagnosis of WD. Since the symptoms are not specific and different organ systems are affected, it is complicated to make differential diagnosis in WD. It is easy to diagnose WD if there are neurological symptoms and Kayser-Fleischer ring. WD should be considered if there is liver disease of unknown cause and incipient movement disorders.

Movement disorders are dominant in neurologic manifestation of WD and dystonia, tremor in the extremities, koreoathetoid movements, rigidity, disarthria and ataxia are often seen (6). Before these characteristic neurological symptoms, insidious symptoms including behavioral changes, success deterioration in school, and difficulty in performing activities which require fine motor coordination may occur. Deterioration of handwriting and micrography may develop (11).

Kayser-Fleischer ring was not detected in our patient. Tremor was the first neurological symptom that caused patient to call off his university education. Before diagnosis of WD, behavioral changes, decline in school success and deterioration in fine motor movement disorders had developed in our case which was consistent with the literature. Liver function tests were normal. Although liver function tests were normal in our case, deterioration in liver function tests is frequently seen in patients with WD who have central nervous

system involvement. However, these patients are frequently asymptomatic (6).

Depression, movement disorders such as tremor and ataxia facilitated the diagnosis of WD in our patient five years ago. Therefore, there was no need to examine patient with diagnostic tools. However, neuropsychometric tests and SCID-II diagnostic tests were applied to the patient due to personality changes lasting for one year.

Onset of symptoms in WD usually occurs in the second or third decade (6,10). Psychiatric and behavioral abnormalities may be seen in 30 to 100% of the patients and these are initial symptoms in two thirds of cases (12-14). Personality changes and aggression are the most common symptoms and are followed by depression. Anxiety, cognitive changes, psychosis and catatonia are rarely seen (12,13).

In our case starting age was 20 consistent with the literature. The disease presented with psychiatric symptoms like depression and dysphoric mood but his symptoms were not relieved with antidepressant treatment. These psychiatric findings were personality changes and depression, which were reported as the most frequent psychiatric symptoms in the literature. WD patients frequently apply to a psychiatrist (10). If WD presented with psychiatric symptoms, the diagnosis often delays (5). Although WD is a rare disorder, when there are rapidly starting depressive symptoms, personality changes and tremor; it is important to keep WD in mind (10).

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