

# A rare case of Fahr's syndrome with dissociative amnesia

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## Abstract

**Background:** Fahr's syndrome (FS) is a movement and psychiatric disorder characterized by symmetrical and bilateral calcification of the basal ganglia. Dissociative amnesia is a rare psychiatric disorder that occurs after stressful events and is classified as a dissociative disorder. In addition, dissociation is the process by which the mind conceals stressful events from consciousness. Several neuropsychiatric manifestations in FS have been reported. However, due to their rarity, FS and dissociative amnesia can be misdiagnosed.

**Case:** A 43-year-old female presented with Parkinsonian symptoms and an inability to recall important personal information. FS and dissociative amnesia were diagnosed after a thorough medical, psychiatric, and diagnostic investigation. Subsequently, correction of calcium and phosphate levels by  $\alpha$ -hydroxy-vitamin-73 D3 and psychotherapy were administered for two-months until there were no neuropsychiatric symptoms reported.

**Conclusion:** This research highlights a rare case of FS with dissociative amnesia which pathophysiology is not completely understood due to limited research.

**Keywords:** basal ganglia calcification; dissociative disorders; fahr's syndrome; hypoparathyroid; movement disorders.

## Introduction

Fahr's disease (FD) is a neurodegenerative disease characterized by abnormal symmetrical and bilateral calcified deposits in the basal ganglia and cerebral cortex which causes extrapyramidal symptoms (Lee et al., 2018). Its various symptoms, ranging from extrapyramidal to psychiatric abnormalities have been reported, with Parkinsonism being the most common (Abubakar & Saidu, 2012; Ooi et al., 2019). It affects young to middle-aged adults with a prevalence of <1/1,000,000 worldwide (Saleem et al., 2013). The presence of parathyroid endocrinopathies is termed Fahr's syndrome (FS) (Perugula & Lippmann, 2016).

FS with depression, delusional-manic disorder, dementia, flattening of affectivity, hypochondria, and schizophrenia was reported in previous research (Calabrò et al., 2014; El Hechmi et al., 2014; Naqvi et al., 2017; Roiter et al., 2016; Samuels et al., 2018; Savino et al., 2016). Furthermore, parathyroid endocrinopathies and Parkinsonism can be pathognomonic, but FS has no pathognomonic psychiatric symptoms. In several cases, new and unexpected psychiatric disorders have been reported (Asokan et al., 2013). In this case, a 43-year-old female was presented with FS and an inability to recall important autobiographical information but no prior evidence of significant cognitive decline indicating unreported and unexpected psychiatric disorders of dissociative amnesia.

## OPEN ACCESS

### Jurnal Keperawatan Padjadjaran (JKP)

Volume 10(3), 224-227  
© The Author(s) 2022  
<https://doi.org/10.24198/jkp.v10i3.2134>

#### Article Info

Received : October 05, 2022  
Revised : December 24, 2022  
Accepted : December 26, 2022  
Published : December 31, 2022

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#### Citation

Muhammad, F., Arpin, W., Pratama, Y.S., Lestari, C.H., Budianto, P., Danuaji, R., Hutabarat, E.A.J., Putra, S.E., & Hafizhan, M. (2022). A rare case of Fahr's syndrome with dissociative amnesia. *Jurnal Keperawatan Padjadjaran*, 10(3), 224-227. <https://doi.org/10.24198/jkp.v10i3.2134>

#### Website

<http://jkp.fkep.unpad.ac.id/index.php/jkp>

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E-ISSN: 2442-7276

P-ISSN: 2338-5324

Muhammad,F., et al. (2022)

**Case Presentation**

A 43-year-old female was presented to a hospital with resting tremors and gait disturbance for two years. The companion disclosed that the patient had a radical thyroidectomy 20 years ago and occasionally wanders away from home and does not recall self-identity. These occurrences became apparent after the passing of the patient’s husband four years ago. The symptoms were causing significant social impairment, and if received condolences, the patient would relapse. The amnesia is retrograde and occurs abruptly. The patient is unaware of the amnesia and requires five minutes to recall all of personal information. Hallucinations, delusions, or mood disturbances have also not been experienced. There was no history of alcohol consumption, medication abuse, drug abuse, seizures, head injury, metabolic, systemic diseases, and neuropsychiatric disorders in the family. The patient’s mini-mental state examination (MMSE) score was 29 out of 30 indicating no mild cognitive impairment was found and dissociative amnesia was diagnosed based on the signs, symptoms, and findings.

On observation, the resting tremor was pin-rolling and had a frequency of approximately 4 to 6 Hz. The patient struggled to get out of a chair and stood with a flexed posture. The gait shown was characterized by small-shuffling steps with a retained arm swing. A structural brain organic lesion and metabolic abnormalities due to the radical thyroidectomy were suspected. The results of a blood diagnostic workup showed 9,640/ $\mu$ l leukocyte, 12.2 g/dl hemoglobin, 119 mg/dl blood sugar, 35 mg/dl urea, 1.13 mg/dl creatinine, low parathyroid hormone (PTH) (7.9 pg/ml), low calcium (0.57 mmol/l), 3.27 mmol/l potassium, 144.14 mmol/l sodium, and 102.87 mmol/l chloride. As shown in Figure 1, the computed tomography (CT) scan revealed bilateral symmetrical large areas of calcification over the corona radiata, basal ganglia, and dentate nucleus, which was suggestive of FS. FS secondary to

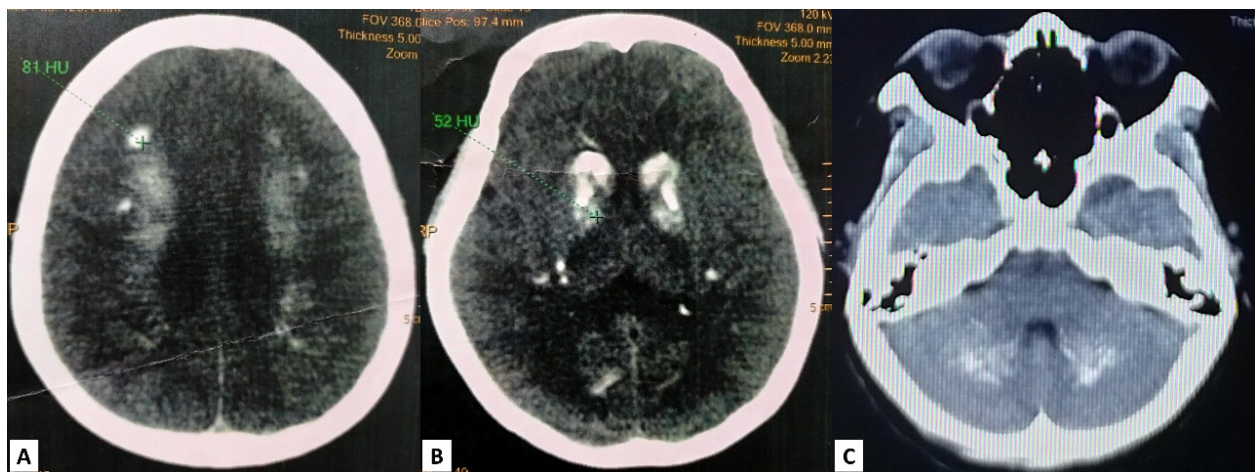
hypoparathyroidism was diagnosed in the absence of autosomal dominant inheritance in the family history.

**Results**

The treatment was focused on the patient’s specific pathology as well as symptomatic therapy. The movement disorders resolved with the administration of  $\alpha$ -hydroxy-vitamin-D3 5000 IU o.d. and corticosteroid dexamethasone 0.5 mg o.d. for two months. Furthermore, we implemented novel approach of supportive ventilation psychotherapy for two months or more as needed. This psychotherapy provides the widest possible opportunity for the patient to express what is in his heart so that he feels relieved and his complaints are reduced. Therapist attitude is to be a good listener and full of understanding. Topic of discussion is problems that become the main stress. Psychotherapy, hypnosis, family and group therapies were also used to treat dissociative amnesia. This was suggested so that the patient feels safe, and the companion was educated on the need to reacquaint her with personal memoirs and belongings. In addition, re-traumatization factors should be discussed with the family or relatives, as well as the importance of avoiding them. During the two-month post-treatment follow-up, the patient was still cooperative by showing good mental and emotional adaptation during ventilation psychotherapy, and her family, also reported that no amnesic symptoms. Furthermore, the post-treatment MMSE score was 29 showing no decline and indicating her cognitive remained alert and good.

**Discussion**

The diagnosis of FS must be distinguished from FD because of the differences in prognosis and



**Figure 1. Axial non-contrast CT-scan depicts bilateral symmetric calcifications in corona radiata (A), basal ganglia (B), and both dentate nuclei (C).**

treatment (Perugula & Lippmann, 2016). The age of onset in the fourth or fifth decade, a family history consistent with autosomal dominant inheritance, and the absence of metabolic systemic disorders are all diagnostic criteria for FD. FS should be considered in the presence of metabolic or other systemic disorders that could be explained as an underlying cause of calcification. The presence of metabolic or other systemic disorders that could be explained as an underlying cause of calcification should be considered as FS (Perugula & Lippmann, 2016). FD requires only symptomatic therapies, however, FS should be treated with symptomatic therapies directed at the underlying primary causes (Saleem et al., 2013).

The neurological manifestations of Parkinsonism are usually found in FS (Grover et al., 2011). The other common neurological signs and symptoms include headache, vertigo, stroke-like events, seizures, syncope, spasticity, dysarthria, chorea, dystonia, and myoclonus (Savino et al., 2016). Symptomatic or asymptomatic bilateral symmetric calcification can occur in various conditions. Therefore, clinicians should carefully exclude other potential diagnoses because the size of calcification affects manifestation, particularly dementia and extrapyramidal symptoms (Saleem et al., 2013).

Psychiatric disorders are common in FS, with 40% of patients presenting them as their initial symptom (Saleem et al., 2013). According to previous research, mood disorder, psychotic symptoms, and cognitive disturbance are the most common features (Calabrò et al., 2014; El Hechmi et al., 2014; Naqvi et al., 2017; Roiter et al., 2016). An incidental finding of FD in a hypochondriac patient was also reported in previous research (Samuels et al., 2018). In addition, there was an unusual and previously unreported presentation of fugue, retrograde memory loss, and autobiography amnesia in our case. The patient's amnesia was examined to see if it could be attributed to the calcification. The coincidental onset of amnesia following the patient's family loss, as well as the typical FS symptoms, increased the diagnostic difficulty. However, the coexistence of organic and psychological factors is common in amnesic patients.

Previous research recommended the identification of structural calcinosis-causing amnesia in the anatomical structure of the memory processing area (Lucchelli & Spinnler, 2002). The calcification of the patient's brain did not involve the declarative memory processing area, such as the hippocampus, medial temporal lobe, or diencephalon (Engelhardt, 2020). There has been an established relationship between brain calcification and psychiatric symptoms. The presence of brain calcification and progressive subarachnoid space dilatation is associated with widespread psychiatric symptoms (Lauterbach et al., 1998). Moreover, the stress trigger of her husband's death was believed to be the cause of neurotic psychiatric disorders such as dissociative disorders.

Movement disorders in PTH-related FS can be resolved by correcting the calcium and phosphate levels (Abubakar & Saidu, 2012). Corticosteroid therapy also reversed the neurological deficits (Saleem et al., 2013). Meanwhile, levodopa efficacy in FS with Parkinsonism is reportedly poor (Asokan et al., 2013). Psychotherapy has become the treatment of choice since no evidence-based treatments for dissociative amnesia are available. Medication can also be used to alleviate symptoms of anxiety and depression (Staniloiu & Markowitsch, 2014). However, lithium was not administered because it may increase the risk of seizures in the patient (Saleem et al., 2013).

## Conclusions

We conclude that dissociative amnesia is a rare presentation of FS. Although the brain calcification did not affect the memory-processing area in this case, the history of family loss is a known cause of dissociative amnesia. There are no known cases in which these diagnoses coexisted. Therefore, further research is recommended to explore how brain calcinosis causes dissociative amnesia. We recommend the use of  $\alpha$ -hydroxy-vitamin-D3, dexamethasone and supportive ventilation psychotherapy to alleviate the hypocalcemia-caused FS symptoms and dissociative disorders, respectively.

## Declaration of Interest

All authors declare that they have no conflict of interests.

## Acknowledgment

The authors are grateful to all neurologists and psychiatrists of Dr. Moewardi General Hospital and UNS Teaching Hospital for providing feedback and technical support.

## Funding

No financial support or grant was received.

## Data Availability

Available on reasonable request from the corresponding author's email [faizal9m@student.uns.ac.id](mailto:faizal9m@student.uns.ac.id).

## Ethics statement

Written informed consent was obtained from the patient. The copies of the written consent and ethical approval (Health Research Ethics Committee of Dr. Moewardi General Hospital No.221/HREC/2022) are available for review by the Editor-in-Chief of this journal.

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