Teaching Video NeuroImages: Wilson Disease Presenting With Catatonia

Roshni Abee Patel, MD, and Meagan Bailey, MD, MS

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Correspondence

Dr. Patel
roshnipatel1988@gmail.com

A 29-year-old man with a history of depression on fluoxetine presented with 4 months of progressive abdominal pain and hypoactive encephalopathy. On examination, he was non-responsive, bradykinetic, spastic, and had catatonia with waxy flexibility (video 1). Bush-Francis Catatonia Rating Scale score was 28 (table). He had new liver cirrhosis; serum ceruloplasmin was low and 24-hour urine copper was elevated. MRI revealed abnormal signal in basal ganglia and pons (figure). Two heterozygous mutations in the *ATP7B* gene (c.1934T > G and c.207 G > T) confirmed the diagnosis of Wilson disease. Zinc and penicillamine were initiated for copper reduction; catatonia did not respond to benzodiazepines. He was discharged home but required full care from family (nonambulatory, required help with all activities of daily living), and neurologic status was unchanged 3 months after discharge. Catatonia is a rare and underappreciated manifestation in Wilson disease¹; early recognition is essential for prompt initiation of chelation therapy.²

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Disclosure

The authors report no disclosures relevant to the manuscript. Go to Neurology.org/N for full disclosures.

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Video

Table Bush-Francis Catatonia Rating Scale With Subscores

Signs and symptoms	Patient score ^a
Excitement	0
Immobility/stupor	2
Mutism	3
Staring	3
Posturing/cataplexy	3
Grimacing	3
Echopraxia/echolalia	0
Stereotypy	0
Mannerisms	0
Verbigeration	1
Rigidity	2
Negativism	0
Waxy flexibility	3
Withdrawal	2
Impulsivity	0
Automatic obedience	1
Mitgehen	0
Gegenhalten	0
Ambitendency	3
Grasp reflex	0
Perseveration	3
Combateness	1
Autonomic abnormality	1
Total score	28

^a All items are rated on scale of 0–3.

Figure MRI Fluid-Attenuated Inversion Recovery

Hyperintensities seen in bilateral caudate and lentiform nuclei (A) and dorsal pons (B).

Appendix Authors

Name	Location	Contribution
Roshni Patel, MD	Rush University Medical Center, Chicago, IL	Designed and conceptualized study, major role in acquisition of data, drafted the manuscript for intellectual content
Meagan Bailey, MD, MS	Rush University Medical Center, Chicago, IL	Revised the manuscript for intellectual content

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