

disorder. The neurologic signs and symptoms reflect the predilection for basal ganglia (e.g., caudate, putamen) involvement in the brains of affected persons. Parkinson disease or other movement disorders may be mistakenly diagnosed.

Psychiatric Presentation In psychiatric presentations, changes in personality (irritability, anger, poor self-control), depression, and anxiety are common symptoms. Typically, patients presenting in this fashion are in their late teens or early twenties, a period during which substance abuse is also a diagnostic consideration. Wilson's disease should be formally excluded in all teenagers and young adults with new-onset psychiatric signs.

Ocular Manifestations The eye is a primary site of copper deposition in Wilson's disease, producing a pathognomonic sign, the Kayser-Fleischer ring (**Fig. 415-1**), a golden to greenish-brown band in the peripheral cornea. This important diagnostic sign first appears as a superior crescent and then develops inferiorly and ultimately becomes circumferential. Slit-lamp or optical coherent tomography examinations are required to detect rings in their early stage of formation. Copper can also accumulate in the lens and produce "sunflower" cataracts. Approximately 95% of Wilson's disease patients with neurologic signs manifest the Kayser-Fleischer ring compared to ~65% of those with hepatic presentations. Copper chelation therapy causes fading and eventual disappearance of corneal copper.