

## Brain atrophy and neurological impairment in Wilson's disease

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**Objective:** To determine whether brain volume was associated with neurological and functional impairments and with copper overload markers in patients with Wilson's disease. **Methods:** In 48 treatment-naïve patients, we assessed neurological and functional impairments, with the Unified Wilson's Disease Rating Scale; measured normalized brain volumes, based on magnetic resonance images; and assessed copper overload indices, i.e., the concentration of non-ceruloplasmin-bound copper and the presence of corneal copper deposits. We correlated brain volume measures with neurological and functional impairment scores and copper overload indices. **Results:** Neurological and functional impairments correlated with all brain volume measures, including the total brain volume and the volumes of white matter and grey matter (both peripheral grey matter and deep brain nuclei). Moreover, higher non-ceruloplasmin-bound copper concentrations were associated with lower brain volumes, and patients with corneal copper deposits had significantly lower brain volumes than patients without these deposits. **Conclusions:** Our findings provided the first *in vivo* evidence that the severity of brain atrophy is a correlate of neurological and functional impairments in patients with Wilson's disease and that brain volume could serve as a marker of copper toxicity.