

Transcranial brain parenchyma sonography in Wilson's disease and healthy controls: quantification and correlations with clinical parameters

G. G. Tribi, M. C. Trindade, K. J. Almeida, R. C. Alves, D. C. de Andrade, E. T.

Fonoff, M. J. Teixeira, E. R. Barbosa, E. J. L. Alho, E. Bor-Seng-Shu

Division of Neurology and Neurosurgery, University of Sao Paulo (USP) School of Medicine, Brazil

Transcranial sonography (TCS) allows easy access, rapid, low-cost, and radiation free imaging of brain parenchyma. In Wilson's disease (WD), TCS has shown an even higher sensitivity than conventional MRI. We compared TCS findings in patients with predominantly neurological, treated Wilson's disease (n=40/m=22) and healthy, matched controls (n=49/m=29) and correlated TCS findings with clinical data, including: serum copper and iron parameters, Unified Wilson's Disease Rating Scale (UWDRS), Addenbrooke's Cognitive Examination-Revised, Mini Mental State Examination, and Beck Depression Inventory (BDI). Highly significant differences between patients and controls were found for the hyperechogenicity of lenticular nucleus, substantia nigra, thalamus, and midbrain tegmentum/tectum, and for the values of the midbrain axial area and of the third ventricle width. The echogenic area sizes of lenticular nucleus, substantia nigra, and caudate nucleus allowed highly accurate discrimination between patients and controls (areas under the curve 95.4%, 79.4%, and 80.5%, respectively). In WD, both dystonia and dysarthria correlated with the hyperechogenicity of lenticular nucleus, the midbrain axial area, and the third ventricle width. Parkinsonism, UWDRS total and neurological scores, and reduced cognitive performance correlated with an increased third ventricle width. Symptoms of depressed mood (BDI15) correlated with a reduced midbrain axial section area. Male sex correlated with substantia nigra echogenic area and serum ferritin levels. TCS hyperechogenicity can clearly differentiate treated WD patients with neurological manifestations from healthy subjects. Particular TCS findings correlate with various dimensions of WD symptoms. The discriminative capacity of TCS in detecting brain involvement should be further tested in early-stage WD cohorts.