

To investigate the deep-gray nuclei susceptibility-weighted imaging filtered phase shift in patients with Wilson's disease

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Target Audience: Radiologists, neurologists and pediatricians

Purpose: The aim of this study was to evaluate the susceptibility-weighted imaging filtered phase shift in brain gray nuclei of Wilson's disease.

Methods: 23 Wilson's disease (WD) patients (15 males and 8 females, range = 6 - 35 years, mean = 18.48 ± 8.42 years) and 23 age- and gender-matched healthy controls were recruited. All the WD patients were diagnosed basing on their clinical examinations, laboratory and MRI findings. All the images were performed on a 3.0-T system (Signa HDx, GE Medical System, USA) equipped with a standard eight-channel head coil. The susceptibility-weighted images (SWI) were performed parallel to the AC-PC line and covered the nuclei of the basal ganglia and mesencephalon. The protocols was set as follows: repetition time (TR) = 38 ms, echo time (TE) = 25 ms, flip angle = 20° , matrix = 448×256 (Nx × Ny), FOV = 24 cm × 24 cm. The data processing were operated on a separate workstation (ADW4.3, GE), phase images were high-pass-filtered to create corrected phase images with a central matrix size of 64×64 , in order to get corrected phase image. The regions of interest were identified and hand-drawn according to the anatomical structures by a trained neuroradiologist. Phase values were measured for the bilateral brain gray nuclei on corrected phase image.

Results: Compared with healthy controls, WD patients showed a trend of negative phase shift in all regions of interest, and significantly lower phase value was found in bilateral PU (left $p = 0.007$, right $p = 0.006$), CA (left $p = 0.011$, right $p = 0.001$), TH (left $p < 0.001$, right $p = 0.009$), RN (left $p = 0.029$, right $p = 0.027$) and SN (left $p = 0.003$, right $p = 0.024$), neither left nor right GP ($p > 0.05$) showed significant difference.

Discussion: Former studies have confirmed the negative relationship between paramagnetic mineralization and phase value. So the negative phase shift of our study indicating that there is paramagnetic mineralization primarily deposit in these regions. Copper and iron are paramagnetic ion. Wilson's disease is a copper overloading disease, and is considered there is iron accumulation in WD brain. One reasonable possibility is that the negative phase shift may be the result of copper and iron interaction.

Conclusion: Abnormal negative phase value was significantly increased in brain gray nuclei of WD patients. That gives evidence about paramagnetic mineralization accumulating in brain gray nuclei in WD.

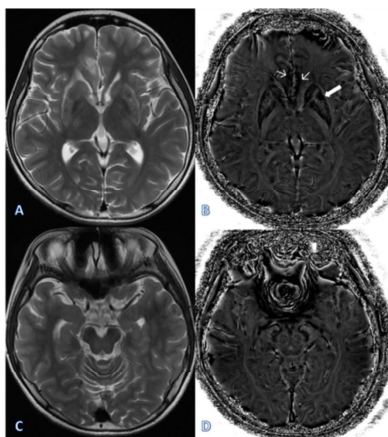


Fig 1: T2-weighted axial MR images (A, C) reveal bilateral symmetric high signal intensity in bilateral caudate, putamen and the tegmental area of midbrain, as well as the lateral margin of putamen, thalamus. SW corrected phase images (B, D) show markedly dark paramagnetic signals in lentiform nucleus, caudate, substantia nigra, red nucleus. In lentiform nuclei, multiple concentric dark foci (thick arrow) and dark stripes on bilateral frontal cortex (thin arrow) are noted.

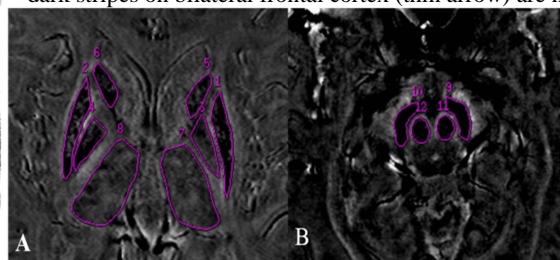


Fig 2: Display the basal ganglia (A) and the midbrain (B) of a patient with WD, drawing 12 gray matter areas as representative regions of interest. 1, 2 = bilateral PT, 3, 4 = bilateral GP, 5, 6 = bilateral CA, 7, 8 = bilateral TH, 9, 10 = bilateral SN, 11, 12 = bilateral RN.

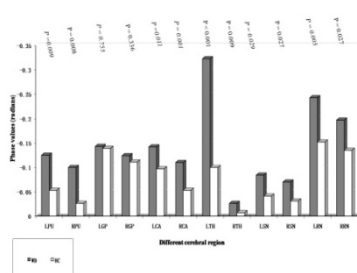


Fig 3: Phase shift of bilateral ROIs between WD and healthy controls. Gray column corresponds to the mean phase value of WD patient ($n = 23$) and white column corresponds to the mean phase value of healthy controls ($n = 23$), $p =$ significant difference of phase shift of ROIs between WD and healthy controls ($p < 0.05$, two-tailed t-test). All the phase value data measured from ROIs of WD patients were lower than those of control subjects (table 1, Fig 5), and significantly negative phase shift was found in bilateral PU, CA, TH, RN and SN. There is no significant difference in left and right GP.