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## Cerebral abnormalities in myotonic dystrophy. Cerebral blood flow, magnetic resonance imaging, and neuropsychological tests

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**OBJECTIVE**--To study **cerebral abnormalities in myotonic dystrophy** (MD) and determine the different patterns of **cerebral** function **in** patients with MD with maternal (mMD) vs paternal (pMD) **in**heritance. **DESIGN**--Patients with MD and normal controls were studied with neuropsychological testing, magnetic resonance imaging, and single photon emission computed tomography. **SETTING**--Studies were done at Harbor-UCLA Medical Center, Torrance, Calif. **PATIENTS AND OTHER PARTICIPANTS**--Twenty-two consecutive-patients with MD, 11 of whom had pMD and eight mMD, and 10 normal controls were studied. Diagnoses were made on the basis of family history, electromyography, and clinical examinations. Normal subjects **in** the same age distribution were studied for comparisons. **RESULTS**--We found significantly lower neuropsychological performance and **cerebral** blood flow **in** the patients with MD compared with the controls. Patients with mMD had statistically lower scores on IQ tests and more extensive **cerebral** hypoperfusion when compared with those with pMD. **Changes in cerebral** blood flow were most severe **in** the frontal and temporoparietal association cortex. **Cerebral** blood flow measures strongly correlated with IQ. **CONCLUSIONS**--Patients with mMD had earlier onset of disease and lower IQs than the pMD group. The pattern of **cerebral** perfusion **in** the mMD group was consistent with a diffuse brain injury, while **cerebral** perfusion **in** pMD showed more **minor changes**. These findings emphasize the cognitive differences between mMD and pMD.

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