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[BioMedicine] Taiwan Biomedical Researchers Show Hearing Impairment Is Authentic Symptom of Huntington's Disease

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Academia Sinica Newsletter (2011/03/22) A team of biomedical scientists from Academia Sinica, National Yang-Ming University, Tri-Service General Hospital, Chang Gung Memorial Hospital, and the Veterans General Hospital recently reported that hearing loss is an authentic symptom of Huntington's disease (HD). In addition, the team found that brain-type creatine kinase (CKB), an enzyme important in supplying energy to cells, was reduced in the cochlea in the inner ear of mice with HD. Most importantly, treatment with creatine supplements ameliorated the hearing impairment of HD mice, hinting that creatine may be an effective treatment for hearing problems in HD patients. The study was published in the Journal of Clinical Investigation on March 14, 2011.

Huntington's disease (HD) is a neurodegenerative disorder that usually becomes established in middle age. Clinical features of HD include uncontrollable writhing movements known as chorea, cognitive impairment, and psychiatric syndromes. HD is caused by an autosomal dominant mutation on either of an individual's two copies of a gene called Huntingtin, which means that any child of parent with HD has a 50% risk of inheriting the disease. The worldwide prevalence of this disorder is estimated to be 5-10 cases per 100000 people.

In the study led by Dr. Yijuang CHERN, a Research Fellow at the Institute of Biomedical Sciences, Academia Sinica, and Dr. Chih-Hung WANG, a clinician of Tri-Service General Hospital, 19 HD patients (aged 40–59) were assessed for hearing loss using pure-tone audiometry (PTA) and auditory brainstem responses (ABR). Hearing loss was found to be considerably greater in HD patients. Two mouse models of HD also showed hearing loss when tested in the same way as humans, leading researchers to conclude that hearing loss is an authentic symptom of HD.

Analyses showed that mutant huntingtin (Htt) was present in the organ of Corti (the spiral organ of the inner ear) of HD mice, which might interfere with its normal functioning. Further analyses revealed reduced expression of the enzyme CKB in the cochlea of HD mice. Treatment with creatine supplements improved the hearing impairment of HD mice, suggesting that creatine may be useful for treatment of hearing abnormalities in HD patients.

The full-text of the study entitled "Dysregulated brain creatine kinase is associated with hearing impairment in mouse models of Huntington disease" is available at the Journal of Clinical Investigation website at:
http://www.the-jci.org/publiTron.php?series_id=71&action=review_series

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Further Information:
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