Possible therapy for one form of inherited nerve dysfunction

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Hereditary neuropathies are common nervous system conditions characterized by progressive loss of muscle control and/or sensory function. There are no effective treatments. However, work in mice, by a team of researchers led by Florian Eichler, at Massachusetts General Hospital and Harvard Medical School, Boston, has revealed a rational candidate oral therapy for one hereditary neuropathy -- hereditary sensory and autonomic neuropathy type 1 (HSAN1).

HSAN1 is thought to be caused by the accumulation of molecules known as deoxysphingolipids, which are toxic for cultured sensory neurons. Eichler and colleagues found that oral administration of L-serine to mice with a disease that models HSAN1 reversed the accumulation of deoxysphingolipids and reduced the severity of neuropathy.

As the accumulation of deoxysphingolipids in humans with HSAN1 was also reversed by oral administration of L-serine, these data provide a rationale for the authors suggestion that L-serine supplementation could provide a treatment option for HSAN1.

In an accompanying commentary, Steven Scherer, at the University of Pennsylvania, Philadelphia, agrees with Eichler and colleagues that a clinical trial of L-serine as a treatment for HSAN1 should be done but he cautions patients to remember that the approach has not yet been proven to be effective.

More information: Oral L-serine supplementation reduces production

of neurotoxic deoxysphingolipids in mice and humans with hereditary sensory autonomic neuropathy type 1, www.jci.org/articles/view/5754 … 2bc381dd8e09d15886c4