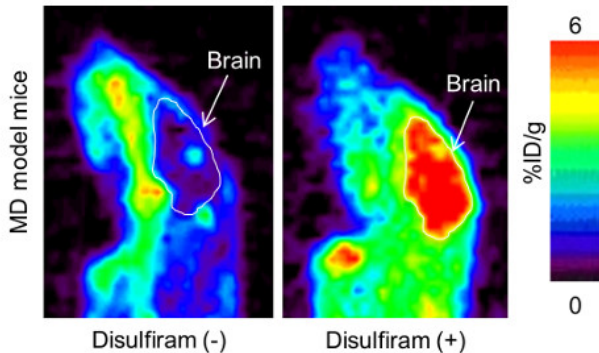


# Using PET scanning to evaluate therapies of Menkes disease

26 March 2014



Sagittal section of brain PET image at 4 hrs after  $^{64}\text{CuCl}_2$  injection with disulfiram or D-penicillamine in MD model mice.

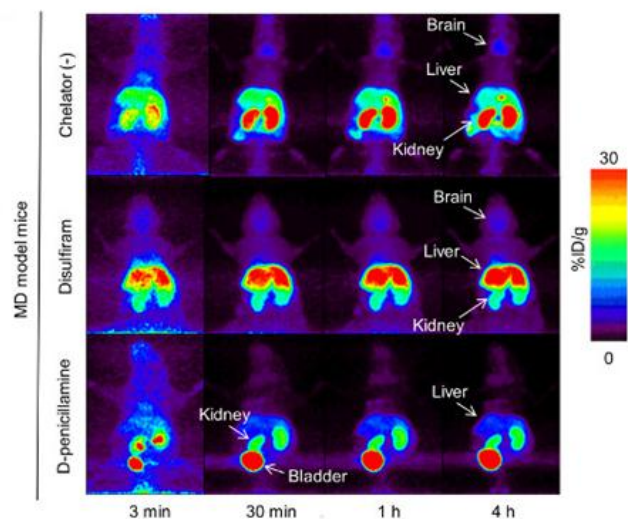
Scientists at the RIKEN Center for Life Science Technologies have used PET imaging to visualize the distribution in the body of copper, which is deregulated in Menkes disease, a genetic disorder, using a mouse model. This study lays the groundwork for PET imaging studies on human Menkes disease patients to identify new therapy options.

Menkes disease, though rare, is a fearsome [genetic disorder](#). Most affected babies die within the first few years of life. The disease is caused by an inborn fault in the body's ability to absorb [copper](#). The standard treatment today for the 1 in 100,000 babies affected by the disorder is to inject copper, but this therapy has limited efficacy. Eventually the treatment becomes ineffective, leading to neurodegeneration, and the copper accumulates in the kidneys, sometimes leading to renal failure.

As a result, treatments have been sought that enhance the accumulation of injected copper in the brain while preventing its accumulation in the

kidney. Recently, disulfiram, a drug developed to treat alcoholism, has been suggested as a therapy for Menkes disease, since one of its actions is to enhance this copper accumulation in the brain.

Now, in a landmark study published in the *Journal of Nuclear Medicine*, scientists at the RIKEN Center for Life Science Technologies in Kobe, Japan, in collaboration with paediatricians from Osaka City University and Teikyo University, have used [positron emission tomography](#) (PET) to show that a combination of copper injections and disulfiram or D-penicillamine allows a greater movement of copper to the brain, where it is needed, without accumulating in the kidneys.



Coronal section of whole-body PET image at 4 hrs after  $^{64}\text{CuCl}_2$  injection with disulfiram or D-penicillamine in MD model mice.

In the study, the researchers used Menkes Disease model mice, which have an inborn defect in [copper metabolism](#), and injected copper-64, a radioactive

isotope of copper, into the mice. They then used PET scanning, a non-invasive procedure, to visualize how the copper moved throughout the body. They compared mice injected with copper alone to mice injected with copper along with one of two other drugs, disulfiram or D-penicillamine, and the distribution of the copper throughout the body was observed for a four-hour period.

The results showed that the mice given copper along with disulfiram had a relatively high concentration of copper in the brain without a significant increase in the kidneys. Surprisingly, it showed that the amount of copper going to the brain in mice treated with disulfiram was actually higher than in those treated with copper alone, suggesting that the drug has an effect on the passage of copper through the blood-brain barrier.

According to Satoshi Nozaki, one of the co-authors, "This study demonstrates that PET imaging can be a useful tool for evaluating new treatments for Menkes disease." Looking to the future, he said, "Based on this study, we are planning to conduct clinical PET studies of patient with Menkes disease."

**More information:** Shiho Nomura, Satoshi Nozaki, Takashi Hamazaki, Taisuke Takeda, Eiichi Ninomiya, Satoshi Kudo, Emi Hayashinaka, Yasuhiro Wada, Tomoko Hiroki, Chie Fujisawa, Hiroko Kodama, Haruo Shintaku, Yasuyoshi Watanabe, "PET Imaging Analysis with  $^{64}\text{Cu}$  in Disulfiram Treatment for Aberrant Copper Bio-distribution in Menkes Disease Mouse Model", *The Journal of Nuclear Medicine*, 2014, [DOI: 10.2967/jnumed.113.131797](https://doi.org/10.2967/jnumed.113.131797)

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