

International team discover clue to Friedreich's ataxia, devastating nervous system disease

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(Medical Xpress)—A new form of iron may hold the clue that leads to treatment for a fatal inherited nervous system disease that can cause gait disturbance, speech problems, heart disease, diabetes and other symptoms.

In a study published in [Proceedings of the National Academy of Sciences](#) today, researchers unravel a thread of evidence about Friedreich's ataxia (FA), a rare condition that strikes when sufferers are aged from five to 15.

Co-authors Professor Tim St Pierre and Dr Lucia Gutierrez of The University of Western Australia's BioMagnetics Group, are internationally acclaimed experts in the roles of iron in the body and ways to measure and detect iron.

With PhD student Adam Fleming at UWA and fellow researchers from the University of Sydney led by Professor Des Richardson, as well as institutions in Canada and Spain, they have found a yet-to-be named new form of iron in the heart of the mice with the disease.

"The iron looks like a mineralised form of iron and phosphate," Professor St Pierre said. "It's a type of rust that isn't an iron oxide."

The UWA team and their colleagues around the world used techniques including Mössbauer spectroscopy and magnetic susceptibility measurements to try to identify the iron-containing substance that was initially shown up under [electron microscopy](#). The experiments were carried out in the livers and hearts of affected mice at temperatures as low as 5 Kelvin, or minus 268 degrees Centigrade.

In FA patients there is an absence or reduction of the iron-[binding protein](#) frataxin in the cells, Professor St Pierre said.

"A change in [iron metabolism](#) has been suspected to be part of the model of damage in patients, whose cells' mitochondria ([cellular energy](#) supply) have more iron than usual while there is less than usual in the cytosol (liquid found inside cells)," he said. An excess of iron where it shouldn't be can cause problems, while iron deficiency also causes problems. Iron excess in particular organs causes an accumulation of damaging free radicals."

More information: www.pnas.org/content/early/2011/12/15/1215349109.abstract

Provided by University of Western Australia

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