Plant extract hope for infant motor neurone therapy

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(Medical Xpress)—Researchers from Plymouth University Peninsula Schools of Medicine and Dentistry have been part of an international team led by the University of Edinburgh, who have identified that a chemical found in plants could reduce the symptoms of a rare muscle disease that leaves children with little or no control of their movements.

Their study is published today (3rd March 2014) in the *Journal of Clinical Investigation*.

The research team have found that a plant pigment called quercetin – present in some fruits, vegetables, herbs and grains – could help to prevent damage to the nerves associated with the childhood form of motor neurone disease, spinal muscular atrophy (SMA).

SMA, also known as 'floppy baby syndrome', is a leading genetic cause of death in children. It affects approximately one in 6,000 to 10,000 children and around half of children with the most severe form will die before the age of two. There is currently no cure for this kind of neuromuscular disorder.

The condition is caused by a mutation in a gene that is vital for the survival of nerve cells that connect the brain and spinal cord to the muscles, known as motor neurons. Until now, it was not known how the mutation damages these cells and causes disease.

The study reveals that the mutated gene affects a key housekeeping

process that is required for removing unwanted molecules from cells in the body. When this process doesn't work properly, molecules can build-up and cause problems inside the cells.

Children with SMA experience progressive muscle wastage and loss of mobility and control of their movements. The disorder is often referred to as 'floppy baby syndrome' because of the weakness that it creates.

The team has found that the build-up of a specific molecule inside cells – called beta-catenin – is responsible for some of the symptoms associated with the condition.

In tests on zebra fish, mice and fruit flies (the latter in Plymouth), scientists found that treating the disease with quercetin – which targets beta-catenin – led to a significant improvement in the health of nerve and muscle cells.

Although quercetin did not prevent all of the symptoms associated with SMA, researchers hope that it could offer a useful treatment option in the early stages of disease.

Professor Tom Gillingwater from the University of Edinburgh, who led the study, said: "This is an important step that could one day improve quality of life for the babies affected by this condition and their families. There is currently no cure for this kind of neuromuscular disorder so new treatments that can tackle the progression of disease are urgently needed."

Dr. Iain Robinson, Associate Professor in Neuroscience at Plymouth University Peninsula Schools of Medicine and Dentistry, added: "By working to understand how SMA operates at a molecular level we have been able to apply a naturally occurring compound which targets the one of the key culprits, with the potential to introduce an effective drug

therapy for children in the early stages of the disease."

Provided by University of Plymouth