Methods of Inducing and Preventing Neurofibromatosis in Schwann Cells

Background
The neurofibromatoses are genetic disorders of the nervous system that primarily affect the development and growth of neural cell tissues. These disorders cause tumors to grow on nerves and produce other abnormalities such as skin changes and bone deformities. Although many affected persons inherit the disorder, between 30 and 50 percent of new cases arise spontaneously through mutation. These disorders have been classified into two types: neurofibromatosis type 1 (NF1) and neurofibromatosis type 2 (NF2). Neurofibromatosis type 2 is caused by a mutation on chromosome 22q12 in the nf2 gene that lead to loss of expression of schwannomin (Sch), also called merlin, which is a tumor suppressor. Symptoms include, acoustic neuromas on the vestibulocochlear nerve leading to hearing loss. Patients with NF2 may also develop other brain tumors, as well as spinal tumors. The current invention describes a model system for identifying new therapies for NF2. This invention also describes a method for preventing inactivation of the Schwannomin’s tumor suppressor activity in Schwann cells.

Invention
The invention provides a method for inducing neurofibromatosis type 2 (NF2) in Schwann cells for testing of new treatment drugs, as well as a method for preventing the inactivity of tumor suppressors via a blocking agent, tyrphostin AG825.

Application
The invention can provide a model for neurofibromatosis (NF2) which would allow for testing of new therapeutic agents.

Advantages
• Provides reliable laboratory methods to induce NF2 modeling.
• The blocking agent is a possible agent for further pharmaceutical options.

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Selected References

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