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(54) Title: COMPOUNDS AND METHODS FOR MODULATING ATXN1

(57) Abstract: Provided are compounds, methods, and pharmaceutical compositions for reducing the amount or activity of ATXN1 RNA in a cell or subject, and in certain instances reducing the amount of ATXN1 in a cell or subject. Such compounds, methods, and pharmaceutical compositions are useful to ameliorate at least one symptom or hallmark of a neurodegenerative disease. Such symptoms and hallmarks include gait and limb ataxia, cognitive impairments, difficulty with speaking and swallowing, atrophy of the cerebellum and brainstem in magnetic resonance imaging (MRI), neurochemical abnormalities in the cerebellum and brainstem detected via magnetic resonance spectroscopy (MRS), and death within 10-15 years of symptom onset. Such neurodegenerative diseases include Spinocerebellar ataxia type 1.