

# Margaret Ibe, Biology

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Faculty Mentor: Dr. Martin Katz, Ophthalmology  
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## Behavioral assessment of mouse models of neurodegenerative disorders

The neuronal ceroid lipofuscinoses (NCLs) are a family of progressive hereditary neurodegenerative disorders characterized by accumulation of lysosomal storage bodies, cognitive decline, loss of motor function and premature death. We are using mouse models of the NCLs to develop therapies for these diseases. To assess the effectiveness of therapies in treating the NCLs, quantitative measures of phenotypic disease features are needed. We hypothesized that mice with NCL would show deficits in associative learning abilities. To determine whether this is the case, we compared the ability of affected and normal mice to associate the presence of light with a food reward using a T-maze. Mice in 5-7 week, 9-11 week, and 14-15 week age cohorts for both subject groups were tested. Affected mice performed significantly worse than age-matched controls in every age cohort examined, even though no overt signs of behavioral deficits were apparent in any of the ages tested. Our results indicate that the associative learning test is a sensitive phenotypic indicator in the NCL mouse model that will be useful in assessing the effectiveness of therapeutic interventions. This test will likely be useful in characterizing other mouse models of neurodegeneration as well.

This project was completed as part of a Capstone requirement.