Molecular and Behavioral Characterization of surf1-/- Zebrafish

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Leigh Syndrome (LS) is a rare mitochondrial disorder caused by mutations in the SURF1 gene, leading to reduced Complex IV enzymatic activity, impaired oxidative phosphorylation, and severe motor deficiencies. This study aims to develop a zebrafish (Danio rerio) model for LS using CRISPR/Cas9-generated surf1 mutants. Key methodologies included genotyping, DNA extraction, Polymerase Chain Reaction (PCR), gel electrophoresis, molecular imaging, and activity assays to assess motor function. A total of 168 zebrafish from a heterozygous incross were genotyped, identifying 29 mutants. Activity assays revealed a significant reduction in swimming activity among Day 5 surf1 mutants compared to their wild-type and heterozygous siblings, with one one of the Day 3 rep 1 having a similar trend with no significant and Day 3 rep 2 having significance. These findings suggest that surf1 mutants exhibit motor impairments, supporting their potential as an LS model. Future research will further investigate these behavioral deficits to replicate results and assess antioxidant therapies' effects using the mutants as models.

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