New Results

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Hexafluoro slows retinal degeneration and improves visual function in zebrafish models of Usher syndrome 1F

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ABSTRACT

Usher syndrome is the leading genetic cause of deafblindness, affecting hundreds of thousands of people worldwide. The deafness can be addressed with hearing aids or cochlear implants, but there is currently no treatment for the vision loss, which is due to progressive degeneration of retinal photoreceptors. Studies in animal models of Usher syndrome have shown that photoreceptor degeneration is exacerbated by exposure to bright light, and other studies have shown that light-induced photostress reduces mitochondrial function. We previously synthesized hexafluoro and showed that it is a potent Sirt3 activator that promotes mitochondrial respiration. Here we examined the efficacy of hexafluoro as a potential therapeutic for treatment of vison loss in a zebrafish model of Usher syndrome type 1F, which exhibits early and severe vision defects along with vestibular dysfunction as seen in Usher type 1 pathology. We find that hexafluoro improves visual function, reduces photoreceptor degeneration, and protects the retina against exposure to bright light in this USH1F model.

Competing Interest Statement

The authors have declared no competing interest.