Wolfram syndrome is caused by spelling mistakes (mutations) in a gene known as *WFS1* (4p16.1, OMIM 606201) and the majority of people with this condition experience progressive loss of vision from early childhood. This is due to damage to a group of specialised cells in the retina that help transmit visual information from the eye to the vision centres in the brain. These specialised cells are known as "retinal ganglion cells" (RGCs) and as they exit the back of the eye, they form the optic nerve. As these RGCs are gradually lost, the optic nerve becomes pale (optic atrophy) and vision starts to deteriorate. Wolfram syndrome is an important cause of severe visual impairment among children and young adults and there is currently no treatment to prevent vision from getting worse.

The *WFS1* gene affects a protein (wolframin) located within a specific compartment of the cell known as the "endoplasmic reticulum" (ER). It is thought that the lack of wolframin has a major detrimental impact on mitochondria, which are the powerhouses responsible for producing most of the energy that a cell needs to survive. Research into Wolfram syndrome has not yet clearly revealed the mechanisms by which *WFS1* mutations cause cell death and ultimately loss of vision. In order to achieve a research breakthrough, we need a much better understanding of the interactions between the ER and mitochondria, as well as better animal models for testing the efficacy of potential drug treatments for Wolfram syndrome.

After spending much effort in the lab, we have developed a zebrafish model of Wolfram syndrome that replicates the RGC loss and visual dysfunction seen in individuals with this genetic condition. The availability of a zebrafish model for Wolfram syndrome provides a powerful tool to better understand the disease mechanisms that result in RGC loss and to screen for drugs that could potentially block this from happening in an effort to preserve vision.