Title: Targeting inflammation to rescue cone photoreceptors in Retinitis Pigmentosa (RP)

Abstract: Recent studies demonstrate a major pathogenic role of inflammatory processes in neurodegenerative diseases of the Central Nervous System (CNS), even when the aetiology is not primarily inflammatory. An acute phase of microglial activation needed for the effective removal of misfolded proteins and dead cells is followed by a local release of toxic cytokines that, in turn, trigger an escalating, vicious cycle of inflammation, which becomes chronic and worsens the phenotype.

Data from this laboratory demonstrate an unsuspected role of the inflammatory and immune response in the progression of Retinitis Pigmentosa, a disease that is not inflammatory in aetiology but in which a strong molecular signature left in the retina by the inflammatory and immune system responses is revealed by transcriptome analysis. Our working hypothesis is that this response, carried on by reactive retinal microglial and monocytes infiltrating from the blood stream, contribute to create a microenvironment hostile to cell survival, ultimately leading to the bystander death of retinal cones and to blindness. We are developing several anti-inflammatory strategies, based upon administration of trophic factors, natural molecules and steroids, to decrease the inflammatory-immune response, promoting the integrity of the blood-retinal barriers and delaying the degeneration of cones, thus prolonging the time window of useful vision.

While we work on state-of-the art mouse models of RP, our clinicians collaborators lead parallel trials on human RP subjects, evaluating the integrity of retinal barriers with high resolution imaging and testing the most promising strategies for the treatment of this (still orphan) disease.

References:


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