



Spotlight on vision

Foreword

Retina mediates the first steps in vision. All pathological events that affect the photoreceptor structure and function result in impaired vision. Photoreceptor degeneration of either genetic or age-related origin is a major cause of vision impairment and blindness worldwide. Inherited retinal dystrophies (IRDs) affect approximately one in every 3000 individuals and represent the most frequent inherited forms of human visual handicap. Age-related macular degeneration (AMD) is a leading cause of irreversible blindness and visual disability in the elderly. Up to one third of people aged 75 and older suffer from a form of AMD and, owing to the rapidly ageing population, this figure is projected to double in the next decades. Except for the neovascular complications of age-related macular degeneration, there is no known effective treatment that can prevent or reverse the vision loss in degenerative eye diseases. To address this largely unmet medical need, a large array of innovative therapies is under development.

This thematic issue of the *Comptes rendus de l'Académie des sciences* was thus inspired by the recent advances in basic, clinical and translational vision research. It aims at emphasizing the value and importance of integrative multidisciplinary approaches in preventing retinal degeneration diseases and restoring vision. It represents only a sample of the numerous research programs conducted in several laboratories and others that were invited but could not contribute to this issue.

The past two decades have been marked by exceptional progress in the understanding of the retinal biology and pathophysiology, and the identification of the genes and mutations underlying retinal degeneration events. The main themes of this thematic issue focus on mechanisms mediating the highly organized retinal layering, as well as on the genetic causes of IRDs and retinal phenotype of Usher syndrome, the most common genetic cause of combined auditory–visual impairment. Articles in this issue highlight promising innovative approaches to prevent, treat, and

manage blinding diseases, including retinitis pigmentosa and other IRDs, Leber hereditary optic neuropathy, and AMD. Special attention was dedicated on the potential and challenges of gene therapy for retinal disorders. The first safety and efficacy clinical studies of gene therapy in patients with Leber congenital amaurosis provided consistent foundation for gene therapy approaches for other forms of retinal degeneration, such as choroideremia, Stargardt macular degeneration and Usher syndrome type 1B. The emerging concept of “mutation-independent” approaches to save vision is discussed and exemplified by the discovery of rod-derived cone viability factor (RdCVF), which has been shown to induce cone survival and to preserve the function of cone photoreceptors in animal models of inherited retinal dystrophies, and is now in translation as a potential therapeutic agent. Restoring vision by retinal prosthesis has been investigated for several years now and demonstrated gain of some vision in blind individuals. Combining different therapeutic strategies, e.g., gene replacement/optogenetics/retinal prosthesis/neurotrophic compounds, may lead to enhancing the therapeutic potential of these emerging therapeutic solutions.

This thematic issue of the *Comptes rendus de l'Académie des sciences* would not have been possible without the efforts of our contributors and colleagues, including the patient work of Katia Marazova and of the journal editors. We hope that it will demonstrate the vitality and potential of ophthalmic research.

Disclosure of interest

José-Alain Sahel is a founder and consultant for Pixium Vision and GenSight Biologics, and a consultant for Sanofi-fovea and Genesignal.

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