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OPA1 functions in mitochondria and dysfunctions in optic nerve

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Titre	OPA1 functions in mitochondria and dysfunctions in optic nerve
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Auteur	Lenaers, Guy [1], Reynier, Pascal [2], El Achouri, Ghizlane [3], Soukkarieh, Chadi [4], Olichon, Aurélien [5], Belenguer, Pascale [6], Baricault, Laurent [7], Ducommun, Bernard [8], Hamel, Christian [9], Delettre, Cécile [10]
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Résumé en anglais	OPA1 is the major gene responsible for Dominant Optic Atrophy (DOA), a blinding disease that affects specifically the retinal ganglion cells (RGCs), which function consists in connecting the neuro-retina to the brain. OPA1 encodes an intra-mitochondrial dynamin, involved in inner membrane structures and ubiquitously expressed, raising the critical question of the origin of the disease pathophysiology. Here, we review the fundamental knowledge on OPA1 functions and regulations, highlighting their involvements in mitochondrial respiration, membrane dynamic and apoptosis. In light of these functions, we then describe the remarkable RGC mitochondrial network physiology and analyse data collected from animal models expressing OPA1 mutations. If, to date RGC mitochondria does not present any peculiarity at the molecular level, they represent possible targets of numerous assaults, like light, pressure, oxidative stress and energetic impairment, which jeopardize their function and survival, as observed in OPA1 mouse models. Although fascinating fields of investigation are still to be addressed on OPA1 functions and on DOA pathophysiology, we have reached a conspicuous state of knowledge with pertinent cell and animal models, from which therapeutic trials can be initiated and deeply evaluated.
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