Author Response: Increased mtDNA Copy Number Protects Against LHON

Although we appreciate the major interest of Josef Finsterer and Sinda Zarrouk-Mahjoub in our recent publication in *Investigative Ophthalmology and Visual Science* entitled "High Mitochondrial DNA Copy Number Is a Protective Factor from Vision Loss in Heteroplasmic Leber's Hereditary Optic Neuropathy (LHON)," we wish to clarify two issues. First, in our published studies, our findings strongly support the concept that in unaffected LHON subjects there is an increase of mitochondrial DNA (mtDNA) copy number in peripheral blood, as already reported in *Brain* and in *Acta Myologica*, and we also acknowledged a possible contribution of mtDNA haplotype. Our working hypothesis is in keeping with work by others. A recent review is also in line with our data.

Second, we did not overlook that idebenone is a useful drug in LHON during the acute stage of the disease, thereby preventing further vision loss and promoting recovery of vision. We agree that commencing treatment shortly after the onset of symptoms is likely to have the best therapeutic effect, whereas it is hard to weight the usefulness of therapy to treat the healthy relatives.

We hope that we have fully clarified our opinion.

Angelica Bianco  
Luigi L. Palese  
Silvana Guerriero  
Vittoria Petruzzella

Dipartimento di Scienze Mediche di Base, Neuroscienze e Organi di Senso - Università degli Studi Aldo Moro, Piazza G. Cesare, Bari. E-mail: vittoria.petruzzella@uniba.it

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