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| **Date** | **Relevant past medical history** | |
|  | Low vision in the left eye since childhood due to esodeviation which was never operated on. History of arterial hypertension. Two relatives on the mother’s side in the family had a similar episode of vision loss and then improvement at a younger age. Unfortunately, these relatives live in another country and were not available for screening and genetic testing. | |
|  | **Summaries from initial and follow-up visits** | **Interventions and therapy** |
| June 2010 | VA loss on the previously healthy right eye |  |
| June 2010  (2-3 weeks after the onset of the right eye) | Hospitalization at Clinic. VA RE counting fingers on 3 meters, LE: counting fingers at 2,5 m Color vision RLE 0/15. VF Bilateral visual field constriction of 15-20 deg, enlarged right blind spot and excluded left blind spot. On fundoscopy right optic disc was pink with sharp borders, left slightly paler temporally FA: no leakage, EF: P 50 amplitude was reduced, lower on amblyopic eye, The N95 was decreased, agin more on the amblyopic eye decreased N95 wave more on the amblyopic, N95/P50 ratio RE 1.52 LE:1.27, delayed and decreased VEP P100 more on the amblyopic eye. | Systemic corticosteroid therapy for 3 days Solumedrol i.v. (1g/day), no improvement.  MRI of the head and brain, no signs of demyelination, aqp4 negative, antiMOG negative, excluded all other possible signs of theoptic atrophy (infectious, paraneoplastic, compressive etc) |
| October 2010 (4 months after the onset) | VA RLE 0,1 Color vision RLE 1/15, VF REscotoma in lower half of visual field LE scotoma on nasal visual field. On fundoscopy both optic discs were still pink. Decreased sensitivity and extentric fixation on microperimetry. | Bilateral amblyopia suspected due to the absence of the optic disc pallor |
| June 2011  (12 months after the onset) | VA RLE 0,1 Color vision RLE 1/15, central scotoma bilaterally. On fundoscopy temporal pallor of the optic discs started to show, thinning of the inner retina on OCT bilaterally and thinning of the pRNFL on the RE T ans TS, LE temporal half of the optic disc. | Hereditary optic neuropathy was suspected, and blood taken for genetic analysis |
| December 2011 (1.5 years after the onset) | Slight VA improvement RE 0.2 LE:0.1 Color vision RE 0/15, LE 1/15, smaller scotomas in visual field bilaterally. On fundoscopy both optic discs pale, thinning of the inner retina on OCT bilaterally, further progression of the pRNFL thinning. | Gentic testing for 3 common mutations revealed presence of the common mutation |
| June 2012  (2 years after the onset) | Significant VA improvement RE 0.8, LE 0.1 Color vision RE 3/15, LE 1/15, significant reduction of the visual field bilaterally. On fundoscopy both optic discs paler, thinning of the inner retina on OCT bilaterally, further progression of the pRNFL thinning, with still preserved nasal segment on both eyes. EF: P 50 amplitude improved, but was still lower on amblyopic eye, The N95 was decreased on the amblyopic eye, normal on the LHON but in the level of the baseline, N95/P50 ratio RE 0.83 LE:0.74, delayed (more on amblyopic) and decreased (more on LHON) VEP P100 wave. |  |
| March 2018  (8 years after the disease onset) | Complete VA recovery on the RE 1.0, LE 0.1 Color vision RE 6/15, LE 1/15, further reduction of the visual field scotomas bilaterally. On fundoscopy both optic discs were paler, with thinning of the inner retina on OCT bilaterally in the macular region, slight progression of the pRNFL thinning in the TI segment on LHON eye, and no progression in other segments in comparison to 2012. EF: P 50 amplitude normal on both eyes, still lower on amblyopic, The N95 was borderline decreased on the amblyopic eye, normal on the LHON but in the level of the baseline, N95/P50 ratio RE 1.04 LE:1.05, borderline delayed and still significantly decreased VEP P100 wave (more on the amblyopic eye). Microperimetry showed significant improvement in sensitivity. |  |

Table S5. Timeline of the disease progression in Patient 3