A 13-year-old girl with a history of nystagmus from 4 to 18 months of age developed intense photophobia. Examination showed poorvision (0.8 logarithm of the minimum angle of resolution in both eyes). Fundus photographs (A, B), autofluorescence imaging (C), andspectral domain OCT (D) showed stable, deep, coloboma-like central macular lesions in both eyes. We observed absent cone responses butnormal rod responses on full-field electroretinogram. Molecular genetic testing detected a pathogenic c.1561C>T (p.Gln521\*) homozygousmutation in the exon 13 of ATF6.